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The Maltese Dental Journal

The Dental Probe



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Editorial

By Dr David Muscat

Dear colleagues,

Following the DAM AGM and the elections we have a new committee. The new committee is as follows

- Dr Noel Manche
President
- Dr Adam Bartolo
Vice President, SAC Representative and substitute representative on the Mater Dei Training Committee
- Dr Thomas Grixti
Treasurer, Federation Representative
- Dr Gabrielle Ferriggi
Secretary, Public Relations Officer
- Dr Nicholas Busuttill Dougall
IT Officer, Representative on the

Mater Dei Education Committee, substitute representative on the SAC

- Dr Simon Muscat
CPD Officer
- Dr Deborah Mifsud
Membership Officer, Second IT Officer
- Dr David Muscat
International Relations Officer, Editor the Dental Probe
- Dr Edward Fenech
Social Events Officer

This is an exciting dynamic pro-active team!

Dr Yvette Zahra was featured in a 'Trailblazers in Health' calendar

highlighting 14 women working in Health. This was launched by the 'Women for Women foundation', an NGO that seeks to support women's well being and economic independence. The calendar celebrates the achievements of exceptional women who are leading the way in the health sector.

The cover picture is by Dr Gabrielle Ferriggi and is entitled 'Longing.'

David

Dr David Muscat B.D.S. (LON)
Editor / Secretary, P.R.O. D.A.M.

The DAM Christmas Party 2024

20th December 2024



SAINT APOLLONIA 2025

The Dental Association of Malta celebrated the feast of St Apollonia at the Church of St Agatha in Rabat. A mass was held followed by a tour of the church and the crypt.

The original church was built in 1504 and was replaced by a larger one on the same site in 1670. In 1894 the statue of Saint Agatha was elevated on a high pedestal above the altar. There is an underground crypt in a chamber on the right hand side of the altar where the remains of The Priest and Missionary Mgr Joseph De Piro – the founder of The Missionary Society of Saint Paul – are found.

The statue of Saint Agatha is the very same that the Knights raised on the Mdina Bastions during a raid by the Ottomans. It is said that on seeing the statue the Ottomans fled. The church was built in her honour and the statue was then transferred from the Church in Mdina to Saint Agatha's church in Rabat.

The crypt under the church was cut into live rock and was a natural cave that was enlarged in the 4th and 5th centuries. There are frescoes in the crypt from the 4th to the 15th centuries.

It is said that Saint Agatha had hidden in the catacombs when she fled Sicily. Saint Agatha was martyred in Catania on 5th February in the year 251AD. Saint Apollonia was martyred in Alexandria Roman Egypt on 9th February in 249 AD.

Saint Agatha is the patron Saint of Bellfounders, breast cancer patients, nurses/wet nurses, bakers, jewellers, rape victims, martyrs, single laywomen, sufferers of sterility, victims of torture, natural disasters, fire, eruptions of Mount Etna and Volcanic Eruptions.

Saint Apollonia is the patron Saint of dentists and dental diseases. The Saint Apollonia Event was organized by the DAM with the cooperation of Father Mark, the DAM Chaplain, and Dr Lino Said. 🇲🇹



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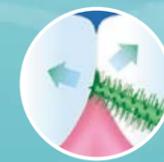
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'Ponte della Malvasia Vecchia' won the bronze medal overall. It was taken in Venice and the idea came from an extremely fleeting moment in Luchino Visconti's film 'Death in Venice'

DR IVAN PADOVANI: A TALENTED DENTIST AND PHOTOGRAPHER

Dr Ivan Padovani took part in the Malta Photographic Society's 59th annual exhibition at the Malta Society of Arts in Valletta. Dr Padovani featured several of his pictures such as 'Pawlu's Field', which reached the finals in the Sony

Global International Competition and was exhibited at Somerset House in London. 'Ponte Della Malvasia' was one of the exhibition's standout images and won third prize overall. The other two were 'Early Morning Serenade' and 'Three Worlds.'



'Early Morning Serenade' was also shot in Venice, on a cold morning in February 2020, literally a few days (less than a week) before the Covid pandemic broke out.



'Pawlu's Field' also grabbed the judges' attention. This one reached the finals in the Sony Global International competition, earlier this year, and was exhibited at Somerset House in London at the time



'Three Worlds' was an opportunity that presented itself out of nowhere, in Delhi, India. It was a rapid grab shot because, in the Islamic world, many men take a dim view of having members of their family photographed

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- During Aligner Insertion
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DONE.

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KEY BENEFITS OF THE CURAPROX ALIGNER FOAM:

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- Plaque-fighting action
- Tackles bad breath
- Moisturises teeth surfaces
- Antibacterial properties
- Refreshing mint flavour



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THE DENTAL ASSOCIATION OF MALTA

Administrative Report for the year 2024



By Dr David Muscat, Secretary

In 2024 the Dental Association of Malta committee was composed of 9 members namely:

- Dr Edward Fenech
President
- Dr Adam Bartolo
Vice President and Government affairs co-ordinator
- Dr Noel Manche
Treasurer
- Dr David Muscat
Secretary, Public Relations Officer and Editor of The Dental Probe Journal
- Dr Nicholas Busuttill Dougall
IT Officer
- Dr Thomas Grixti
Federation Representative
- Dr Ann Meli Attard
CPD Officer
- Dr Audrey Camilleri
European Liaison Officer
- Dr David Vella
Events co-ordinator

The committee held 12 committee meetings during the year in addition to the AGM.

The DAM organised several courses and lectures during the year:

- A Basic Life Saving Course was organised. This involved a full day of CPD lectures, an online course with an online MCQ exam and several hands on courses of several small groups. This course has to be repeated every two years.
- The DAM, in conjunction with Page Technology Ltd, organised a course at VOCCO HQ in Cuxhaven Germany. This was a course involving lectures and hands on teaching. Several of the dentists who attended the course wrote scientific articles based on what was learnt and these featured in Dental Probe Journal issues in 2024.
- The DAM organised a full day hands-on CPD course in Injection Moulding as well as a half day CPD course in Paediatric dentistry. Both these courses were done in conjunction with Cherubino Ltd.
- CPD lectures on Digital Dentistry and Periodontal disease were also organised.

All the events organised were full to capacity.

The DAM is planning further Basic Life Saving Courses as well as CBCT courses and a series of Periodontal lectures.

The European Liaison Officer attended two EU meetings abroad and briefed the DAM on European Dental Developments.

The Federation Representative attended several Federation meetings.

A Saint Apollonia Event was held in Rabat – a mass, a tour of a monastery followed by a lunch. 🇲🇹

International Relations Officer Report for 2024



By Audrey Camilleri
International Liaison Officer,
Dental Association of Malta

On 22nd November 2024, representatives of CED Member, Affiliate Member and Observer associations met in Brussels, Belgium for the CED General Meeting, under the chairmanship of President Dr Freddie Sloth-Lisbjerg and I attended representing Dental Association of Malta.

Delegates were updated on upcoming CED communication activities and the latest developments at the EU policy level

1. CORPORATE DENTISTRY

Corporate dentistry has been on the rise in Europe and is likely going to remain a reality among many European countries.

CED wishes to outline its stance on corporate dentistry and its core principles in relation to the current and future corporate dentistry status quo.

The BTF IM members discussed the topic of corporate dentistry and highlighted that this is an issue that will continue to exist and develop in the future.

BTF developed a document offering advice and supporting national dental associations in relation to raising awareness and educating students and young dentists about the possible ways of exercising the profession – as a liberal practitioner vs. as an employed dentist within corporate dentistry, in particular

highlighting the challenges and risks related to corporate dentistry.

The document was placed for adoption at the November General Meeting under the name CED Statement Corporate Dentistry and the Dental Profession.

2. EDUCATION AND PROFESSIONAL QUALIFICATIONS

The CED received on 28 May 2024 an invitation to provide input to a public consultation carried out by the European Commission DG GROW (Directorate General for Internal Market, Industry, Entrepreneurship and SMEs) for the preparation of the implementation report of the Professional Qualifications Directive 2005/36/EC (PQD).

The Chair explained that the implementation report was a formal obligation of the European Commission under Article 60(2) of the PQD Directive 2005/36/EC, to monitor and assess the effective implementation and challenges the Directive represents in all EU Member States. Such a report is published every five years.

The general objectives included identifying gaps, issues and challenges; monitoring progress; assessing effectiveness, assessing efficiency, preparing the ground for future policy initiatives; and outlining enforcement measures. The official implementation report was reported by the Chair to be foreseeably published in Q2 of 2025.

This consultation was explained to be an important opportunity to call for the amendment of the Professional Qualifications Directive to introduce strengthened clinical training requirements. The main incorporated CED positions and language included:

- A distinction made to clinical practice through a mandatory minimum of 5 out of 10 semesters (for at least 5 years of study) and 6 out of 12 semesters (for at least 6 years of study) dedicated to clinical training on patients.
- Addition of the distinction between student operators (individual execution of clinical procedures) and student observers/assistants when carrying out mandatory clinical practice in basic dental studies.
- The inclusion of an additional clinical supervised practice year (vocational training) following the completion of the dental degree and prior to registration as an independent dental professional.
- The call for the inclusion of periodontology under the Directive's requirements for dental specialties, harmonizing the minimum length of training between periodontologists and recognized specialist dental practitioners.

The Chair reminded members that this call for inclusion does not, in any way, push countries to recognise periodontology, but allows for the automatic recognition of

diplomas from countries currently recognising periodontology.

WG EPQ is waiting for follow-up from the consultation and the possibilities for providing further input.

External relations and collaboration with educational partners

The Chair also touched upon the renewed and strengthened collaboration with the Association for Dental Education in Europe (ADEE) and the European Students' Dental Association (EDSA).

3. DENTAL MATERIALS AND MEDICAL DEVICES

- **An updated CED Statement on Medical Devices Regulation and chairside CAD/CAM procedures:** rights of dentists not to be defined as manufacturers was done with the idea of providing direct reference to the definition of health institution (as outlined per the Medical Devices Regulation, Article 2(36) – 'Definitions'), and to also highlight the undeniable right of dentists to own and use CAD/CAM.

- **Dental technicians and CAD-CAM:** The WG held discussions on the latest reactions from dental technicians on the CAD-CAM issue and the dental profession. The conversation focused on the report provided by the Fédération Européenne des Patrons Prothésistes

Dentaires (FEPPD) to the CEN/TC 55 (European Committee for Standardisation/Technical Committee 55 – Dentistry).

In the report, the organisation representing the dental technicians once again questions definitions related to dental practices (e.g. 'health institution').

The WG DMMD discussed the reaction by FEPPD and the Chair encouraged the members to share any information and examples of similar national situations and experiences. The WG members also re-examined the MDCG (Medical Devices Coordination Group) document Guidance on the health institution exemption under Article 5(5) of Regulation (EU) 2017/745 and Regulation (EU) 2017/746, referred to in the FEPPD report. The WG was reminded that the guidance in question is also not legally binding.

The WG once again underlined and agreed that dental practices fall clearly under the health institution definition, according to the official legislation (MDR) and in relation to the MDCG Guidance document above; the WG DMMD also highlighted the fact that when using a CAD/CAM system a new medical device is not actually "manufactured" or produced, but a mass-produced

Continues on page 14

International Relations Officer Report for 2024

Continues from page 13

device (a Dental ceramic-resin block), which was already available on the market, is modified by the dentist so that it is adapted to the specific needs of individual patients).

• **Surveys in relation to medical devices:**

Survey on availability of medical devices, with healthcare professionals as a target group – after delays, the survey, managed by the Austrian Public Health Institute on behalf of the European Commission, was launched following the summer period, with deadline of 31 October.

The general objective of the study is to monitor and analyse the availability of medical devices on the EU market in the context of the implementation of medical devices and in vitro diagnostic medical devices Regulations from the perspectives of key stakeholders.

The WG DMMD has been actively involved on this topic and as such, continued engaging on the survey. The CED encouraged the CED members to provide direct replies to the survey as well (in light of the focus on national issues with availabilities of medical devices).

Survey on electronic instructions for use of medical devices:

the European Commission is currently considering allowing the use of Electronic Instructions for Use for all professional use devices (i.e. devices that are used by healthcare professionals). EU Regulation 2021/2226 allows Instructions for use (IFU) for certain medical devices such as

(active) implantable or software to be delivered in electronic format.

For devices covered by the EU Regulation 2021/2226 it is always possible to request a paper copy free of charge. Many manufacturers already provide IFU in electronic format in addition to paper. This is notwithstanding the possibility to request a paper copy free of charge.

The scope of the survey was to gather feedback on the potential extension of scope to all professional use devices.

The WG DMMD prepared a reply, highlighting that 1) Electronic instructions for use should be encouraged as much as possible, 2) The preference for instructions should be in the hands of the healthcare professional and based on the individual national circumstances, 3) In cases when the manufacturer goes out of business, access to instructions should be possible, in electronic and/or paper form (and again based also on the individual preferences of the healthcare professional and the relevant national situation).

• **Dental materials:**

The WG continues to monitor the status quo on various dental materials based on the latest updates and information. During its latest meeting, the WG discussed the topic of silicone monomers.

Silicone monomers Octamethylcyclotetrasiloxane (D4), Decamethylcyclopentasiloxane (D5) and Dodecamethylcyclohexasiloxane (D6) may be listed as persistent

organic pollutants under the UN Stockholm Convention on Persistent Organic Pollutants (POP).

This Convention aims to eliminate the manufacture and use of substances listed therein. Through the POP nomination, the European Commission aims to ban the direct use of D4, D5 and D6 in personal care products globally, which account for less than 2% of their total applications. However, such a nomination would endanger the production of silicone polymers, representing over 98% of the uses of these substances.

These substances are critical intermediates used predominantly in the production of silicone polymers, which have essential applications across many key sectors; use for dental impressions currently benefits from derogations under existing EU restrictions (which were last updated fairly recently, in May 2024).

As a first step in exploring this issue, the WG DMMD agreed for the CED to reach out to FIDE to check whether they are informed on this topic.

Cobalt: according to information from the dental technicians' reports, the REACH Committee has been re-evaluating Cobalt. As such, the Cobalt Chromium alloys status as reprotoxic, carcinogenic and mutagenic, may also be reconsidered.

4. ORAL HEALTH

Ageing and oral health:

The WG agreed to initiate the work around the drafting of a new CED document on ageing and

oral health. The topic of healthy ageing and specific health and oral health issues was justified as a topic of growing importance of international and European concern. This was linked to the growing importance given to continuing fighting antimicrobial resistance (AMR) from the European Union.

The Chair briefly presented the new Health at a Glance Report by the European Commission. This year's edition was dedicated to Promoting Healthy ageing and longevity whilst tackling health workforce challenges and included a set of important data on dental care and workforce.

Antimicrobial Resistance (AMR):

CED has resumed its participation in meetings of the AMR Stakeholder Network Group by the European Public Health Alliance (EPHA), after an interruption of meetings due to internal difficulties on EPHA's side. The Chair highlighted that the 2030 targets on AMR are yet far from being reached.

External meetings:

In addition to previously mentioned meetings, the CED was also present for the European Medicines Agency's (EMA) latest online HCPs POG meeting (meeting of eligible Healthcare Professionals Learned Societies Policy Officers' Group) on 17 September 2024.

Sugar:

The Chair updated the GM on the initiated work on updating the CED position on Sugar. He also explained the rationale behind the presentation of the FDI Policy Statement on the Reduction of Sugar Consumption, adopted at the FDI General Meeting in September, for endorsement by the CED General Meeting. The objective of the endorsement of

the FDI Paper was for the CED to recognise the important document produced and adopted at the FDI General Assembly in September, and to endorse the positions of the document ahead of the development of the CED own Resolution on sugar.

5. E-HEALTH

European Health Data Space – ongoing updates:

the expected final adoption of the EHDS in the European Parliament is in December, the act is to be published in the EU official journal in early 2025, and entry into force is expected in February 2025. Once the legislation is finalised and published, it would be useful for the CED national members to reach out and to ask their contacts from Ministries of Health as to what the national plans are for the implementation of the file and what other implications are envisioned for different stakeholders for their specific national level.

Artificial Intelligence (AI):

- Update on existing CED Resolution on AI in Dentistry: At its latest meeting, the WG members discussed and agreed that it would be useful to begin an update to the existing CED policy document on Artificial Intelligence (dating back to 2020), considering that this is a field that is constantly changing and developing.
- The Chair had an interview with the consultancy EY on Study on the economic impact of applying Artificial Intelligence in healthcare in Europe in end of October.

6. PATIENT SAFETY, INFECTION CONTROL AND WASTE MANAGEMENT

Vaccinations:

On behalf of the Chair, the CED members' national contributors

to the questionnaire Vaccination policies for dentists and other dental professionals in Europe (disseminated in 2023) were approached for final clarifications and validations to their replies.

In cooperation with the Directorate for Research, Studies, and Documentation, National Public Health Organization, Athens, Greece, a paper for publication under the title Vaccination Policies for dentists and other dental professionals in Europe has been prepared by Prof Tzoutzas, based among other, on the information on the vaccination questionnaire. The paper is envisioned to be published soon.

Dentistry and waste:

In the first part of the year, the WG re-opened the work on a CED Statement on waste management and sustainability in dentistry, which aims to ensure that the dental profession engages in the discussion on waste management in a proactive and timely manner and that it can respond to new legislative and regulatory initiatives that may impose restrictions and conditions to the dental office. The statement was successfully adopted during the May 2024 GM. The WG intends to continue gathering information and strengthening the CED-level discussion on this topic.

EU legislation on waste:

The WG continues following several relevant legislative pieces: the Proposal for a revised Urban Wastewater Directive; the Packaging Waste Regulation Proposal; the Proposal for a revision of the list of groundwater and surface water pollutants. The WG agreed to monitor the legislations as they progress, and to react if there are any concerns in relation to dentistry. 🏠

THE 3 SHAPE TRIOS SUPERCLASS

MASTERING THE DIGITAL WORKFLOW WITH TRIOS 5

By Bart Enterprises
 Presented By Professor Hani Tohme, 3 Shape Kol And ITI Fellow
 Thursday 5 December 2024 at Salina Resort
 Summarised By Dr David Muscat

The objectives of the course were to efficiently use the Trios 5 IOS and integrate the digital tools into clinical practice. Single crowns, smile design, implant planning, full arch rehabilitation and partial and removable dentures were all described.

There was a sizeable audience and the delivery was impressive.

Ergonomics of scanning as well as techniques to optimize scan accuracy and minimizing errors were described.

Scanning and integrating facial references with 2 D smile design and transferring the 2 D smile designs into 3 D models were shown.

A digital implant planning workflow for single and partial edentulism and an overview on prosthetically driven implant planning was shown.

The dual scan concept and the digital flow for full arch rehabilitation was shown with clinical cases.

SALIENT POINTS

With Trios one can scan a post and core. It depends on the depth of field. With 3 Shape one can also choose the tooth colour. The latest version allows for occlusal caries detection – but not interproximal caries as they have dispensed with the extra tip as studies have shown that the tip was not so efficient.

The next Trios version will focus on photogrammetry with extra oral photos incorporated to take implant positions.

The advantages of Trios 5:

1. Calibration free
2. Smart haptic feedback(vibration)
3. LED to increase readiness(will tell you if you are scanning)

4. Remote control button(no need to touch a mouse.
5. Scan several patients daily
6. The Trios 5 is 12% lighter and 21% smaller than the Trios 3.
7. A one time use sleeve can be used.
8. Scratch free sapphire glass
9. Sealed mirror(no dust or dirt)
10. Closed autoclavable tip
11. Pod for placement-charge and battery
12. The zoom feature - not really triangulating -software is zooming your image

THE BOPT CONCEPT

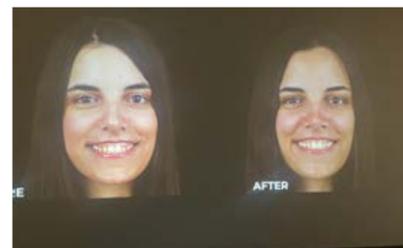
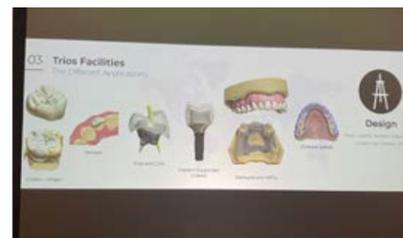
A knife edge margin on a natural tooth – no chamfer or shoulder. 0.5 mm sub gingival – put on a temporary crown and wait for 6 weeks. The soft tissue will heal. Remove temporary crown and take impression without cord.

CBCT

- You can do a quadrant, open mouth or closed mouth.
- Always do an OPEN MOUTH with opening a minimum of 4mm. CBCTs have to be calibrated at least 3 times a year otherwise you will lose accuracy. The quadrant is no longer recommended – take a full arch.
- Use cotton rolls so that you can visualise the soft tissues.
- CBCT requires open mouth, no quadrant, and retraction is mandatory.

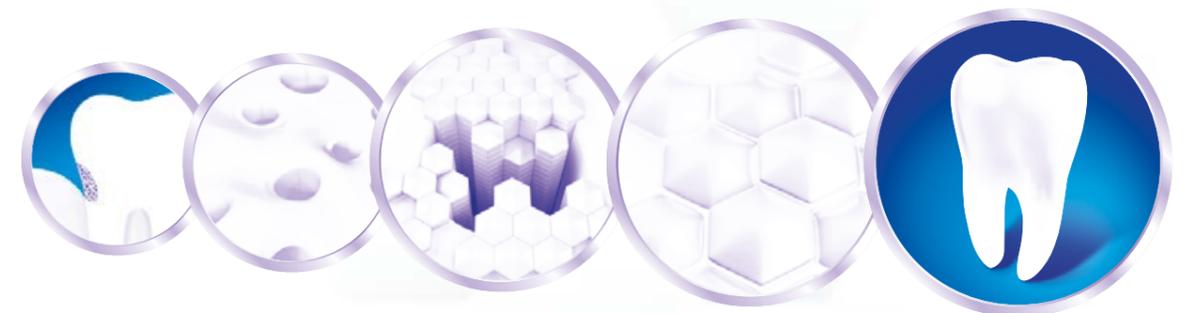
Always scan the full arch even for a single implant.

With dentures use fiduciary markers on the buccal flanges and the palate. For a denture CBCT this has to be on a foam. For the bite take two bite indices and do the CBCT with the bite indices. Use a dual scan protocol. Use flowable composite for markers.



DEEP REPAIR

& DAILY PROTECTION FOR SENSITIVE TEETH



CAUSE OF SENSITIVITY PAIN

TINY HOLES IN DENTINE

GETS DEEP INSIDE THESE HOLES

REPAIRS SENSITIVE TEETH

HELPS PROTECT FROM PAIN



LIFE'S TOO SHORT FOR SENSITIVE TEETH

OSTEORADIONECCROSIS OF THE JAW: A CASE REPORT

A case report by Leonard Schembri
Supervisor: Dr Adam Bartolo
University of Malta, Department of Dental Surgery

ABSTRACT

Osteoradionecrosis of the jaw is a serious complication following radiotherapy defined by the failure of bone to heal within a period of three months. The following case report presents the case of a 71-year-old male who presented to the clinic with an osteoradionecrotic lesion in the body of the mandible. Upon presentation this was the third time the patient experienced an osteoradionecrotic lesion in the area for which he was treated using hyperbaric oxygen therapy prior to sequestration of the lesion and extraction of an adjacent mobile tooth.

The report also highlights and discusses the risk factors, pathogenesis and treatment modalities of osteoradionecrosis of the jaw including hyperbaric oxygen therapy, surgical interventions and medical treatment with pentoxifylline.

KEYWORDS

- Osteoradionecrosis
- Mandible
- Hyperbaric Oxygen Therapy

INTRODUCTION

Osteoradionecrosis of the jaw is a serious complication that may occur following radiation therapy in the head and neck. It is defined as exposed, irradiated bone that fails to heal within three months in the absence of evidence of a persistent or recurrent tumour. (Chronopoulos, Zarra, Ehrenfeld, & Otto, 2018).

Radiological evidence of necrosis is also required for the classification and diagnosis of osteoradionecrosis severity.

The definition, as well as the mechanism of pathogenesis, risk factors, clinical staging and treatment protocols still require further investigation.

Potential risk factors for the condition can be divided into patient-related (such as a history of alcohol use, smoking, sex and diabetes mellitus), tumour-related (site of the primary tumour, T-stage and nodal status) and treatment related factors (such as radiotherapy technique, concurrent chemotherapy and pre-radiotherapy surgery). (Kubota, et al., 2021).

This report presents the case of a 71-year-old male who presented with a recurring episode of osteoradionecrosis that occurred 3 years following radiotherapy in the left side of the mandible.

The purpose of the case report is to review the characteristics of osteoradionecrosis, risk factors and options for management.

CASE PRESENTATION

A 71-year-old male reported to the dental teaching clinic noting pain in the left side of the jaw. The patient noted that this was recurring and had first appeared approximately 3 years ago following treatment of a squamous cell carcinoma which was treated with chemotherapy, surgery and radiotherapy.

Following the radiotherapy the patient developed osteoradionecrosis in the left side of the jaw. He then underwent treatment with hyperbaric oxygen which significantly aided the condition.

The patient is diabetic and smokes 2 packets of cigarettes a day, both of which are factors that increase the risk for osteoradionecrosis. In addition, the patient also suffers from hypertension.

Intraorally the examination showed exposed necrotic bone in the left side of the mandible in the molar area which was 15mm in length (Figure 1). The patient also had grade 1 mobility in tooth 35. Saliva production was also reduced, and the patient's oral hygiene was found to be poor.

A panoramic radiograph was also taken which revealed radiolucency indicating necrosis of the bone which mesiodistally extended from the distal of tooth 35 to the mesial border of the ramus. The radiolucency also extended from the alveolar ridge to below the mandibular canal (Figure 2).

Given the clinical and radiographic findings the stage of the disease

can be defined as stage II division A according to the classification by Schwartz and Kagan since both the cortical and underlying medullary bone are necrotic and there is minimal soft tissue ulceration.

The full thickness of the mandible is not involved and thus it is not classified as a stage III. (Schwartz & Kagan, 2002)

Differential diagnoses for the condition include a possible recurrence of the tumour, osteomyelitis or bisphosphonate related osteoradionecrosis of the jaw. (Chronopoulos, Zarra, Ehrenfeld, & Otto, 2018)

The treatment plan for the patient was to provide hyperbaric oxygen therapy prior to the extraction of the mobile tooth and possibly sequestration.

The surgical intervention would be reassessed following hyperbaric oxygen due to the association of the lesion to the mandible as well as the depth of the lesion which would increase the risk of fracture due to the amount of bone that would have to be removed.

The patient however opted to cease hyperbaric oxygen treatment after having attended for the first two sessions of twenty. On the follow up visit however the condition was noted to have improved with part of the necrotic bone having detached.

DISCUSSION

Osteoradionecrosis in the mandible is a debilitating complication of radiotherapy which has become a key part of management for many head and neck malignancies.

As it develops it causes trismus, neuropathic pain and chronic

drainage along with other side effects of radiation therapy such as xerostomia, dysphagia and decreased tongue mobility. (Adam S. Jacobson, 2010).

As mentioned previously it is commonly defined as exposed irradiated bone that fails to heal over a period of 3 months without any evidence of persistent or recurrent tumour although there is no standard and unified definition.

Various risk factors predispose patients to the development of osteoradionecrosis a few of which were present in this case, namely the age of the patient being over 55, smoking and suffering from diabetes.

Other host factors seen in the patient include existing dental disease and poor oral hygiene. The occurrence is however largely dependent on the type and amount of radiation.

Microorganisms are not thought to play a major role however as mentioned poor oral hygiene is a predisposing factor and it is hypothesised to be the reason for there being a higher incidence of osteoradionecrosis in dentate patients as opposed to edentulous patients due to the higher bacterial load. (Holley, Keenan, & Militsakh, 2018) (Kubota, et al., 2021)

The disease occurs more commonly in the mandible when compared to other sites in the head and neck. The reasons for this are thought to be that the mandible is at an increased risk of ischemic radionecrosis due to it consisting of cortical bone with a singular blood supply in the inferior alveolar artery.

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OSTEORADIONECCROSIS OF THE JAW: A CASE REPORT

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The mandible is also the most common site exposed to higher radiation doses during radiotherapy. Osteoradionecrosis can occur within a few months to up to 30 years following radiotherapy and mainly arises following trauma such as extractions or even denture trauma.

However any manipulation of irradiated bone may result in the development of osteoradionecrosis thus emphasising the importance of carrying out necessary examination and treatment prior to initiation of radiotherapy.

Surgery as modality of treatment for the condition may also not be indicated for this reason especially if opting for a more conservative approach to treatment. (Holley, Keenan, & Militsakh, 2018)

There are multiple theories as to the pathogenesis of osteoradionecrosis the first of these was that osteoradionecrosis was caused by local injury and infection following radiation exposure.

The exposed tissues in the mandible develop inflammation, hyperaemia and obliterative endarteritis which is the inflammation of the inner lining of an artery which causes occlusion of said artery. (Dekker H, et al., 2018)

This led Marx to put forward the triple H hypothesis which proposed that exposure to radiation caused hypoxia, hypocellularity and hypovascularity meaning that osteoradionecrosis occurs as a result of microvascular damage causing a damaged supply. This theory formed the basis of the use of hyperbaric oxygen as a treatment for osteoradionecrosis. (Marx, 1983)

A more recent theory that has been put forward is the radiation induced fibro atrophic theory. This suggests that radiation causes injury to endothelial cells both directly and through the generation of free radical oxygen species (ROS). This then leads to micro vessel necrosis causing vascular thrombosis and subsequent tissue loss.

The injured endothelial cells also cause ROS mediated cytokine production which in turn together with ROS and radiation cause unregulated fibroblast activation of which the myofibroblasts persist and cause dysregulation of collagen metabolism and fibrosis.

Fibrosis causes obliteration of the inferior alveolar artery meaning a lack of blood supply to the mandible. The combination of the death osteoblasts after irradiation, their failure to repopulate along with the excessive proliferation of myofibroblasts causes a reduction in the bony matrix and replacement with fibrous tissues.

Myofibroblasts will eventually undergo apoptosis causing the bone to remain poorly vascularised and fibrosed. Physical trauma to these now fragile tissues that have been exposed to radiation causes a late surge of reactivated inflammation meaning they have a tendency to develop osteoradionecrosis. (Lyons & Ghazali, 2008)

In terms of treatment there is no universally accepted treatment protocol for osteoradionecrosis. There are however several staging systems that have been introduced to aid in the management of osteoradionecrosis such as those developed by Marx in 1983 and more recently by Schwartz and Kagan in 2002.

For cases with a lower stage the disease may be treated conservatively meaning the institution of good oral hygiene, analgesics and antibiotics. This has been found to be effective in 25-44% of cases. Ultrasound therapy is another treatment modality that has been used for the treatment of osteoradionecrosis.

First proposed by Harris in 1992 it induces angiogenesis and improves blood flow to muscles and ischemic varicose ulcers. The use of ultrasound for 15 minutes per day for 40-100 applications along with debridement showed healing in 48% of cases. (Rice, Pozlyzois, Ekanayake, Omer, & Stassen, 2015)

Hyperbaric oxygen therapy (HBO) is another treatment modality for osteoradionecrosis that is primarily used as an adjunct to surgery and is found to have little effect when being the sole treatment for the disease.

It is based on Marx's theory that osteoradionecrosis is caused by hypoxia, hypocellularity, and hypovascularity. The mode of action of HBO is to increase the blood-tissue oxygen gradient therefore increasing the diffusion of oxygen into the hypoxic tissues. In turn this causes stimulation of the proliferation of fibroblasts, angiogenesis and collagen formation. It is also seen to have bacteriostatic and bactericidal effects.

The steep oxygen gradient generated by HBO promotes neovascularisation as well as improvement in the function of white blood cells and fibroblasts which in turn promote wound healing. (Akbar, 2023).

The outcomes of treatment are mostly dependant on the stage of the disease with patients in stage I having the best results with some healing through the use of HBO

alone. Patients who had reached stage II also see improved results with HBO however it may be less useful for patients in stage III where it seems to be less effective for bone healing at this stage.

However HBO may improve the surrounding tissues thus having a positive effect on post-surgical wound healing. (Dieleman FJ, 2017)

Protocols for the use of HBO therapy for osteoradionecrosis vary but most predominantly the most used is 20-30 sessions of 90 minutes at 2.5 ATA preoperatively and 10 sessions postop. (Dhanda, Beshara, Machon, & Parmar, 2009) HBO therapy can be administered using a multiplace or monoplace chamber.

The former pressurises the chamber with air and the patient then breathes oxygen through a mask whilst the latter fills the chamber with pressurised air and the patient then breathes the oxygen directly. (Niinikoski, 2004).

As mentioned, the patient in this case report had previous success with HBO however even in the absence of extractions or other dental treatment he experienced a second episode of osteoradionecrosis due to the latent effect of the condition.

The patient opted to not continue HBO treatment due to the length and number of sessions that the treatment entails. This is a significant disadvantage of the treatment.

There are also questions for more advanced cases of osteoradionecrosis and the use of HBO as these cases require radical surgical intervention and the use of HBO will only delay treatment without benefit. (Rice, Pozlyzois, Ekanayake, Omer, & Stassen, 2015) (Adam S. Jacobson, 2010)



Figure 1 Intraoral photograph showing exposed necrotic bone in the lower left molar area



Figure 2 Orthopantomogram showing the extent of necrotic bone

Studies are also divided on the use of HBO as a prophylactic measure prior to performing dentoalveolar procedures with some studies indicating a lower proportion of individuals being diagnosed with osteoradionecrosis following prophylactic HBO therapy whilst others showed no significant evidence to support the use of HBO therapy. (Dnag B, et al., 2023) (Sultan, Hanna, & Danielle N. Margalit, 2017)

Medical treatment is also being used for the prevention and treatment through the use of pentoxifylline.

It is given either in a dose of 400 mg twice daily for 1 week prior to extraction along with tocopherol 1000

IU once daily and continuing the pentoxifylline for another 8 weeks or when using prior to the removal of sequestrum the same dose regimen is used but for three months prior to the date of the sequestrectomy along with clodronate 1600mg daily.

Pentoxifylline has been shown to dilate blood vessels and inhibit inflammatory reactions in vivo.

It also inhibits proliferation of human dermal fibroblasts and the production of extracellular matrix as well as increasing collagenase activity in vitro. (Lyons & Ghazali, 2008).

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CONCLUSION

Osteoradionecrosis is a debilitating disease which comes following an already difficult time for patients due to malignancy and its associated treatment. This case report highlights the associated risk factors for osteoradionecrosis in particular those observed in the patient as well as the pathogenesis and treatments available for osteoradionecrosis.

Emphasising the importance of preventive measures together with early treatment of osteoradionecrosis as well as the regular follow ups following radiotherapy is key in such cases along with prophylaxis prior to dentoalveolar procedures. Future research should be focused on better standardising staging for the condition as well as establishing the optimum protocols for treatment.

PATIENT CONSENT

Informed patient consent was obtained prior to examination and photographs. Diagnosis and treatment plan were also explained to the patient.

The patient was informed about the right to the confidentiality of his medical information and the potential use of anonymised data for educational or research purposes as well as his right to withdraw consent of the data's usage at any point. A signed consent form is kept with the patient's medical history.

ACKNOWLEDGEMENTS

I would like to acknowledge and give my thanks to my supervisor Dr Adam Bartolo for his guidance in completing this case report. 🙏

REFERENCES

- Adam S. Jacobson, D. B. (2010). Paradigm shifts in the management of osteoradionecrosis of the mandible. *Oral Oncology*, 795-801.
- Akbar, N. T. (2023). The role of hyperbaric oxygen therapy on the management of mandibular osteoradionecrosis: A scoping review. *Journal of International Dental and Medical Research*, 1836-1845.
- Chronopoulos, A., Zarra, T., Ehrenfeld, M., & Otto, S. (2018). Osteoradionecrosis of the jaws: definition, epidemiology. *International Dental Journal*, 22-30.
- Dekker H, Bravenboer N, Van Dijk, D., Bloemena, E., Rietveld, D., Bruggenkate, & Schulten. (2018). The irradiated human mandible: A quantitative study on bone vascularity. *Oral Oncology*, 126-130.
- Dhanda, J., Beshara, D., Machon, & Parmar, s. (2009). Comparisons between UK and European protocols used in the treatment of osteoradionecrosis with hyperbaric oxygen therapy. *British Journal of Oral and Maxillofacial Surgery*, 25-26.
- Dieleman FJ, P. T. (2017). The efficacy of hyperbaric oxygen therapy related to the clinical stage of osteoradionecrosis of the mandible. *International Journal of Oral and Maxillofacial Surgery*, 428-433.
- Dnag B, Gamage S, Sethi S, Jensen ED, Sambrook P, & Goss A. (2023). The role of hyperbaric oxygen in osteoradionecrosis-a prophylactic insight. *Australian Dental Journal*, 171-178.
- Holley, T., Keenan, D., & Militakh, O. (2018, July). Osteoradionecrosis. *Current Otorhinolaryngology Reports*, 6, 285-291.
- Kubota, H., Miyawaki, D., Mukumoto, N., Ishihara, T., Matsumura, M., Hasgeawa, T., ... Sasaki, R. (2021). Risk factors for osteoradionecrosis of the jaw in patients with head and neck squamous cell carcinoma. *Radiation Oncology*. doi:https://doi.org/10.1186/s13014-020-01701-5
- Lyons, A., & Ghazali, N. (2008). Osteoradionecrosis of the jaws: current understanding. *British Journal of Oral and Maxillofacial Surgery*, 653-660.
- Marx, R. (1983, May). Osteoradionecrosis: a new concept of its pathophysiology. *Journal of Oral and Maxillofacial Surgery*, 283-288.
- Niinikoski, J. H. (2004). Clinical Hyperbaric Oxygen Therapy, Wound Perfusion, and Transcutaneous. *World Journal of Surgery*, 307-311.
- Rice, N., Pozlyzois, I., Ekanayake, K., Omer, O., & Stassen, L. (2015). The management of osteoradionecrosis of the jaws. *The Surgeon*, 101-109.
- Schwartz, H. C., & Kagan, R. (2002, April). Osteoradionecrosis of the Mandible. *American Journal of Clinical Oncology*, 25(2), 168-171. doi:0.1097/00000421-200204000-00013
- Sultan, A., Hanna, G. J., & Danielle N. Margalit, N. C. (2017). The Use of Hyperbaric Oxygen for the Prevention and Management of Osteoradionecrosis of the Jaw: A Dana-Farber/Brigham and Women's Cancer Center Multidisciplinary Guideline. *The Oncologist*, 343-350.

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A RARE FIND: REPORTING A CASE OF ORAL LEIOMYOMA

A case report by Sarah Bellizzi, Master of Dental Surgery – Year V

Supervisor: Mr Nicolas Bezzina BChD, MFDSRCS(Eng), PGDip(Conscious Sedation), MOSRCSEd(Oral Surgery)

University of Malta, Faculty of Dental Surgery

ABSTRACT

As originally outlined by Virchow in 1854, a leiomyoma is a benign smooth muscle neoplasm. While this is infrequently encountered in the upper aerodigestive tract, it is more commonly found in the genitalia, skin, and gastrointestinal tract. This condition can present at any age; however, it is frequently encountered during the fourth and fifth decades of life, with no preference for a particular gender. (Douglas Gnepp, Jus/n Bishop - Gnepp's Diagnos/c Surgical Pathology of the Head and Neck-Elsevier (2020), n.d.)

Here, diagnosis primarily relies on histological investigations using specific stains through immunohistochemistry which verify smooth muscle origin. 'Conservative' surgical removal stands to be the most effective treatment approach in such cases, with likelihood of recurrence being highly unlikely. (Kaur & Gondal, 2011)

In this case report, a 46-year-old male presented to the Oral Surgery Department at Mater Dei Hospital following referral by his General Dental Practitioner. The patient presented with a mobile, palpable lump that was diagnosed as an Oral Leiomyoma. The lump was located at the base of the sulcus in the lower right buccal segment region and had been present for more than three months.

KEYWORDS

- Leiomyoma
- Oral cavity
- Smooth muscle
- Benign

INTRODUCTION

Oral Leiomyoma is a term which refers to a benign smooth muscle neoplasm which occurs in the upper aerodigestive tract.

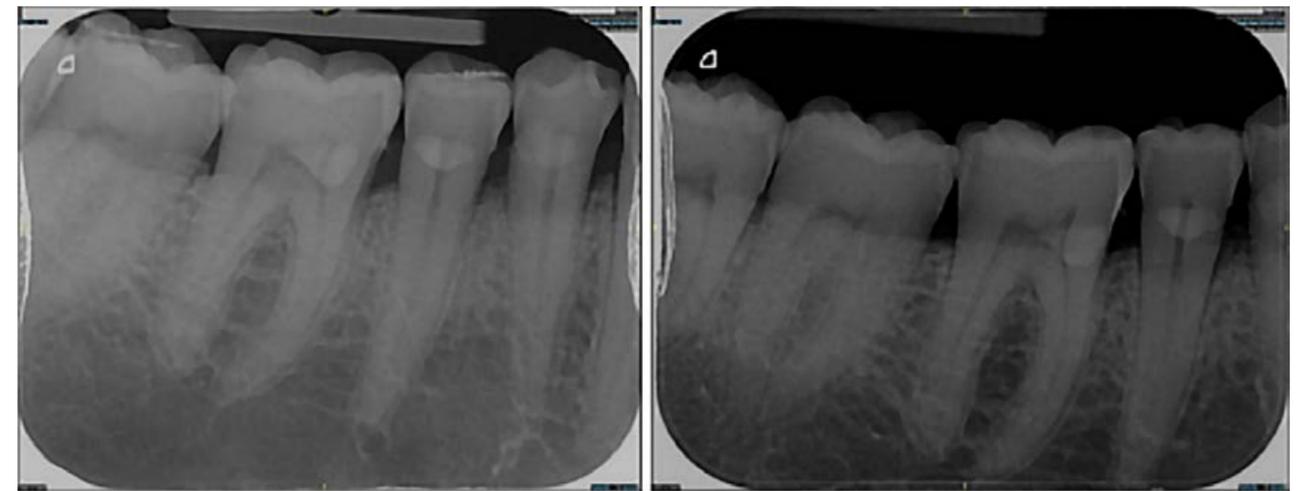
Because of its rarity, there is a scarcity of literature on this condition. The objective of this case report is to illustrate a case of a 46-year-old male who has been diagnosed with Oral Leiomyoma.

Additionally, this will also highlight clinical and pathological characteristics of the condition, offering insights into differential diagnosis and providing recommendations for the most suitable treatment options in such cases.

CASE PRESENTATION

A 46-year-old male presented to the Oral Surgery Department at Mater Dei Hospital following referral by his General Dental Practitioner (GDP). The patient did not report any specific complaints himself. The patient stated that his GDP referred him due to a mobile, palpable lump in the lower right area. He added that this had been present for over three months.

Additionally, he mentioned that the lump remained unchanged and did not cause any discomfort till that point in time. Patient was a non-smoker and a social alcohol user. No known drug allergies reported.



Figures 1 & 2 – Peri-apicals showing absence of any abnormalities in the lower right sextant

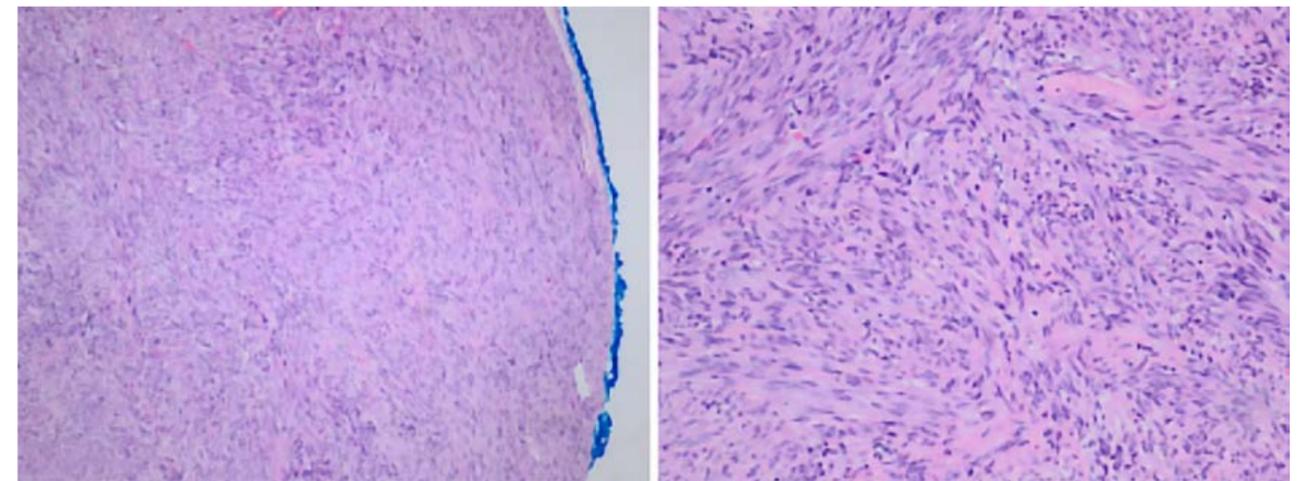


Figure 3 – Proliferation of bland spindle cells arranged as intersecting fascicles. The tumour reaches the inked surgical margin (x40 magnification)

Figure 4 – High power view showing the plumped spindle cells without cytological atypia & inconspicuous mitotic activity (x100 magnification)

Medical History:

Recurrent Depressive

Disorder Drug History:

- Lithium Carbonate (Camcolit®) Tablets Modified Release 400mg
- Quetiapine Tablets 200mg
- Venlafaxine Capsules Modified Release 75mg

The clinical examination was initiated by an extra-oral examination, during which no abnormalities were identified upon examination and palpation.

Subsequently, during the intra-oral examination, a 15mm firm, rubbery mass was observed in the lower right buccal sulcus area around tooth 46. However, it was determined that this lump showed no connection to any of the teeth in the sextant.

Furthermore, all the teeth in the lower right sextant showed a positive response to cold testing, thus, confirming that there was no link between the lump and any of these teeth.

Several peri-apical radiographs were taken for this sextant, and showed no abnormal findings, thus providing confirmation of the absence of any such connection. (Figures 1&2)

Following the patient's oral presentation, the following differential diagnoses were considered:

1. Fibroma
2. Neurofibroma
3. Lipoma
4. Leiomyoma
5. Leiomyosarcoma (Nguyen & Frydrych, 2017)

An Excisional biopsy was carried out with the patient under local anaesthesia, using two cartridges of 2% lidocaine containing 1:80,000 epinephrine. An incision was made in the epithelial tissue over the lump and the lump was enucleated out as a single mass. This is a crucial step in such cases to avoid any future potential recurrences.

A 3.0 resorbable suture (Vicryl) was employed aiming for primary closure of the wound. Following the surgical procedure, post-operative instructions were provided to the patient. Subsequently, the specimen was then sent to the histopathology laboratory for further analysis to acquire a conclusive histological diagnosis.

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This was transported in 10% formaldehyde solution to be picked up in immunohistochemistry. The patient's recovery after the surgical procedure proceeded without any complications. The patient was reviewed four weeks after the procedure took place where no clinical abnormalities were detected.

The patient was informed on the histological diagnosis, which confirmed the presence of an Oral Leiomyoma extending to the surgical margins without any signs of malignancy. (Figures 3&4) This was supported through immunohistochemistry which reported a 'strong diffuse expression' of α -smooth muscle actin and no expression of the S100 protein.

An appointment for a three-month post-surgical review was scheduled for the patient to evaluate and re-assess the clinical situation due to the possibility of recurrence.

DISCUSSION

As originally outlined by Virchow in 1854, a leiomyoma is a benign smooth muscle neoplasm. While this is infrequently encountered in the upper aerodigestive tract, it is more commonly found in the genitalia, skin, and gastrointestinal tract. (Douglas Gnepp, Jus/n Bishop - Gnepp's Diagnos/c Surgical Pathology of the Head and Neck-Elsevier (2020), n.d.)

This condition can present at any age; however, it is frequently encountered during the fourth and fifth decades of life, with no preference for a particular gender.

Nevertheless, both Kaur et al. and G. Sfasciotti et al. suggest that this

demonstrates a preference towards the female gender. (Douglas Gnepp, Jus/n Bishop - Gnepp's Diagnos/c Surgical Pathology of the Head and Neck-Elsevier (2020), n.d.; Gianluca et al., 2011; Kaur & Gondal, 2011)

It has been suggested by Pratibha et al., that the recent discovery of sex steroid receptors in the leiomyomas suggest the possibility that the development of these tumours might be influenced by hormones and as a result showing a more female predilection. (Sharma et al., 2018)

Leiomyomas in the head and neck region are believed to arise mainly from vascular smooth muscle where most of the cases can be classified into two primary types:

- (1) Solid leiomyoma – a leiomyoma of the deep soft tissue and
- (2) Vascular leiomyoma, alternatively termed angioleiomyoma or angioleiomyoma. The vascular type accounts for 75% of all oral cases seen.

A less common variety, termed epithelioid leiomyoma has been documented. (Douglas Gnepp, Jus/n Bishop - Gnepp's Diagnos/c Surgical Pathology of the Head and Neck-Elsevier (2020), n.d.)

Pratibha et al., provides additional details by explaining that the infrequency of these smooth

muscle tumours in the oral cavity is linked to the scarcity of smooth muscle tissue in the oral cavity. In 1938, Scout put forward the idea that the smooth muscle found in the oral cavity originates from the tunica media of the blood vessel walls.

Other sources have suggested origins for smooth muscle tumours in the

oral cavity encompass the ductus lingualis, circumvallate papillae of the tongue, and the smooth muscles of the excretory duct of the salivary glands with heterotopic embryonal muscle tissue. (Sharma et al., 2018)

Most of the reported cases describe an asymptomatic, gradually enlarging mass which feels firm to touch. The colour of the lesion varies depending on the depth and vascularity of such lesion. (Kaur & Gondal, 2011).

Some lesions are occasionally tender or painful, particularly in the case of vascular leiomyoma variants. (Douglas Gnepp, Jus/n Bishop - Gnepp's Diagnos/c Surgical Pathology of the Head and Neck-Elsevier (2020), n.d.)

Such lesions progressively grow over the course of several years and may attain a diameter of several centimetres, although most lesions are relatively small during biopsy.

The solid leiomyoma typically presents as a slow-growing, small, asymptomatic, submucosal nodule with a colour like that of the adjacent mucosa, occasionally displaying a greyish tone. The lesion's surface is typically smooth and rarely ulcerates.

On the other hand, the vascular variant is characterized by a blue or red discolouration, ranging in size from a few millimetres to 3cm, often tender to palpation.

Generally, this is well-defined with the ability to move freely within the tissues of the oral mucosa. (Douglas Gnepp, Jus/n Bishop - Gnepp's Diagnos/c Surgical Pathology of the Head and Neck-Elsevier (2020), n.d.; Sharma et al., 2018)

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A RARE FIND: REPORTING A CASE OF ORAL LEIOMYOMA

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Distinguishing leiomyomas from other mesenchymal tumours poses a challenge due to their comparable clinical features. Therefore, the definitive diagnosis of oral leiomyoma relies predominantly on histopathological examination using haematoxylin and eosin (H&E) stains.

In certain instances, haematoxylin and eosin stains fail to provide conclusive information and thus immunohistochemical tests would have to be conducted to attain a more definitive diagnosis. (Sharma et al., 2018)

This becomes particularly crucial in distinguishing between a leiomyoma and a leiomyosarcoma, with the latter representing the malignant counterpart.

Histologically, all types of leiomyomas are tightly encapsulated and exhibit minimal cellular pleomorphism or mitotic activity.

These are typically composed of numerous intertwining bundles of 'lightly eosinophilic spindled cells' featuring elongated nuclei with a distinct 'cigar shaped' appearance.

Smooth muscle leiomyomas also known as pilar leiomyomas do not show hyperchromatism and mitotic activity.

Furthermore, when compared to those which arise from vascular smooth muscle these have a diminished eosinophilic quantity and give a less conspicuous indication of their smooth muscle origin.

Alternatively, vascular type leiomyomas are notably well defined, composed of clearly

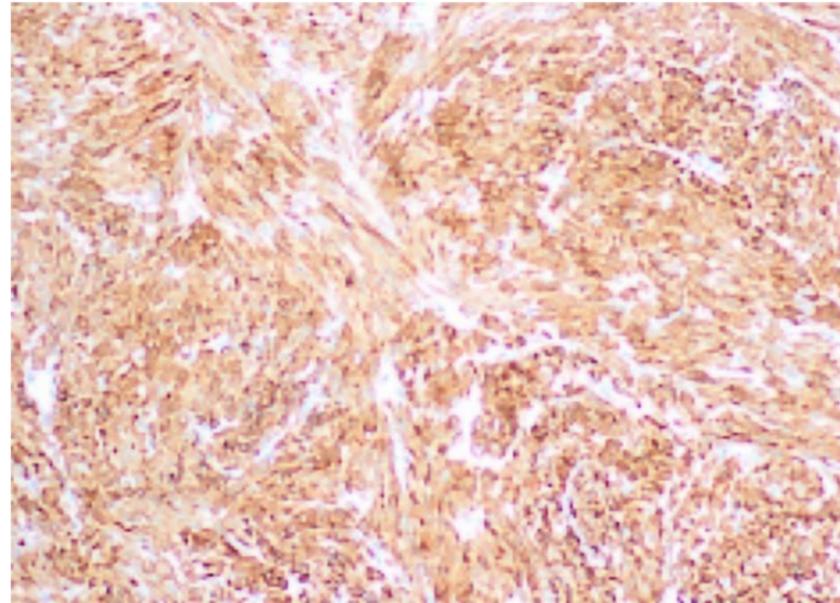


Figure 5 - Strong and diffuse expression of Smooth Muscle Actin by the tumour. Desmin is similarly expressed. (Photomicrograph x100 magnification)

eosinophilic spindled cells featuring the 'cigar shaped' nuclei and perinuclear vacuoles. These cells are organised in fascicles which intersect each other at right angles.

In these lesions however there is absence of both mitotic activity and nuclear atypia. (Douglas Gnepp, Jus/n Bishop - Gnepp's *Diagnos/c Surgical Pathology of the Head and Neck-Elsevier* (2020), n.d.)

In this case report, immunohistochemical studies performed have shown a strong and diffuse expression of SMA by the tumour with Desmin being similarly expressed (figure 5).

Smooth Muscle Actin (SMA) is a 'rabbit monoclonal antibody' designed for laboratory applications, specifically for qualitative identification of smooth muscle actin protein through immunohistochemistry in 'formalin-

fixed paraffin-embedded' tissues. Antibodies will then specifically identify the alpha-smooth muscle actin. (Smooth Muscle Ac/n (SMA) Concentrated and Prediluted Rabbit Monoclonal An/body, n.d.)

In Immunohistochemistry, both pilar and the more non-superficial located leiomyomas exhibit robust expression of smooth muscle actin and varying expression of desmin.

Some leiomyomas sporadically lack expression of such desmin; thus, smooth muscle actin serves as a more reliable screening marker for such leiomyomas irrelevant of the location of such leiomyoma.

In addition to this, there may be occasional positivity for low-molecular weight keratins which was not evident in our case. (Douglas Gnepp, Jus/n Bishop - Gnepp's *Diagnos/c Surgical Pathology of the Head and Neck-Elsevier* (2020), n.d.)

The primary treatment option for benign smooth muscle tumours seems to be surgical excision. In all the leiomyomas documented in the literature to date, surgery has been employed as the sole treatment.

There are no reported instances of recurrence after complete excision, underscoring the importance of meticulous and total removal of such lesions.

In our case, a total resection of the mass was made under local anaesthesia. Following this, the patient did not experience any complications or pathological recurrences to date.

CONCLUSION AND LEARNING POINTS

A Leiomyoma is a benign smooth muscle neoplasm which is hardly encountered in the oral cavity. Clinical and histopathological characteristics here closely resembles that of other mesenchymal tumours.

Consequently, achieving an accurate diagnosis poses a challenge and relies predominantly on histopathological examination. Treatment in such cases involves surgical excision taking into consideration the rarity of recurrence.

In conclusion, reporting such cases in the literature is crucial to enhance the understanding of such condition.

PATIENT CONSENT

In Order to conduct this case report, the patient was presented with relevant information, allowing them to make an informed decision, resulting in acquisition of a written consent, thus ensuring the validity of the consent process.

ACKNOWLEDGEMENTS

I would like to express my deepest and sincere gratitude to my case report supervisor Mr. Nicolas Bezzina, Department of Oral Medicine, Surgery, Pathology and Radiology for his guidance and advice which made completion of this research study possible. It was a great honour and privilege to work under his guidance.

I want to convey my appreciation to Dr. Sandra Betts and Dr. Rebecca Schembri Higgans for their generosity in sharing histological images, which significantly enhanced the quality of this study.

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REFERENCES

- Douglas Gnepp, Jus/n Bishop - Gnepp's *Diagnos/c Surgical Pathology of the Head and Neck Elsevier* (2020). (n.d.).
- Gianluca, S., Roberta Marini, D., Federica Tonoli, D., Maria Paola Cristalli, D., Luca SfasioQ Via Famiano Nardini, G., & Ph, I. (2011). Case report. In *Annali di Stomatologia*. Kaur, G., & Gondal, R. (2011). Oral leiomyoma. *Journal of Oral and Maxillofacial Pathology*, 15(3), 361-362. [hWps://doi.org/10.4103/0973-029X.86727](https://doi.org/10.4103/0973-029X.86727)
- Nguyen, A. P., & Frydrych, A. M. (2017). Oral Leiomyoma in an Adult Male: A Case Report.

- The Open Den/stry Journal, 11(1), 520-526. [hWps://doi.org/10.2174/1874210601711010520](https://doi.org/10.2174/1874210601711010520)
- Sharma, P. S., Naikmasur, V. G., Nandimath, K. R., Burde, K. N., Hegde, V., & Anehosur, V. (2018). Oral leiomyoma: A case report on a rare lesion in oral cavity. *Journal of Oral Diagnosis*. [hWps://doi.org/10.5935/2525-5711.20180001](https://doi.org/10.5935/2525-5711.20180001)
- Smooth Muscle Ac/n (SMA) Concentrated and Prediluted Rabbit Monoclonal An/body. (n.d.). [hWp://biocare.net/](https://biocare.net/)

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PLASMA CELL GINGIVITIS – A CHALLENGE FOR THE DENTAL PROFESSION

A case report by Dr Katrina Marie DeBattista (M.D.S),
General Professional Trainee Mater Dei Hospital

ABSTRACT

Plasma cell gingivitis (PCG) is a rare, benign condition of the gingiva. It has a rapid onset and affects both attached and unattached gingivae. Its main characteristic is sharply demarcated erythematous and oedematous gingivitis which often extends to the mucogingival junction. This report will discuss a case of PCG as a reaction following periodontal treatment and its management.

INTRODUCTION

A 34-year-old male presented at Mater Dei Hospital Dental Department Emergency complaining of generalised white patches and lesions which become covered in blood on brushing. The patient claimed to have no pain or symptoms unless brushing.

CLINICAL CASE

On examination the patient had erythematous swollen gingivae especially on the lower anterior quadrant (figure 1) together with sloughing and ill-defined whitish lesions with a clear background on the attached and unattached buccal mucosa associated with LL5 and 6 (figure 1), UR5 (figure 2), UL3-6 (figure 3).

History of presenting complaint

The patient had undergone periodontal treatment ten days before presentation and was prescribed a chlorhexidine mouthwash and toothpaste. During these ten days the patient was also prescribed an antihistamine however the lesions kept on increasing in size and frequency.

Investigations

Routine blood tests were ordered to exclude potential undiagnosed underlying conditions while a biopsy for histology and direct immunofluorescence

(IMF) was scheduled.

Differential diagnosis

1. Erythema multiforme
2. Vesiculo-bullous disorder – pemphigus, pemphigoid

Biopsy

- Punch biopsy 4mm lower attached mucosa LL3 region
- Punch biopsy 4mm upper left ulcer UL4 region
- Incisional biopsy of perilesional mucosa sent in saline for direct IMF

Following the biopsy, the patient was prescribed prednisolone and analgesics as required. A week later the patient was doing well as symptoms had subsided and lesions were decreasing in size as well.

Biopsy result

The diagnosis for this patient came back as plasma cell gingivitis therefore vesiculobullous lesions were excluded completely. The cause of the lesions was still unclear, however, a plausible cause could have been exposure to chlorhexidine mouthwash and any excipients present in the product. Further more, the patient was advised to take note of any further changes, signs and symptoms, especially when using specific products.

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PLASMA CELL GINGIVITIS – A CHALLENGE FOR THE DENTAL PROFESSION



Figure 1: Clinical photo of ill-defined white patch extending between the LL5 and 6 together with inflamed anterior attached gingiva



Figure 2: Clinical photo of white patch surrounded by a well-defined erythematous border associated with the UR5



Figure 3: Clinical photo of white patch on attached mucosa UL5-6 and erythema surrounding the gingival margin of UL3

Continues from page 31

Management

When managing an erythematous gingival lesion caused by plasma cell gingivitis one must consider whether the patient has symptoms or not. If the patient is asymptomatic the clinician may simply just review and monitor the lesion regularly for any changes or symptoms.

Once a lesion becomes symptomatic and the patient starts to complain of pain, burning or even brings up aesthetic concerns, local factors should be addressed, while maintaining good oral hygiene. Topical steroids can also be prescribed to control symptoms.

Moreover, a thorough clinical history should be taken to attempt identifying the causative factor, in which case, the allergen should be removed. Additionally, if the condition is chronic and persistent, a dermal patch test can be used to identify the allergen to avoid it. (Negi et al., 2019)

The management of plasma cell gingivitis is summarised in a flow chart depicted in figure 4.

CONCLUSION

In general, PCG presents a very big challenge to the dental professional as it is very difficult to identify clinically as it can be easily mistaken for other conditions such as those of the vesiculo-bullous type.

Moreover, it may be difficult to treat especially if its cause cannot be clearly identified. However, this condition typically responds quite well to topical and systemic steroids as seen in this case.

DISCUSSION

Plasma cell gingivitis (a.k.a atypical gingivostomatitis, idiopathic gingivostomatitis, plasma cell gingivostomatitis)

Plasma cell gingivitis is a rare, benign condition of the gingiva with a rapid onset in the free and attached gingivae. It is characterised by sharply demarcated erythematous and oedematous gingivitis often extending to the mucogingival junction.

Localised or diffuse oedematous and erythematous gingival tissues

are present and these tend to bleed easily and lose their normal stippling with or without ulceration. (Janam et al., 2012) This condition can affect any gender at any age however there is a higher prevalence in women. It can also affect any area in the mouth. It is usually asymptomatic but may occasionally cause some pain or a burning sensation.

This condition is usually due to a hypersensitivity reaction to some antigen such as flavouring agents which could be found in certain oral hygiene products.

The absence of desquamation and a negative Nikolsy sign usually helps to differentiate the condition from those of the vesiculobullous type. (Joshi & Shukla, 2015)

Early diagnosis is essential as this condition has similar pathologic changes which are also seen in leukaemia, HIV, infection, discoid lupus erythematosus, atrophic lichen planus, desquamative gingivitis or cicatricial pemphigoid. Therefore these conditions must be differentiated through haematology and serological testing. (Janam et al., 2012)

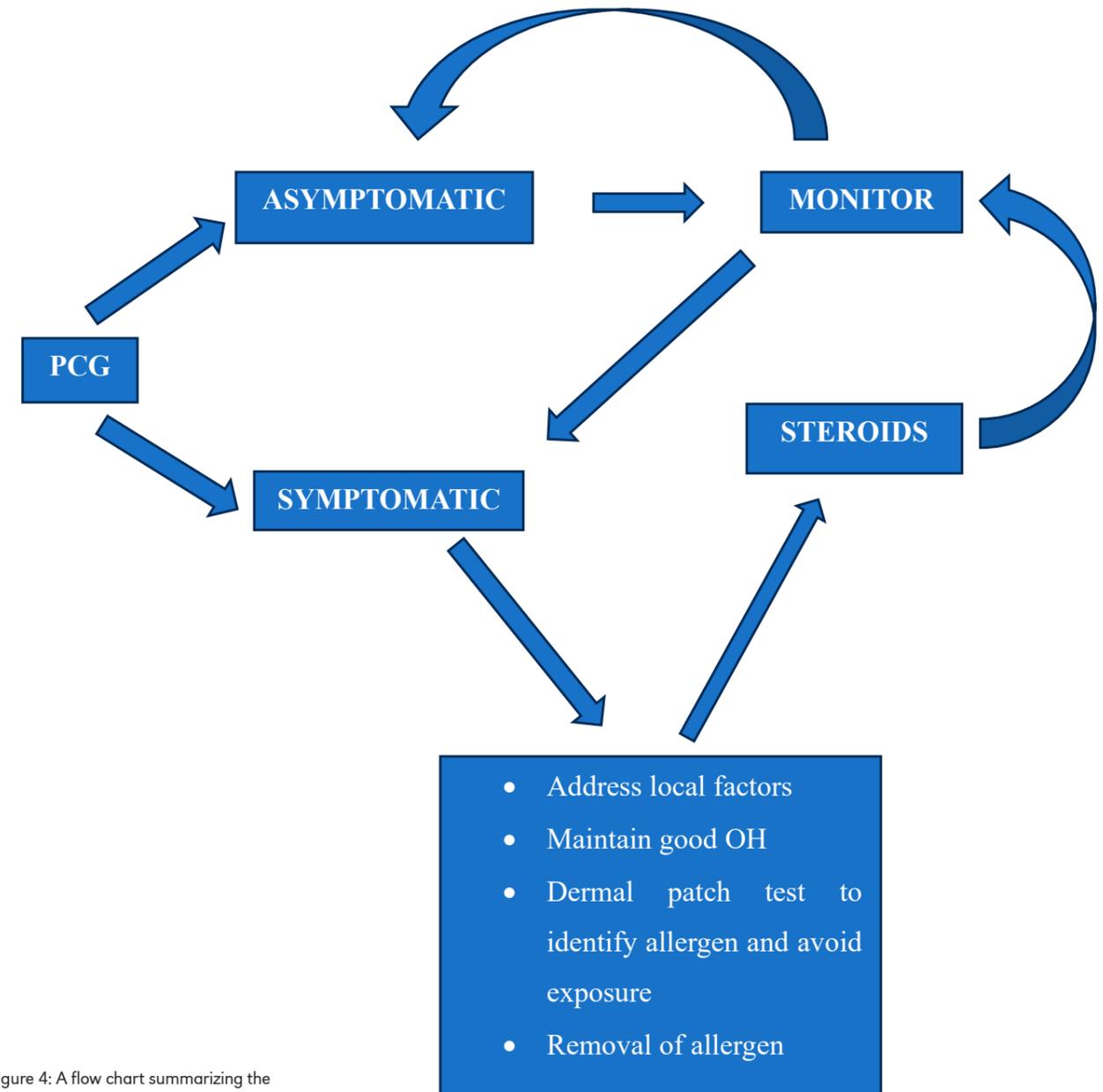


Figure 4: A flow chart summarizing the management of plasma cell gingivitis

BIBLIOGRAPHY

- Plasma cell gingivitis and its mimics (2023) *Pocket Dentistry*.
- Younis RH, Georgaki M, Nikitakis NG. Plasma Cell Gingivitis and Its Mimics. *Oral Maxillofac Surg Clin North Am*. 2023 May;35(2):261-270. doi: 10.1016/j.coms.2022.10.003. Epub 2023 Feb 15. PMID: 36805902.
- Leuci S, Coppola N, Adamo N, Bizzoca ME, Russo D, Spagnuolo G, Lo Muzio L, Mignogna MD. Clinico-Pathological Profile and Outcomes of 45 Cases of Plasma Cell Gingivitis. *Journal of Clinical Medicine*. 2021; 10(4):830.
- Negi BS, Kumar NR, Haris PS, Yogesh JA, Vijayalakshmi C, James J. Plasma-Cell Gingivitis a Challenge to the Oral Physician. *Contemp Clin Dent*. 2019 Jul-Sep;10(3):565-570. doi: 10.4103/ccd.ccd_776_18. PMID: 32308339; PMCID: PMC7150550.
- Belkacem, R. et al. (2024) Plasma cell gingivitis: an enigmatic entity, *International Dental Journal*.
- Janam P, Nayar BR, Mohan R, Suchitra A. Plasma cell gingivitis associated with cheilitis: A diagnostic dilemma! *J Indian Soc Periodontol*. 2012 Jan;16(1):115-9. doi: 10.4103/0972-124X.94618. PMID: 22628976; PMCID: PMC3357019.
- Joshi C, Shukla P. Plasma cell gingivitis. *J Indian Soc Periodontol*. 2015 Mar-Apr;19(2):221-3. doi: 10.4103/0972-124X.145830. PMID: 26015677; PMCID: PMC4439636.
- Lamdari, N., & Pradhan, S. (2012). Plasma cell gingivitis: a case report. *Journal of Nepal Medical Association*, 52(186), 85–87.

NEVOID BASAL CELL CARCINOMA SYNDROME WITH EMPHASIS ON DIAGNOSTIC CRITERIA

A case report by Laura Falzon, Master of Dental Surgery – Year V
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ABSTRACT

Nevoid Basal Cell Carcinoma Syndrome (NBCCS) is a genetic disorder with an autosomal dominant pattern of inheritance. In the majority of patients, diagnosis is relatively straightforward, however in some, it can be challenging because of the variable expressivity of the syndrome. Clinical manifestations include basal cell carcinomas (BCCs), odontogenic keratocysts (OKCs), palmar and/or plantar pits, lamellar calcifications of the falx cerebri, and medulloblastoma at a young age as major criteria, as well as a number of minor criteria. In our case, the diagnosis of the NBCCS was made on the basis of three major criteria; multiple OKCs in the mandible and maxilla, palmar pitting and lamellar calcification of the falx cerebri.

KEYWORDS

- Nevoid Basal Cell Carcinoma Syndrome
- Basal cell carcinomas
- Odontogenic Keratocysts
- Enucleation

ABBREVIATIONS

- BCC Basal Cell Carcinoma
- CBCT Cone Beam Computed Tomography
- CT Computed Tomography
- GA General Anesthesia LA - Local Anaesthesia
- MDH Mater Dei Hospital
- MRI Magnetic resonance imaging
- NBCCS Nevoid Basal Cell Carcinoma Syndrome
- OKC Odontogenic Keratocyst
- OPG Orthopantomogram

INTRODUCTION

Gorlin-Goltz syndrome or Nevoid Basal Cell Carcinoma Syndrome (NBCCS) was first separately described by Jarisch and White in 1894 (Jarisch, 1894, White, 1894). The syndrome was first characterized in 1960, by Gorlin and Goltz. They identified three diagnostic features namely “multiple nevoid basal cell epithelioma, jaw cysts and bifid rib syndrome” (Gorlin & Goltz, 1960). These features were later revised by Rayner et al., who recognized that cysts had to appear together with calcifications of the falx cerebri, or plantar and palmar pits, in order to establish a diagnosis (Rayner et al., 1977). The current diagnostic criteria for establishing the presence of the syndrome were formulated by Evans et al. in 1993 (Evans et al., 1993). These diagnostic criteria will be discussed in depth in this case report.

CASE REPORT

In April 2017, a 35-year-old female was referred to Mater Dei Hospital (MDH) for an Orthopantomogram (OPG) because of right-sided facial swelling. On extra-oral examination, a swelling at the right nasal ala and maxilla was visible. Intra-orally, a labial swelling extending from the upper right central incisor to the upper right first premolar was noted. Teeth 18, 16, 13, 23, 25, 28, 36 and 46 were not visible intra-orally. The upper right lateral incisor was drifted. The OPG revealed a radiolucent lesion which contained the unerupted upper right canine and unerupted upper right third molar (Figure 1). The expansile

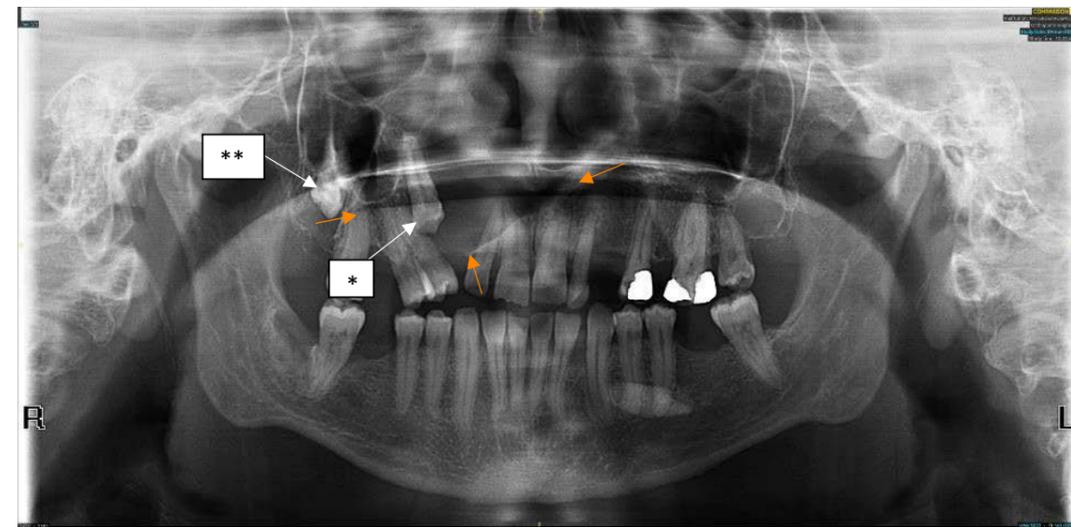


Figure 1: An OPG showing a radiolucent lesion (orange arrows) containing an unerupted upper right canine (*) and an unerupted upper right third molar (**).

lesion had ill-defined borders and was causing displacement of the upper right lateral incisor and upper right premolars. The unerupted teeth were very high up. The patient was referred for Cone Beam Computed Tomography (CBCT) to aid planning for surgery.

In June 2017, enucleation of the radiolucent lesion and extraction on the upper right 3 and 8 were carried out under General Anaesthesia (GA). Histology showed an odontogenic keratocyst (OKC) (Figure 2 and Figure 3). Two months later, in a review appointment, it was noted that the patient had healed well. The risk of recurrence was explained to the patient.

In January 2018 a radiolucent lesion in the upper left third molar area was noted on follow up OPG (Figure 4) and on a CT scan. The CT scan also showed calcification of the falx cerebri. The cyst was treated by enucleation under local anaesthesia (LA). Histology showed a small early odontogenic keratocyst with an adjacent odontogenic fibroma.

In May 2019, a new radiolucent lesion associated with the lower right third molar was detected on an OPG (Figure 5).

A CT taken in September 2019 showed:

- A radiolucent lesion in the upper right quadrant, at the premolar-molar region (possibly a recurrence of the first cyst that was excised)
- A radiolucent lesion in the lower right quadrant at the angle of the mandible
- A radiolucent lesion on the right palatine process

All lesions were enucleated under GA and were diagnosed as odontogenic keratocysts on histology. In the review appointments, the patient was noted to be healing well. Investigation for Nevoid Basal Cell Carcinoma Syndrome was initiated, in view of the multiple OKCs. Plantar pits were present. A brain CT scan, taken in November 2020 confirmed calcification of the falx cerebri (Figure 6).

In July 2021, the patient presented with a flesh-coloured skin nodule on the pre-sternal area, measuring 8mm in maximum dimension. The lesion was excised by a dermatologist. Histology showed a BCC (Figure 7).

OPGs taken in September of 2021 and in September 2022, showed no obvious signs of recurrences, however, the patient needed treatment for submandibular gland sialolithiasis. The patient is being followed up and is being monitored for recurrences and for new lesions.

DISCUSSION

Aetiology

NBCCS is inherited with an autosomal dominant pattern. In the vast majority of cases the disease is caused by a mutation in the PTCH1 gene which is a tumour suppressor gene forming part of the sonic hedgehog signaling pathway.

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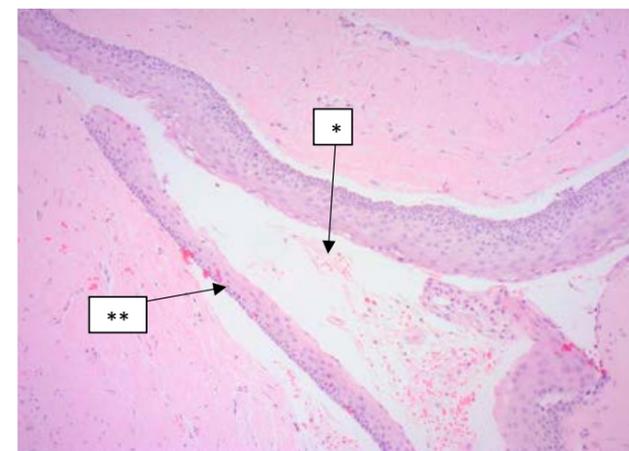


Figure 2: Low power view of the keratocyst of the maxilla. Arrows show the cystic cavity (*), and the lining epithelium (**). (H&E stain, original magnification x100).

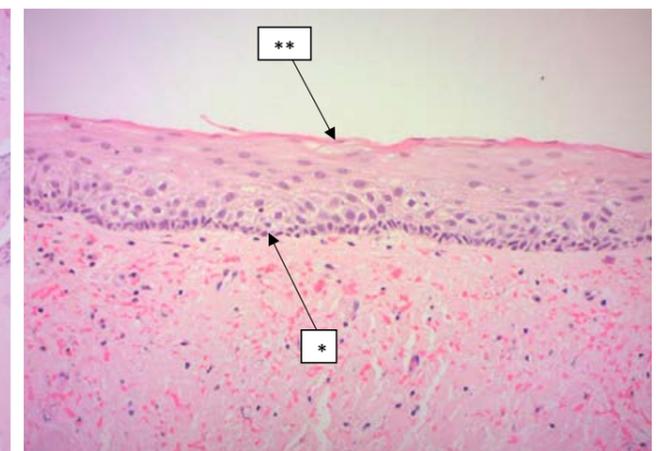


Figure 3: High power view showing the cyst lining epithelium which is only a few layers thick, with a thick prominent basal cell layer (*) and a parakeratinised corrugated surface (**). (H&E stain, original magnification x200).

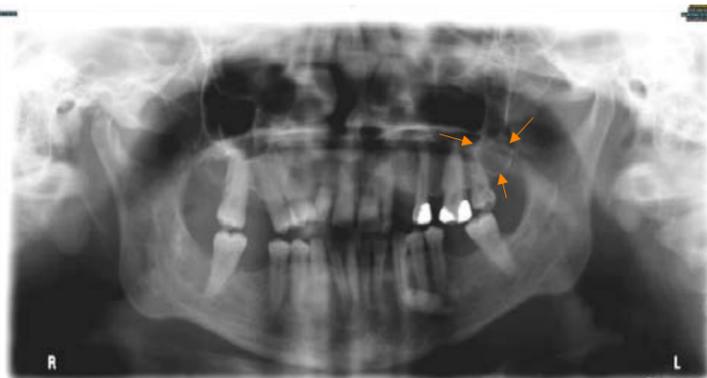


Figure 4: An OPG showing a radiolucent lesion (arrowed) at the upper left third molar



Figure 5: An OPG showing a radiolucent lesion (arrowed) associated with the lower right third molar

Continues from page 35

In around 5% of cases there is an alternate mutation in another component of the same pathway; the SUFU gene. Occasionally, mutations in the PTCH2 gene have been observed in patients with NBCCS (Igaz et al., 2022).

Epidemiology

The prevalence of NBCCS is estimated at 1 per 40,000-60,000. The syndrome affects males and females in equal distribution (Sangera & Grewal, 2018).

Clinical Manifestations

Various signs and symptoms may be linked to NBCCS. These include BCCs, which may show up very early, OKCs, plantar or palmar pits, and lamellar calcifications of the falx cerebri. These manifestations are considered as the major diagnostic criteria. Other clinical features that are considered as minor criteria have also been also reported (Evans et al., 1993, Muzio, 2008).

Diagnosis

In the majority of patients, diagnosis is relatively straightforward, however, it can be challenging in some because of the variable expressivity of the syndrome, particularly in dark-skinned patients, who may not develop basal cell carcinomas. Diagnosis of NBCCS can be made in the presence of: (Evans et al., 1993)

- a. 2 major criteria, or
- b. 1 major criterion and molecular confirmation of PTCH1 gene mutation, or
- c. 1 major and 2 minor criteria.

In our case, the diagnosis of NBCCS was made due to the presence of three major criteria:

1. Multiple OKCs in the mandible and maxilla
2. Palmar pitting
3. Lamellar calcification of the falx cerebri

The patient also subsequently developed a basal cell carcinoma on the pre-sternal area, however, since she was older than 20 years, that doesn't count as a major criterion.

Investigations

The following investigation protocol was recommended by Muzio: (Muzio, 2008).

- Detailed medical, dental and family history - A first-degree relative with NBCCS is a major diagnostic criterion.
- Clinical examination - Oral, skin, interpupillary distance, eyes, head circumference, cardiovascular system, skeletal system, respiratory system, central nervous system, and genitourinary system - to examine for abnormalities such as BCCs, macrocephaly, ocular abnormalities, cleft lip and/or palate, palmar or plantar pitting.
- Genetic testing and DNA analysis
- Radiography - Anteroposterior and lateral skull, OPG, chest, hands, cervical and thoracic spine, pelvic (female). These radiographs are recommended to look for any abnormalities such as rib abnormalities, vertebral anomalies, short fourth metacarpals, and OKCs.
- Ultrasound of the ovaries in female patients - to check for ovarian fibromas.
- Echocardiography in children - to check for cardiac fibromas
- Brain magnetic resonance imaging (MRI) - to check for lamellar calcification of the falx cerebri and to detect any medulloblastomas.

Genetic Testing

NBCCS is an inherited autosomal dominant condition, thus referral to a genetic counselor is a critical component of the ongoing care of the patient. NBCCS is caused when a germline mutation occurs in one of the two PTCH1 genes. Therefore, every offspring of an individual with NBCCS has a 50% probability of inheriting the mutated gene.

Approximately 70-80% of diagnosed patients have at least one affected parent. The remaining 20-30% of cases represent de novo mutations (Muzio, 2008).

The genetic status of the parents affects the risk to a proband's sibling. Siblings are at 50% risk if one parent of the proband has the condition. If the parents do not show signs and symptoms of the

syndrome or if the mutation cannot be demonstrated in their DNA, the risk to siblings of a proband seems to be minimal, however higher than that of the general population (Muzio, 2008). The quantity of OKCs and BCCs encountered in individual patients may depend on particular genetic variations. It is advised that families take part in routine screening. DNA tests may be able to establish whether or not someone in a family has inherited the disorder (Muzio, 2008).

Histopathology

BCCs and OKCs developing in patients with NBCCS are identical to sporadic tumours on histology (Bresler, Padwa, & Granter, 2016). Therefore, diagnosis of NBCCS hinges on clinical, not histological features, hence the development of diagnostic criteria.

Screening and Treatment

Management of patients with the syndrome mandates a multidisciplinary approach. There are two main

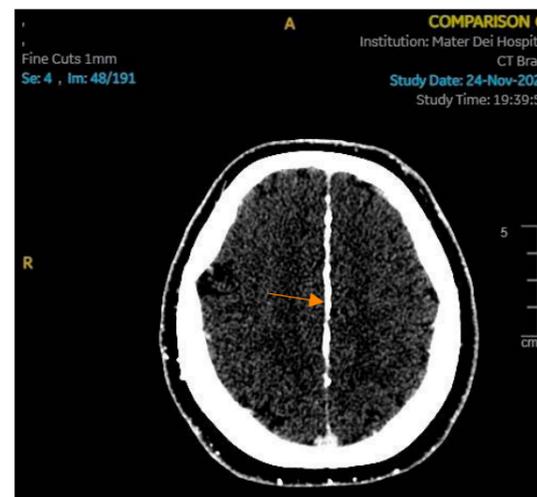


Figure 6: A CT scan of the brain showing calcification of the falx cerebri (arrowed)

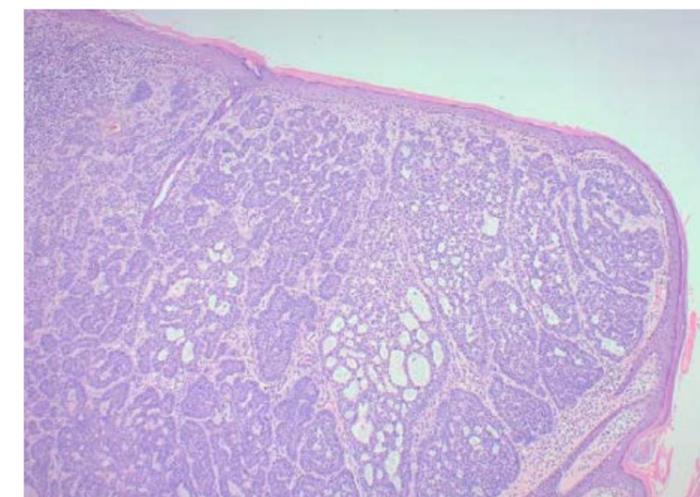


Figure 7: Low power view of the excised pre-sternal lesion showing a nodular and adenoid BCC infiltrating the dermis. (H&E stain. Original magnification x100)

surgical methods that can be used in the treatment of OKCs:

1. Marsupialization – a technique in which the cyst is converted into a pouch so that eventually, the lesion will be decompressed.
2. Enucleation – a reliable and effective technique to treat cysts. Since the cystic capsule is completely removed, the risk of recurrence is reduced. Carnoy's solution is sometimes used as an adjunct for the management of syndromic OKCs. It is safe and effective as it destroys epithelial islands or daughter cysts after enucleation. Application of the solution before enucleation of the cyst or on the exposed osseous wall for three minutes after enucleation, proves to decrease the risk of recurrence. (Chandran et al., 2015)

More aggressive methods such as peripheral ostectomy and resection, should be considered only when there is:

- a recurrence
- a multilocular aggressive intraosseous OKCs
- a diagnosed OKC displaying unusually aggressive clinical behaviour (Lahcen et al., 2018)

Management of BCCs is influenced by the histological subtype, location and size of the lesion. Due to the high tumour burden that is typically experienced, a person with NBCCS may undergo repeated surgery, which can result in significant deformity. Conventional therapies for localized disease include use of topical agents such as imiquimod and 5-fluorouracil.

Radiation therapy is contraindicated in patients with NBCCS. This is because patients with NBCCS are more prone to get BCCs in body parts exposed to high levels of ionizing radiation. Targeted therapy is being

investigated. A phase II trial investigating the effectiveness of vismodegib (a chemotherapy drug that inhibits the hedgehog pathway) in patients with the syndrome found a decrease in the number of BCCs in the treatment group when compared to the control group. This was found to be statistically significant. The majority of the individuals treated with vismodegib however, stopped using it due to adverse effects such as hair loss, muscle cramps and weight loss. In addition, many patients with non-syndromic BCCs that are treated with vismodegib, eventually develop resistance to the drug (Tang et al., 2012).

In another study, Vismodegib provides an alternative to surgery for the management of OKCs.

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TABLE 1: DIAGNOSTIC CRITERIA FOR NEVOID BASAL CELL CARCINOMA SYNDROME (EVANS ET AL., 1993)

	Diagnostic Criteria
Major Criteria	Multiple basal cell carcinomas or single, which occur in patients under 20 years of age
	Odontogenic keratocyst (OKC) of the jaws (with histological confirmation)
	Plantar or palmar pitting
	Lamellar calcification of the falx cerebri
	First-degree relative with NBCCS
	Desmoplastic or nodular medulloblastoma in a child aged < 4 years
Minor Criteria	Macrocephaly
	Congenital abnormalities: frontal bossing, coarse face, cleft lip or palate, moderate or severe hypertelorism.
	Other skeletal abnormalities: marked syndactyly of the digits, marked pectus deformity, Sprengel deformity.
	Radiographical abnormalities: modeling defects of the hands and feet or flame-shaped hands or feet, bridging of the sella turcica, vertebral anomalies such as hemivertebrae, fusion or elongation of the vertebral bodies.
	Ovarian or cardiac fibroma.
	Medulloblastoma.

NEVOID BASAL CELL CARCINOMA SYNDROME WITH EMPHASIS ON DIAGNOSTIC CRITERIA

Continues from page 37

This may be crucial for patients with NBCCS, who may experience speech impairment and facial deformities as a result of multiple surgical procedures. Additional research on the most appropriate maintenance schedules and long-term efficacy should however be conducted (Ally et al., 2015).

Postoperative follow-up is essential. It is recommended that between the ages of 8 and 40, a yearly OPG is performed to help detect the development of new OKCs or their recurrence. It is advised that NBCCS patients get a dermatological exam every three to six months. Sun protection is a crucial preventative practice. Children must have neurological testing every six months since they are more likely to develop medulloblastoma (Chandran et al., 2015).

In our case, all OKCs were surgically treated and any compromised teeth were extracted. The surgical approach was always enucleation. This approach was taken considering the size of the lesions. All cystic lesions removed were sent for histological analysis, and all were found to be OKCs.

The patient was educated about the importance of sun protection as a preventive care measure. This is important as the patient had already developed a basal cell carcinoma.

The odontogenic fibroma that developed in the left posterior maxilla was an incidental finding. Odontogenic fibromas have not been previously described in NBCCS and they are not considered as forming part of the spectrum of the syndrome.

CONCLUSION

The presence of three major criteria; multiple OKCs in the mandible and maxilla, palmar pits and, lamellar

calcification of the falx cerebri; confirmed that our patient represents a case of NBCCS. Identifying the presence of the syndrome early in life will lessen the likelihood of complications and their severity including neoplasia and maxillofacial deformities related to the jaw cysts. Targeted therapy may in future play a role in managing skin lesions and OKCs, however further research is needed in this area. Health specialists especially dentists must have good knowledge of the features of NBCCS so that the patient can be diagnosed and treated early. ■

REFERENCES

- Ally, M. S., Tang, J. Y., Joseph, T., Thompson, B., Lindgren, J., Raphael, M. A., ... Epstein, E. H. (2014). The Use of Vismodegib to Shrink Keratocystic Odontogenic Tumors in Patients with Basal Cell Nevus Syndrome. *JAMA Dermatology*, 150(5), 542. <https://doi.org/10.1001/jamadermatol.2013.7444>
- Bresler, S. C., Padwa, B. L., & Granter, S. R. (2016). Nevoid Basal Cell Carcinoma Syndrome (Gorlin Syndrome). *Head and Neck Pathology*, 10(2), 119–124. <https://doi.org/10.1007/s12105-016-0706-9>
- Chandran, S., Karthikeyan Marudhamuthu, Riaz, R., & B. Saravanan. (2015). Odontogenic Keratocysts in Gorlin-Goltz Syndrome: A Case Report. *PubMed*, 7(Suppl 1), 76–79.
- Evans, D. G., Ladusans, E. J., Rimmer, S., Burnell, L. D., Thakker, N., & Farndon, P. A. (1993). Complications of the naevoid basal cell carcinoma syndrome: results of a population based study. *Journal of Medical Genetics*, 30(6), 460–464. <https://doi.org/10.1136/jmg.30.6.460>
- Gorlin, R. J., & Goltz, R. W. (1960). Multiple nevoid basal-cell epithelioma, jaw cysts and bifid rib. A syndrome. *New England Journal of Medicine*, 262(18), 908–912. <https://doi.org/10.1056/nejm196005052621803>
- Igaz, P., Toth, G., Nagy, P., Dezső, K., Turai, P. I., Medvecz, M., Wikonkal, N., Huszty, G., Piros, L., Toth, E., Bozsik, A., Likó, I., Patócs, A., & Butz, H. (2022). Surprising genetic and pathological findings in a patient with giant bilateral periaxillary tumours: PEComas and mutations of PTCH1 in Gorlin-Goltz syndrome. *Journal of Medical Genetics*, 59(9), 916–919.
- Jarisch, W. (1894). On the doctrine of skin tumors. *Archiv of Dermatology and Syphilis*, 28, 163–222.
- Kumar, N. N., Padmashree, S., Jyotsna, T. R., & Shastry, S. P. (2018). Gorlin-Goltz Syndrome: A Rare Case Report. *Contemporary Clinical Dentistry*, 9(3), 478–483. https://doi.org/10.4103/ccd.ccd_96_18
- Lahcen, K., Jalal, H., Mohamed Kamal, F., Abibou, N., Yassamina, R., & Karim, E. (2018). Odontogenic keratocysts in gorlin-goltz syndrome: how to manage? *Oral Health and Care*, 3(2), 1–3. <https://doi.org/10.15761/ohc.1000140>
- Muzio, L. L. (2008). Nevoid basal cell carcinoma syndrome (Gorlin syndrome). *Orphanet Journal of Rare Diseases*, 3(1), 3–32. <https://doi.org/10.1186/1750-1172-3-32>
- Rayner, C. R. W., Towers, J. F., & Wilson, J. S. P. (1977). What is Gorlin's syndrome? The diagnosis and management of the basal cell naevus syndrome, based on a study of thirty-seven patients. *British Journal of Plastic Surgery*, 30(1), 62–67. [https://doi.org/10.1016/s0007-1226\(77\)90037-6](https://doi.org/10.1016/s0007-1226(77)90037-6)
- Sangera, R., & Grewal, P. (2018). Gorlin Syndrome Presentation and the Importance of Differential Diagnosis of Skin Cancer: A Case Report. *Journal of Pharmacy & Pharmaceutical Sciences*, 21(1s), 222s–224s. <https://doi.org/10.18433/jpps30150>
- Tang, J. Y., Mackay-Wiggan, J. M., Aszterbaum, M., Yauch, R. L., Lindgren, J., Chang, K., ... Epstein, E. H. (2012). Inhibiting the Hedgehog Pathway in Patients with the Basal-Cell Nevus Syndrome. *New England Journal of Medicine*, 366(23), 2180–2188. <https://doi.org/10.1056/nejmoa1113538>
- White, J. C. (1894). Multiple benign cystic epitheliomas. *J Cutan Genitourin Dis*, 12(4), 477–484.

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