A case of glomus jugulare tumour

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GUILD (1941, 1951) DESCRIBED the glomus jugulare as consisting of one or more bodies lying in the adventitia of the dome of the jugular bulb. Since then, it is known that the glomus jugulare is part of the chemoreceptor system, the carotid body being the best known example. The glomus jugulare has a histology which resembles the carotid body. It consists of capillaries separated by a delicate stroma containing numerous epitheloid cells rich in cytoplasm.

Rosenwasser (1945) reported the first tumour arising from this structure when he suggested that "carotid body tumour of the middle ear and the mastoid" might originate in the glomus jugulare. Such tumours are composed of cells in alveolar arrangement with numerous sinusoidal vessels (Latles and Waltner, 1949). Situated in the jugular foramen, they produce symptoms by eroding into the middle ear, or intracranium, especially the posterior cranial fossa causing palsies of the related lower cranial nerves.

The purpose of describing this patient is to discuss the possible diagnosis and review the more common presentations.

P.C.M. U.H. R.N. 080769, a 39-year-old Chinese married woman, when seen on 9th July 1969, first noticed a change in her voice, which became softer and hoarse about a year ago. She also experienced progressive dysphagia in which both liquid and solid food appeared to be stuck at her throat. Together with these symptoms, she had persistent tinnitus and increasing deafness of her right ear. She also complained of blurring of vision, with diplopia on seeing distant objects. These symptoms appeared to have followed one another fairly quickly. She became definitely aware that a lump on the right side of the neck seemed to be growing in size over a period of a month,

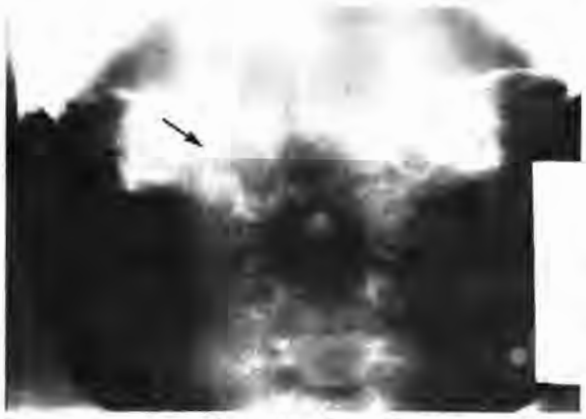

Fig. 1 Tomogram of Jugular Bulb
Arrow indicates site of erosion of Right Jugular Bulb

prior to her being seen in this hospital. On direct questioning, she revealed she had aches on that side of the neck, for years, and this was sometimes initiated by neck movements. She had also noticed a small lump in that region.

For these symptoms, she was admitted to the surgical unit of another hospital on three separate occasions for investigations of a possible naso-pharyngeal carcinoma within the last year. A laryngoscopy revealed a paralysis of the right vocal cord. Biopsy of the post cricoid region showed evidence of slight mild chronic inflammation only. Other examinations, like oesophagoscopy, barium swallow and bronchoscopy, provided no additional information.

When she was first examined, she had multiple lower motor neurone cranial nerve palsy of the right 5th, 6th, 8th, 9th, 10th, 11th and 12th cranial nerves. No papilloedema was seen. A swelling was found on the right side of the neck extending from almost the right angle of the jaw to just across the midline posteriorly. It was pulsatile, which ceased immediately on pressure over the right carotid artery. No bruit was heard over it or the right carotid artery. The right ear drum was thickened and bulging, suggesting presence of a tumour. The rest of the examination provided no signficant findings.

Investigations revealed a haemoglobin of 14 gm%, P.C.V. of 41%, total white cell count of 8,500 cells/cu.mm. with a normal differential count. The E.S.R. was 9 mm/hr. C.S.F. pressure was normal with a normal Queckenstedt test. There were no cells, protein was 22 gm% and sugar was 60 mgs.%. Blood and C.S.F. Kahn tests were negative. Chest X-rays and plain X-ray of nasal sinuses showed no abnormality.

Ordinary X-ray of the skull showed no abnormality. Special views of the jugular foramen with tomography showed a destruction of the base of the skull on the right side (Fig. 1), beginning from just behind and below the tip of the petrous pyramid. The right jugular fossa had lost its outline, being partially eroded and extended. The destructive process extended backwards to involve the posterior cranial fossa behind the region of the posterior tip of the foramen magnum.

X-rays of the mastoids showed that there was bony destruction at the medial and inferior aspects of the right petrous pyramid. The tip itself appeared to have been partially destroyed. A right carotid angiogram, with the needle placed into the proximal portion of the external carotid artery, showed the occipital and maxillary branch both hypertrophied, the former much more than the latter. An abnormal vascular pattern was seen in the right sub-occipital region resembling an A-V malformation (Fig. 2). A large draining vein at the posterior region was seen in sequential films. It revealed a tumour pattern of 10 cm. by 6 cm. extending from the medial margin of the base of the skull. The radiological conclusion appeared to be consistent with a glomus jugulare tumour. Other tumours that might show the vascular pattern included an A-V malformation or a very vascular secondary deposit. A diagnosis of glomus jugulare tumour was made. No biopsy was performed because of the danger of bleeding after even a relatively minor operation.

As the tumour was very extensive and surgical removal not possible, she was given deep X-ray therapy. No apparent change in the size of the tumour was noted during the course of radiotherapy. When reviewed one month later, there was no progression in the neurological symptoms. When last seen by one of us (T.P.D.) on 5th October, 1971, she was feeling well and had returned to her work as a rubber tapper. Her voice was no longer hoarse. She had no tinnitus but there was slight deafness of her right ear. There was no dysphagia, though she had to chew her food slowly before swallowing. She had no complaints regarding her vision.

On examination she appeared in good health. There was no lump in the neck. The only cranial nerve lesions still present were right VIII (slight hearing defect), IX and X (minimal loss of elevation of soft palate), XI (slight weakness of sternomastoid and trapezius) and XII (mild wasting of the right half of tongue and inability to protrude tongue out of mouth). The ear drum was normal and so were the fundi. There were no

Fig. 2a. Right carotid Angiogram Arrow shows early arterial phase of tumour.

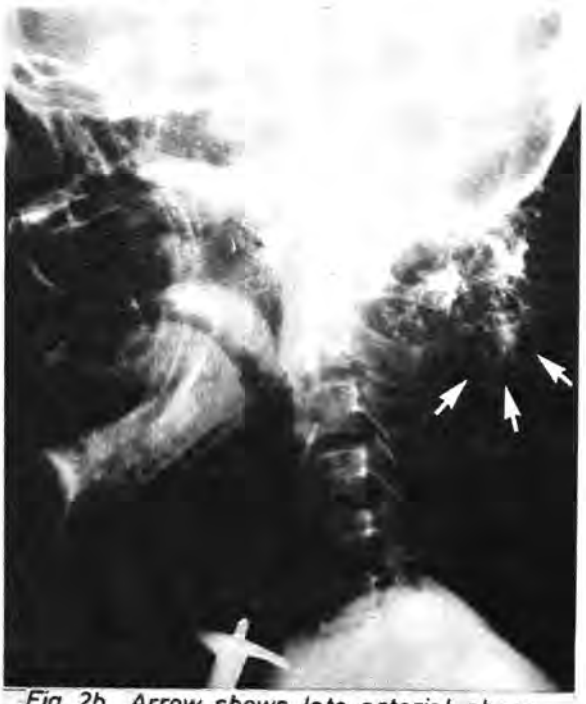

Fig. 2b. Arrow shows late arterial phase of tumour.

enlarged lymph nodes. The rest of the examination was essentially negative.

Discussion

In the excellent reviews by Bickerstaff and Howell (1953), Hanson, Crawford and Cavanagh (1953), it is noted that the tumour is four to five times commoner in women than in men. The long history of symptoms was stressed. About half of the patients had symptoms for more than ten years. The presentations fall into two main groups. The majority of patients reviewed present with aural symptoms, consisting of tinnitus and progressive deafness. Otorrhoea and vertigo are less frequent. Pain in the ear is not common. A striking feature is aural bleeding which sometimes occurs after coughing, sneezing or straining or following interference. Frequently, a red bulging drum is noticed and myringotomy invariably resulted in profuse bleeding. Also a polyp may be seen in the external auditory meatus. Sometimes this follows a myringotomy which permits the tumour to bulge through the drum or it erodes spontaneously through it. The only neurological involvement at this stage may be only a facial nerve palsy.

The other group of symptoms consists of neurological involvement which usually follow several years later. Dysphagia and hoarseness usually make the patients seek medical aid. The ninth, tenth, eleventh and twelfth cranial nerves are most frequently affected; much less common, the sixth and fifth cranial nerves may be involved.

Sometimes, neurological and aural symptoms appear together. Much less commonly, neurological symptoms precede aural manifestations (Revilla, 1948; Capps, 1952). Intracranial spread, usually into the posterior cranial fossa, is indicated by ataxia, incoordination and nystagmus. Sometimes the pyramidal tract may be involved. Raised intracranial pressure may be associated. Intracranial bruit and pulsatile mass below the mastoid and behind the ear are seen in some patients.

In the patient reported here, both aural and neurological symptoms appeared more or less simultaneously. There was a pulsatile lump on the right side of the neck. No aural polyp, haemorrhage or discharge were present. In the second case of Poppen and Riemenscheider (1957), there was no haemorrhagic manifestations. A pulsating tumour was found anterior to the mastoid. In Revilla's patient, bleeding occurred only after exploration of the cerebello-pontine angle. Two patients of Bartels (1949) had no aural bleeding, but both had aural symptoms only.

The possible differential diagnosis consists of

an angiomatous meningioma, an acoustic neuroma, a nasopharyngeal carcinoma. In the early stages, it may be difficult to differentiate them from a tumour jugulare. In most cases, the lower cranial nerve palsies with external aural symptoms exclude other possibilities. Nasopharyngeal carcinoma is common among Chinese. In this Chinese patient, it could have been difficult to decide but the lack of positive findings in the pharynx and post-nasal space, along with a pulsatile tumour, make such a diagnosis extremely unlikely. Besides, the lower cranial nerves are usually involved late in nasopharyngeal carcinoma and headache becomes very prominent by then. This latter symptom was conspicuously absent in this patient. Other diagnosis, like a racemose angioma, may present a problem (Bicksrtaff and Howell).

With better awareness and more refined investigations, glomus jugulare tumours are being diagnosed at an early stage. Special tomography has been used by Kim, S.K. and Cop (1966) and Jane Metz Strickler (1966) to display the jugular foramen. Kohut and Lindsay (1965), in an effort to determine the suitability for surgical removal and to determine tumour of middle ear origin from that which arises from the jugular fossa, employed subtraction angiographic techniques with tomography to great advantage. Gejrot and Lauren (1964) stressed the importance of retrograde jugularography preoperatively in detecting the presence of an intravascular involvement by the tumour of the jugular bulb and the jugular vein.

The treatment in early cases, if possible, is surgical excision (Kim, S.K. et al, McSwanz, Calson, Scott, 1959). Irradiation is indicated when extensive bone resection would be necessary to remove all the tumour. In a review of 38 cases of glomus jugulare tumour concerning the current management, Rosenwasser (1967) divided the cases into three groups. Group I cases are those with intact drum membrane and a small lesion apparently confined to the middle ear space; there is a moderate conduction type of hearing loss. In Group II cases, the middle ear, aditus, antrum and mastoid bone are involved. Group III cases have widespread extension, at times intracranially. In Group I cases, transtympanic removal is advised. Exploratory endaural mastoidectomy is always done in Group II cases. Irradiation is the treatment of choice for Group III cases. The patient reported here obviously belongs to Group III and radiotherapy was selected. She remains well at the end of two years. Encouraging results have been reported for radiotherapy (Alexander et al, 1951 and Copps, 1951).

Summary

A case of glomus jugulare tumour treated by deep X-ray therapy is reported with the patient remaining well at the end of two years. The symptomatology and literature are briefly reviewed.

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