GLOMUS JUGULARE TUMOURS
Their Otological and Neurological Importance

LUIS VASSALLO
B.Sc., M.D., M.R.C.P. (Lond.), M.R.C.P. (Edin.), M.R.C.P. (Glasg.)
Consultant Physician

J. A. HARDING
M.D., F.R.C.S. (Edin.), D.L.O. (Eng.)
Consultant ENT Surgeon,

St. Luke's Hospital,
Malta.

Perusal of the literature pertaining to glomus jugulare tumours reveals the long interval which usually passes between the onset of symptoms caused by these slowly growing tumours and the time of diagnosis. The literature about these tumours and the structures from which they arise has been accumulating for less than thirty years as the glomus jugulare was only discovered by Stacy Guild of the Johns Hopkins Hospital in 1941, and the first case of "a tumour in the middle ear and mastoid bone" resembling a carotid body was recorded by Rosenwassed in 1945. Rosenwasser recognised the relationship to the glomus jugulare structure. By the end of 1956, 172 tumours in the vicinity of the glomus jugulare had been recorded (Steinberg and Holz, 1965). Glomus jugulare tumours are nowadays considered to be the most common tumours of the middle ear cleft (Shapiro & Neues, 1964).

We wish to report two further cases illustrating the main different clinical manifestations of these tumours. The first case is especially illustrative because the real cause of a lower motor neurone facial palsy had been for many years unrecognised. The causative pathology can be missed if a full otological and neurological examination is not carried out in patients suffering from a lower motor neurone facial palsy. This is all the more indicated if there has been no sign of nerve function recovery after a few weeks of conservative treatment.

Case Reports

Case 1. A.S., a 31 year old single Maltese woman had had a left sided lower motor neurone facial weakness of sudden onset seven years before being seen by one of us (J.H.). There had also been initial ear ache. Repeatedly questioned, she insisted that she had not experienced any tinnitus. She was seen originally by her general practitioner who diagnosed an ordinary Bell's palsy and referred her to a unit for facial physiotherapy. No improvement in the facial palsy resulted. She consulted other doctors in the first year of her symptoms. No other diagnosis or form of treatment (such as facial nerve decompression) was suggested to her. She was labelled as having suffered from a Bell's palsy from which she had not recovered.

During the last year she had become aware of an increasing pulsating swelling in the region just below her left ear and had also noted that the left side of her tongue was wasting.

Deafness which had been present in the left ear almost since the origin of her symptoms had not bothered her. Her other ear was perfectly normal.

Because of these new developments, she sought medical advice from a new medical practioner, who referred her to one of us (J.H.) with the diagnosis of an external carotid artery aneurysm.

Examination showed a fully orientated young woman, with evidence of old facial

weakness of lower motor neurone type. (Infranuclear). The lower lobe of the left ear could be seen to pulsate as one looked at the patient from in front. The optic discs were normal. There was no diplopia or other evidence of involvement of the external ocular muscles. Hearing was grossly diminished in the left ear. Audiometric assessment showed a conduction type of deafness. A large pulsating mass could be seen blocking the external auditory canal almost completely. The mass appeared to arise from the floor of the canal. Examination with a Jobson-Horne probe easily provoked bleeding.

Neurological examination (by L.V.) showed weakness of the left side of the soft palate. There was loss of sensation over the left posterior pharyngeal wall and the pharyngeal and palatal reflexes could not be elicited on the left side. There was no hoarseness or evidence of paralysis of the vocal cords. The sternomastoids and trapezii were normal. There was gross left hemiatrophy of the tongue with fasciculation. A bruit was present over the mastoid region. No other neurological abnormalities were found. In particular, there were no signs of cerebellar or pyramidal dysfunction. Her blood pressure was 140/80 mm. Hg. Radiographs of the skull showed marked erosion of the petrous bone on the left side.

It was decided that the best line of treatment would be radiotherapy and the patient was accordingly referred abroad for deep radiotherapy.

Case 2. A sixty five years old woman had noted the development of mild deafness and tinnitus in her left ear since fifteen years. The symptoms had slowly increased with time and she was referred for an E.N.T. opinion because her doctor felt the wall of the meatus was so swollen that this obstructed the insertion of an auroscope. When examined by one of us (J.H.) it was realised that the meatus was occluded by a hard tightly fitting fibrous polyp. This "polyp" was removed on 23.12.68. under general anaesthesia using a Zeiss operating microscope. The consistency of the polyp was hard. Bleeding was severe. It was noted that the polyp was emerging from the hypotympanum through a large perforation of the pars tensa of the tympanic membrane.

Histological examination (Prof. G. Xuereb) confirmed the presence of a glomus jugulare tumour. Microscopical examination showed blood vessels and strands of connective tissue separating largish polyhedral cells with vacuolated cytoplasm. Mitotic figures were infrequent and a few binucleate cells were present.

The patient was referred abroad for deep radiotherapy.

Discussion

These cases illustrate incidentally the two main groups of clinical manifestations which tumours of the glomus jugulare give rise to — i.e. the neurological and the aural types. It is not surprising that the glomus jugulare structure was not discovered till 1941, as it is, after all, an almost microscopical structure measuring 0.5 × 0.25 mm. and lies in the dome of the jugulare bulb, just below the floor of the middle ear, and consists of non-chromaffin staining paraganglion cells regularly arranged. The term glomus has been given to this structure because of the fancied resemblance to a ball of wool or thread. Other similar bodies or glomera have also been found in other sites such as along the tympanic branch of the glossopharyngeal nerve (Jackobson's nerve) and along the auricular branch of the vagus nerve (Arnold's nerve). Guild (1951) claimed to have seen these glomera as far distal on the nerve of Arnold as the site where it crossed the descending part of the facial nerve. The practical significance of these different sites of origin of tumours of this class lies in the fact that the very different early manifestations in various cases may be thus explained. It would be quite reasonable to postulate that the very early involvement of the facial nerve in our first patient may have been due to the tumour originating near the distal part of the nerve of Arnold. Zak (1954) in an exhaustive study of the subject, has emphasised that glomus type tissue may also be found in the brain, dura and optic chiasm. According to Black (1952) one half of all cranial glomus tumours originate from glomera within the temporal bone and the rest from the dome of the jugular bulb.

There is considerable disagreement about the nomenclature to be given to these tumours, and different classifications have been adopted by various authors. Thus attempts have been made to classify these tumours according to their presumed site of origin. It is well known that, on occasions, tumours have been found lying solely within the tympanum. Lundgren (1949) suggested that these should be called tympanic body tumours. Henson, Crawford and Cavanagh (1953) reviewing glomus jugulare tumours from the neurological aspect also agreed with this proposal, as they pointed out that the initial presenting symptoms of tumours arising from the tympanic body are otological. We disagree with this type of anatomical differentiation of origin of these tumours in the petrous temporal region, as it is essentially of hypothetical academic value only and as it tends to obscure the basic clinical picture. After all, as Henson, Crawford and Cavanagh (1953) have also pointed out, the initial symptoms when the site of origin is the glomus jugulare are also aural.

It is nowadays fashionable to use terms for these tumours based on histopathological and functional aspects. Thus the name non-chromaffin paraganglioma of the middle ear was used by Lattes and Waltner (1949) to describe these tumours. As Rosenwasser (1968) rightly points out there is still no definite evidence that these structures possess paraganglionic function or any other known function.

Another term used is that of chemodectoma, originally proposed by Mulligan (1950) who assumed that these tumours were derived from chemoreceptor tissue. A leading article in the British Medical Journal (1964) was also in favour of the generic term "chemodectoma" for these and similar non-chromaffin neuroepithelial cells arising in the adventitia of blood vessels, carotid and the aortic bodies.

The tumours, histologically, consist of clumps of large polyhedral epithelial cells. The stroma is vascular (hence the occasional confusion with angiosarcomata), and contains non-myelinated nerve fibrils and uniform nuclei. These growths rarely metastasise. It is surprising that these tumours with their usual involvement of the jugular venous system do not metastasise more frequenty. The rare reports of metastases show spread to regional lymph glands, liver, lungs, spleen and vertebrae (Taylor, Alford and Greenberg, 1965).

Clinical Features. The main presenting symptoms may be either otological or neurological or a mixture of both. This depends on the exact site of origin of the tumour and also on the direction in which it invades surrounding structures. The broad clinical classification proposed by Black (1964) is excellent. In his Group I, there are only aural symptoms. This is the commonest group. In Group II, neurological symptoms and signs develop after the aural ones. Group III patients show from the beginning a mixture of aural and neurological involvement, while in Group IV, there is neurological involvement first and this is followed later by aural signs.

The most frequent presenting symptoms are those of tinnitus associated with mild loss of hearing on the affected side. The loss of hearing is usually insidious and slowly progressive. It must be noted that sudden loss of hearing may occur. Munson *et al.*, (1967) reported sudden loss of hearing in three of the patients they reviewed. The loss of hearing is of the conductive type as measured audiometrically and can show from 20 to 60 decibel loss. Other otological symptoms that may be complained of, are haemorrhage from the ear, otorrhoea, and otalgia. Otoscopy may reveal a vascular pulsating polyp either behind the ear drum or extending through it. The pulsations can be controlled by digital pressure over the carotid artery. The majority of reports in the literature deal with the otological symptomatology and signs.

Neurological manifestations though important develop, as a rule, after the start of otological symptoms. The development of a sudden lower motor neurone facial nerve palsy without any initial tinnitus, as occurred in our first patient is unusual, as it is customary for tinnitus to precede the neurological deficits. Rosenwasser (1968) in his paper on the therapy of these tumours describes a case (Case 9) which presented with middle ear stuffiness and mild conduc-

tion deafness in the absence of tinnitus. The proportion of cases showing neurological involvement has been variously reported as 40% (Henson, Crawford, and Cavanagh, 1953) and 47% (Munson *et al.* 1967). These two groups of workers analysed 149 cases between them. The neurological picture may either be the less common one of an intracranial space occupying lesion or else that of multiple cranial nerve palsies. The seventh to twelfth nerves are usually involved. If one groups the frequency of cranial nerve palsies analysed by the two above mentioned group of workers, one finds that involvement of the Vth nerve occurred in 10 patients; VIth nerve palsy in 9; Xth nerve palsy in 37 patients; XIth nerve involvement in 31 patients and hypoglossal nerve involvement in 45 patients. The frequency of involvement of the hypologlossal nerve as compared with other cranial nerves is worth underlining. Another frequent finding is nystagmus. Occasional findings are cerebellar or pyramidal signs. Henson *et al.* could find no reported cases of papilloedema (1953). Though this is unusual it does occur, as is demonstrated in the interesting case recorded by Taylor *et al.* (1965) where hydrocephalus and an increased intracranial pressure of 600 mm. CSF was found.

Another peculiar feature of these tumours is their increased incidence in females (Steinberg & Holz, 1965; Munson *et al.*, 1967). Thus Snyder and Maur reported that 70% of their patients were females whereas in the big series analysed by Oberman *et al.*, over 82% of their patients were females. This is in marked contrast to the incidence of chemodectomas elsewhere in the body where the majority of these tumours occur in males. Another odd feature is their greater tendency to occur on the left than on the right side. The tumours of both our patients were on the left side.

The natural history of these tumours is as a rule long. The average development of symptoms in the group of patients analysed by Munson *et al.*, (1967) was 5.5 years. Black (1952) stated that survival for twenty years was not uncommon while Bickerstaff and Howell's patient had remained untreated for 42 years. Steinberg and Holz (1965) reported a case where the growth had been present for almost fifty years before medical advice was requested.

Investigations which are usually performed include biopsy of the aural polyp. In cases where intratympanic location of the tumour is suspected, the technique of clinical examination should include the use of Siegle's speculum (Black, 1964) The inward pressure of the tympanic membrane may delineate the tumour. As the pressure is slowy decreased or increased, a stage is reached at which one may note pulsation of the vascular tumour. Skull radiography is unlikely to be of value in the early stages though erosion of the petrous bone and enlargement of the jugular foremen may be found in more advanced cases. Our first case showed marked erosion of the base of the skull. X-Ray of the mastoid bone may show "cloudliness". At times, tomography may reveal an erosive lesion where conventional films are negative. This is particularly important with regard to the jugular foramen. Newer techniques which have been advocated include the use of retrograde jugulargraphy with contrast media. Some workers feel that this is the only certain means of demonstrating intravascular growth into the jugular bulb and into the internal jugular vein. Others emphasise (Rosenwasser) that negative retrograde jugulargraphy does not rule out the presence of a tumour.

Internal carotid and vertebral arteriograms have been performed on these patients usually with negative results. This is not surprising as the blood supply of these tumours is from the external carotid artery, and Alexander, Beamer, and Williams (1951) were able to demonstrate the tumour in their patient in this way. External carotid arteriography could be done either by direct puncture or by selective catheterisation using the Seldinger technique, via the common carotid artery. Another recent method of radiological investigation is by using the subtraction Zeides des Plantes technique in rapid carotid arteriography.

The symptoms caused by these tumours have been mistaken for those arising from chronic otitis media, chronic secre-

tory otitis media, acute suppurative otitis media and granulomata of the external auditory canal. In the age groups (20 — 50 yrs. especially females), otosclerosis is also not a rare misdiagnosis. Glomus tumours have also been mistaken for carcinomata of the middle ear and nasopharynx. Other tumours that must be considered in the differential diagnosis are haemangiomas, meningiomas and neurilemmomas. When neurological deficits predominate, these may mimic those of primary tumours of the posterior fossa or of the base of the skull.

Treatment is either surgical or by radiotherapeutic means or by a combination of boht, radiotherapy preceeding surgery to reduce the vascularity of the growth. Small intratympanic tumours can be excised using the transmeatal approach, thus preserving the normal anatomy of the middle ear. With larger tumours a radical mastoidectomy may be required for their removal. For the tumours extending intracranially, posterior fossa craniotomy has been at times advocated. The procedure is as a rule very dangerous. This type of tumour is perhaps best treated with deep radiotherapy. The amount of radiotherapy necessary has been variously described from series to series. It is nowadays customary to give maximum supervoltage radiotherapy. Another method of treatment for small tumours that has been reported in the last two years is cryosurgery. The main advantage of this form of treatment is the reduction in bleeding and therefore it may prove to be a useful adjunct.

References

ALEXANDER, E., BEAMER, P.R., WILLIAMS, J. (1951) J. Neurosurg., *8*, 515.

BICKERSTAFF, E.R., HOWELL, J. (1963). Brain *76*, 576.

BLACK, J.I.M. (1952). J. Laryng., *66*, 315.

BLACK, J.I.M. (1964). Proc. R.S.M., 57, 943.

GEJTROT, T. & LAUREN, T. Acta Oto-laryng., *57*, 1

GUILD, S.R. (1941) Anat. Record. (Suppl. 2) *79*, 28.

GUILD, S.R. (1951) Arch. Otolaryng., *88*, 32.

HENSON, R.A., CRAWFORD, J.V., CAVANAGH, J.B. (1953) J. Neurol., Neurosurg. Psychiat., *16*, 127.

LATTES, R. & WALTNER, J.C. (1949). Cancer, *2*, 447.

LEADING ARTICLE (1964), Brit. Med. J., *1*, 792.

LUNDGREN, N. (1949). Acta oto-laryng., Stockh., *37*, 367.

MULLIGAN, R.M. (1950). Amer. J. Path., abstract, *28*, 680.

MUNSON, F.A., BROWN, H.A., HARNSON, E.G., JR., SIEKERT, R.G. (1967). Laryngoscope, *77*, 218.

ROSENWASSER, H. (1968) Arch. Otolaryng., *88*, 6.

ROSENWASSER, H. (1945). Arch. Otolaryng., *41*, 64.

SHAPIRO, M.J. & NEUES, D.K. (1964). Arch. Otolaryng., *79*, 219.

SNYDER, G.G., & MAUR, B. (1961). Arch. Otolaryng., SA, 54.

STEINBERG, N., E HOLZ, W.G. (1965). Arch. Otolaryng., *82*, 387.

TAYLOR, D.M., ALFORD, B.R., GREENBERG, S.D. (1965) Arch. Otolaryng., *82*, 5.

ZAK, F.G. (1954). New York J. Med., *54*, 8