

Atypical presentation of a Rib Chondrosarcoma

Samuel Zahra, Kevin Schembri

Abstract

Background: This case report represents a variation from the typical characteristics of an uncommon cardiothoracic pathology, i.e rib chondrosarcomas. Usually this pathology is seen in patients over 40 years of age and grows relatively slowly, taking around 2 years to present clinically.

Case Presentation: Our patient was an asymptomatic and healthy 27 year old male who presented with a large right sided thoracic mass. CT (Computed Tomography) scanning revealed an inhomogeneous lesion around 6cm in diameter, arising from the 8th right rib, with no obvious signs of aggressive type of growth and no evidence of metastatic spread. The decision was taken to excise the lesion based on clinical and radiological evidence. Histopathological analysis was carried out at two centres and reported a Grade II 55mm x 48mm x 43mm show with a variably cellular tumour consistent with chondrosarcoma. The case was discussed with the oncological team who advised no need for further treatment given histology and radiological report, except clinical and radiological surveillance.

Conclusions: Chondrosarcomas are the 3rd commonest type of bone tumour however it is considered rare for them to originate from the ribcage in a young individual over a relatively short time span. CT scanning is considered the gold standard image and surgery as the main form of management.

Keywords

rib, chondrosarcoma, cardiothoracic, oncological

Background

Chondrosarcomas represent a heterogeneous group of bone tumours, common trait being the ability to produce chondroid matrix.¹ The majority of these tumours arise from the pelvis or long bones.²⁻⁴ Malignant primary tumours of the thoracic wall account for 4.5-8% of all bone tumours with chondrosarcomas representing 40%.⁵ Biological behavior varies according to site and grade.² CT scanning is considered to be the gold standard for diagnosis and surgical planning¹, with the commonest finding showing a low density mass with coarse calcifications³. We report a case of a chondrosarcoma located on the right 8th rib, in a young healthy male patient, whose only complaint was the rapidly growing mass.

Case Presentation

27 year old Caucasian patient, presented to the emergency department, complaining of a lump he noted on his right side of his chest. The lump grew from non-palpable to palpable over a timespan of 2 months. It was not painful, did not discharge and had visible punctum. No history of trauma was given and it was not affecting his daily activities.

Patient claimed he regularly smokes 2 packets of tobacco a day but did not note any worsening shortness of breath, cough, sputum, fever or weight loss.

Patient had no prior medical or surgical history. He worked in the delivery system and lived with his mother. A family history of high blood pressure and diabetes was present.

On examining the patient cardiovascular examination revealed normal heart sounds and bilateral vesicular air entry despite his smoking habits. Abdominal examination revealed a 3 finger

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breath hepatomegaly but no signs of jaundice or other stigmata of hepatic pathology.

The lesion itself was found to be located on the right side of the lower chest wall. It was medial to the mid-axillary line with no obvious signs of erythema. The lump was solid in nature, non-tender and had a well defined smooth border which could be traced as originate from the 8th right rib.

Routine blood results were taken including a Full blood count and inflammatory markers- which failed to show signs of ongoing inflammatory process.

A Chest X-ray was ordered and exhibited a solid mass originating from the lower right chest wall (Figure 1). This was followed by a CT scan to get more detailed radiological information.

The CT report commented on the presence of an inhomogeneous mass about 6cm in diameter, arising from the 8th right rib with no obvious signs of aggressive type of growth. Note was also made of the mass pushing on the right liver lobe (Figure 2,3).

Our main differential was some form of rib tumour, suspecting a rib chondroma or chondrosarcoma.

Surgery

The decision was taken to remove the tumour surgically by the cardiothoracic department. This involved using a general anaesthetic to gain access to the lesion. The lesion in theatre was noted to be a well defined mass originating from the 8th right rib. It was highly vascular and there was no indication that the mass was in contact with the rib superior to it (Figure 4). Medially the mass was abutting the right liver lobe and was displacing it but again no signs of obvious liver involvement was noted. The lesion was removed together with a 4cm segment of unaffected bone medially and laterally (Figure 5).

The defect created was reinforced using non-absorbable sutures and the lesion was sent to histopathology and a chest drain was inserted.

Histology report

Macroscopic: A rib segment which measures 95mm in length and 10mm in diameter. A firm, nodular, lobulated tumour occupies the middle third of the rib. The tumour measures 55mm x 48mm x

43mm and is 27mm away from the closest surgical margin. The tumour has a diffusely homogenous chondro-myxoid cut surface.

Microscopic: Sections from the rib lesion show a variably cellular tumour composed of atypical chondrocytic cells set in a predominantly chondroid tumoural matrix. The chondrocytes exhibit mild-to-moderate nuclear pleomorphism and both binucleate and trinucleate forms are readily identified. However, no appreciable mitotic activity is present. In areas, neoplastic non-mineralised and mineralised osteoid formation is seen, in which entrapped tumour cells are present. Mucomyxoid degeneration of the stroma is visualised in areas. The tumour focally infiltrates the bone marrow (Figure 6,7,8).

Diagnosis

Excision of right 8th rib tumour:
Chondrosarcoma, grade 2.

Oncology

The results were then discussed with the oncological team who advised, that chemo-radiotherapy or further resections were unnecessary.

Discussion

Chondrosarcomas are considered to be very rare malignant tumours when growing from the ribs. Typically they present in an older age group, however the patient in this case was 27 years at the time of presentation.^{1,6} Described as slow growing, the patients' tumour grew from clinically palpable to pre-resection size in less than 2 months.

Diagnosing rib chondrosarcomas involves a combination of clinical and radiological investigations. With respect to imaging, CT is considered to be the golden standard, even though most lesions will likely be seen on a plain chest X-ray.⁶ In fact biopsy is not required prior to surgery, highlighting the importance of CT imaging to help deciding to proceed with surgical intervention as the next step.

The most effective treatment is surgical resection with a healthy surgical margin as was done in the situation of our patient³, with adjuvant chemo/radiotherapy therapy not playing a major role

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in managing these sort of tumours.⁵ The outcome from surgical management for rib chondrosarcomas is more favourable when compared to chondrosarcomas originating from other sites in the body; with a 5 year mortality quoted at 10%, local recurrence at 17% and metastatic rate of 12%.⁶ The oncological outcome after surgery is worse in tumors

>5 cm, in tumors with positive resection margins and grade 3 chondrosarcoma.⁷ The patient post procedure will require physical examination and imaging chest X-ray every 3-6 months for the first 5 years. This constitutes the final part of the management plan, surveillance and monitoring.

Figure 1: Lesion seen on AP CXR located in the distal portion of the right rib cage

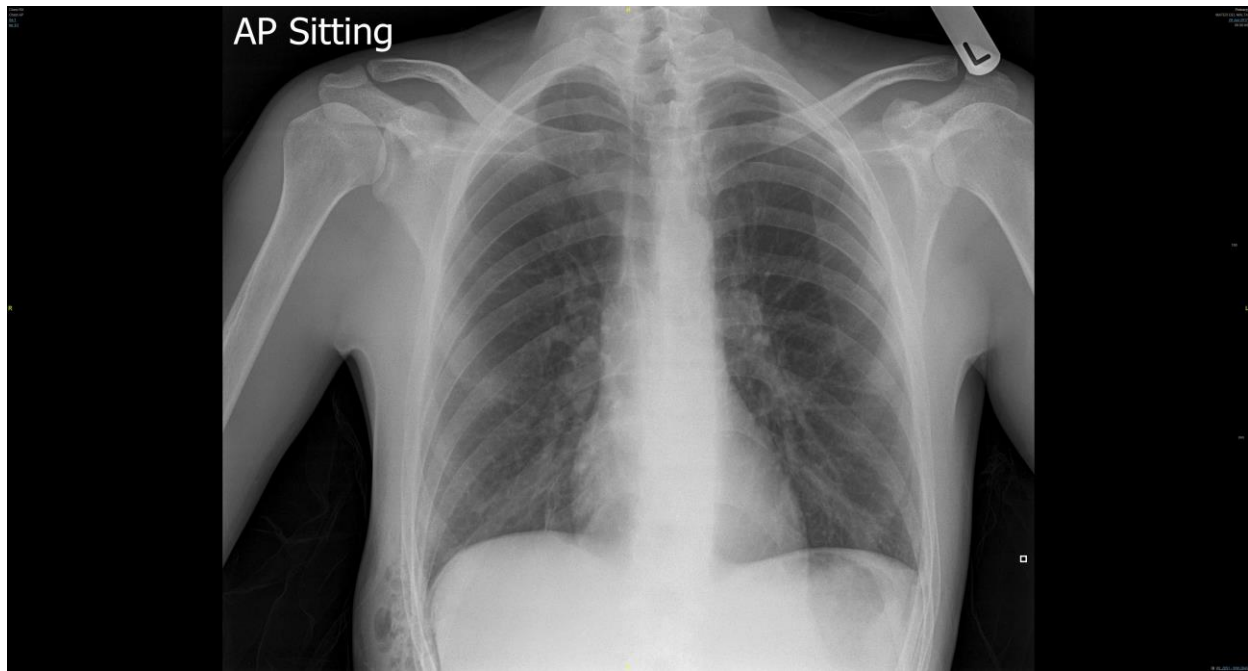


Figure 2: Sagittal CT scan showing lesion growing from the 8th right rib

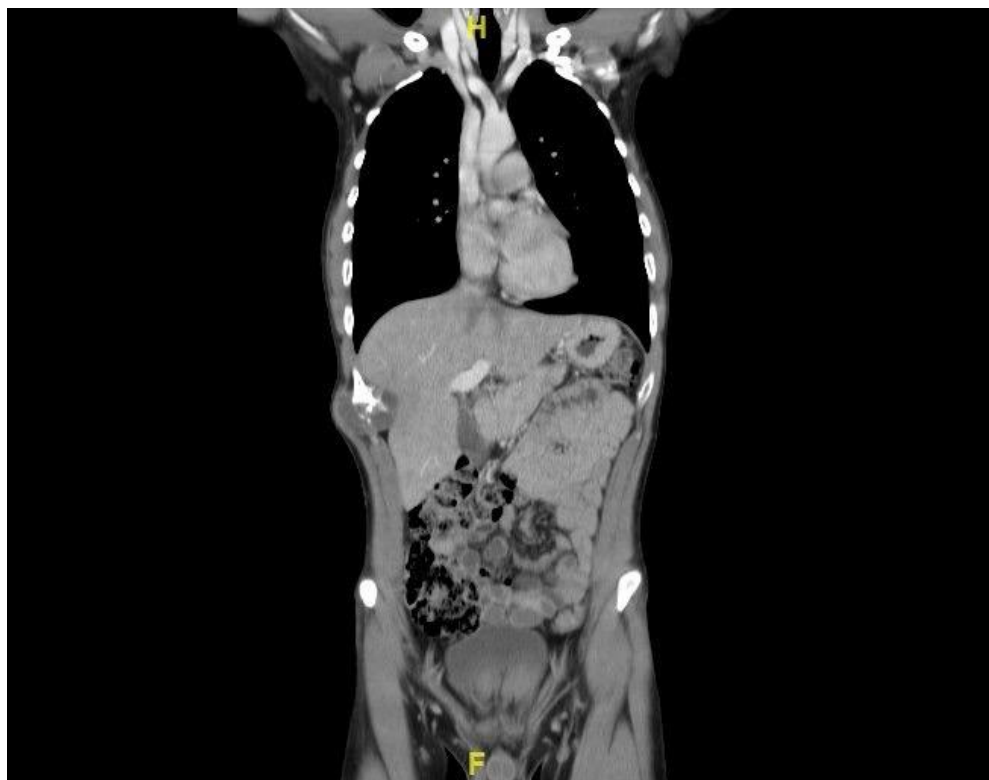


Figure 3: Coronal CT scan showing extent of lesion compressing the right lobe of the liver

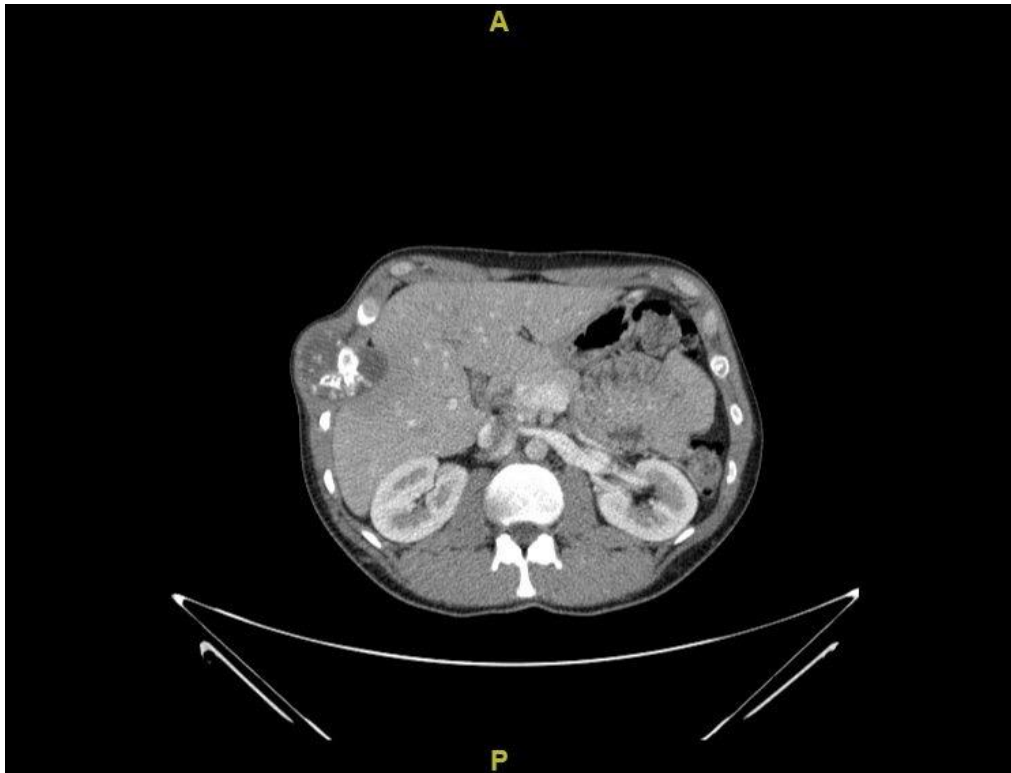


Figure 4: Intraoperative view of the tumour



Figure 5: Gross histological specimen

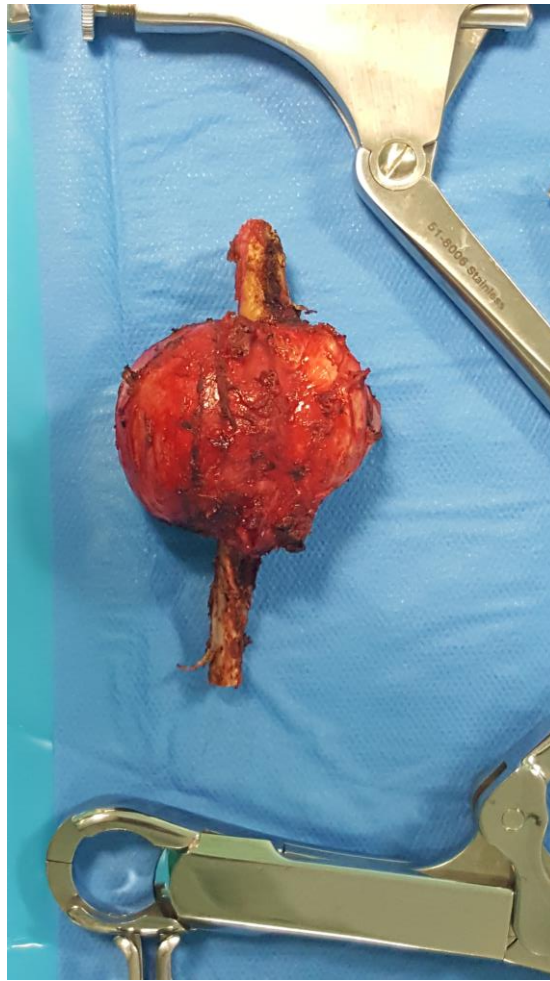
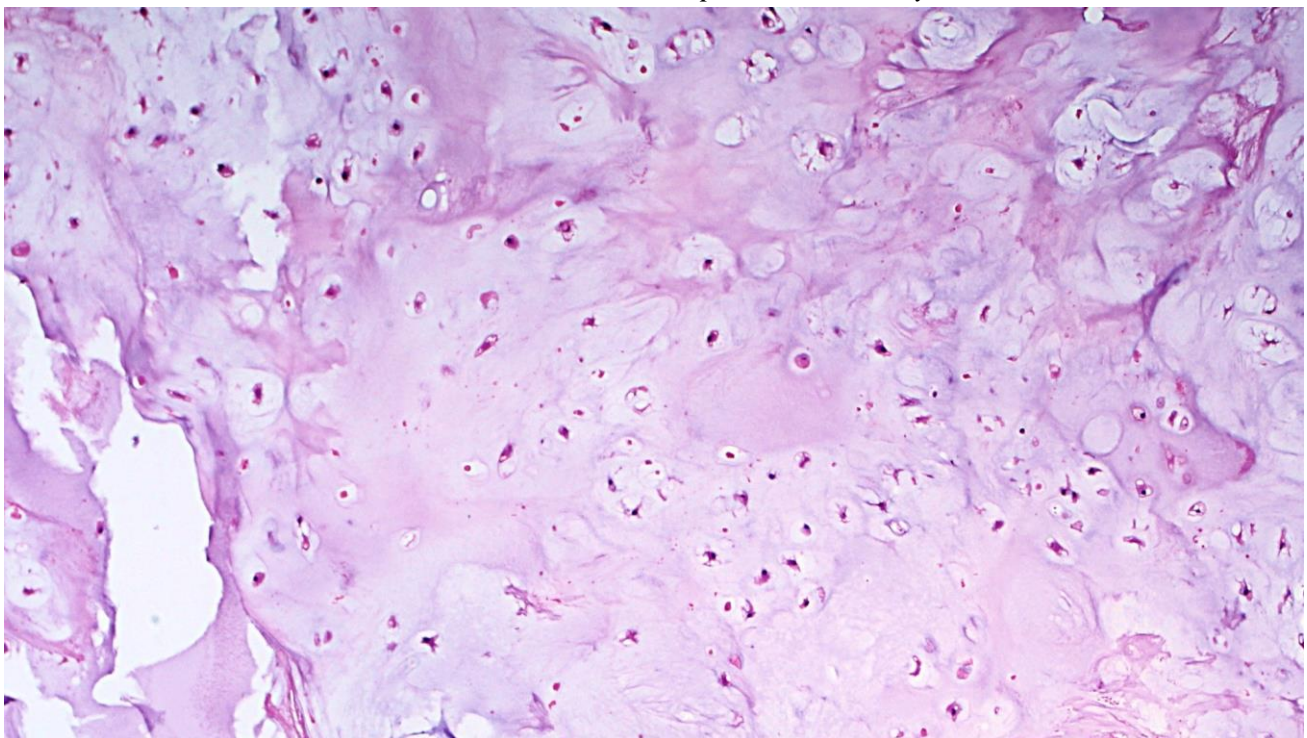


Figure 6: H&E stained slide at x200 magnification which demonstrates the chondroid matrix of the tumour, in which are set numerous neoplastic chondrocytes



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Figure 7: H&E stained slide at x400 magnification. The atypical nature of the chondrocytes wherein the neoplastic cells are pleomorphic, hyperchromatic and have a somewhat stellate morphology.

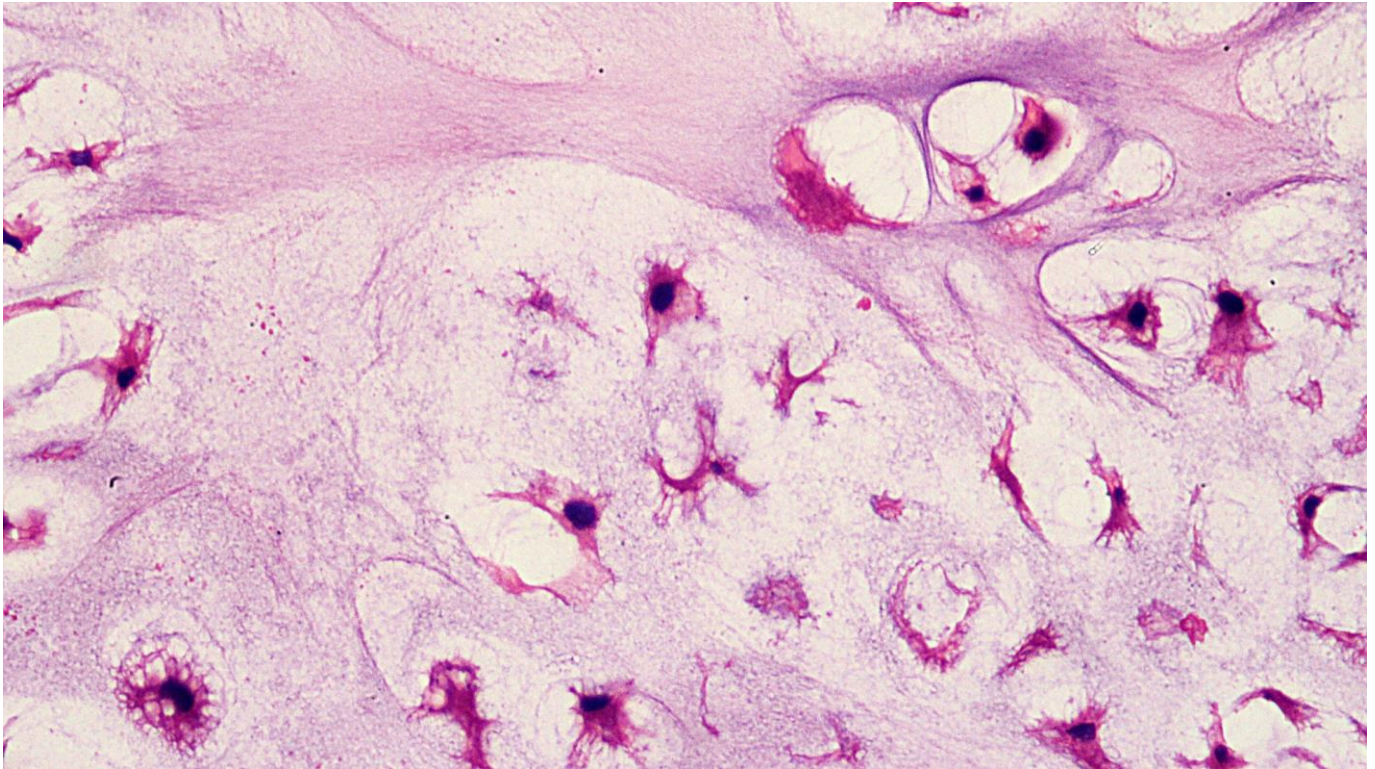
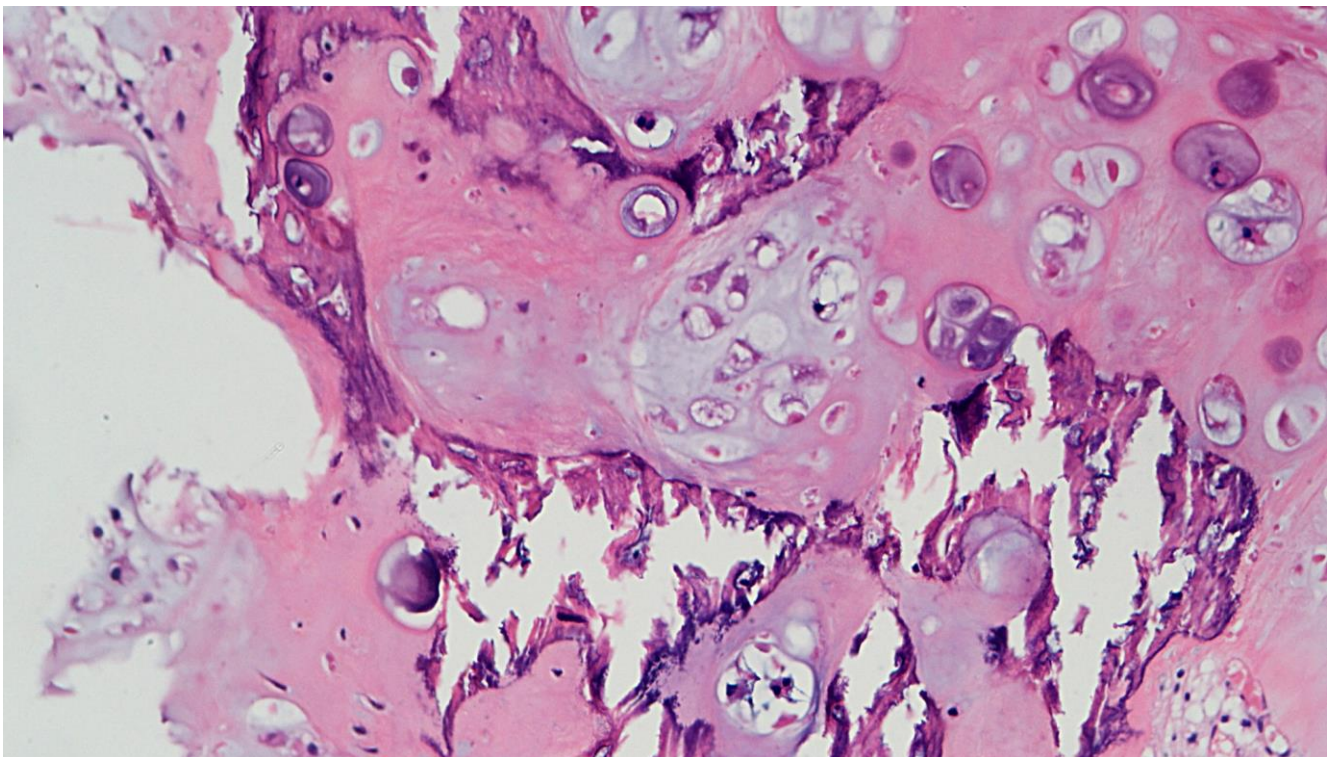


Figure 8: H&E stained slide at x200 magnification. Foci of calcification were also present within the tumour which, in areas, appeared somewhat ossific in nature, which is what prompted referral of the case abroad for further differentiation between chondrosarcoma (within which one is not allowed ossification from a histopathological standpoint) and chondroblastic osteosarcoma. This differentiation is not easy and is aided by molecular and genetic tests (eg: IDH mutations) which are not performed locally.



Conclusions

Chondrosarcoma represent a heterogeneous group of malignancies. Chondrosarcomas account for 20% of bone sarcomas, of which 3.1% arise from the chest wall and 1.8% from the rib. There is a slight male predominance with mean age 47 ± 17.2 years.⁷ Surgical resection of both primary and recurrent chondrosarcoma of the rib is effective and the mainstay method of treatment.

Learning Points:

- Chondrosarcomas are the 3rd commonest type of bone tumour however it is extremely rare for them to originate from the ribcage in a young individual over a relatively short time span.
- CT scanning is considered the gold standard image and surgery as the main form of management.
- Aim is to achieve good resection margins to get R0 resection. Oncological management in the form of chemo-radiotherapy is rarely beneficial and active surveillance with progressively longer intervals form the crux of patient management.

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