A CASE OF GLOMUS JUGULARE TUMOUR WITH UNUSUAL NEUROLOGICAL COMPLICATIONS

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Glomus jugulare tumours, originating either in the middle ear or primarily as intracranial tumours,[4] with extension and involvement of multiple cranial nerves, are not rare. Involvement of CNS tracts is uncommon. The case presented here is an example of a proven case of glomus jugulare tumour in which numerous attacks of left hemiplegia and one attack of right hemiplegia occurred, all with recovery. A still more unusual feature was transient, severe, involvement of respiratory function.

In the territory of the carotid blood flow, glomus bodies[1] are normally present as minute structures of 1·5 mm. average diameter.[2] They consist of well-circumscribed areas of epitheloid cells surrounded by a highly vascular stroma, lying in the adventitia of the jugular bulb or along the course of the nerve of Jacobson and the nerve of Arnold.[2] They may occur at other sites in and about the region of the petrous temporal bone.[2] Their function is unknown, but they may play some part in the chemo-receptor mechanism of circulatory control.[5] Histologically, glomus bodies closely resemble the carotid and aortic arch bodies, and since they do not stain with chrome stains, they have been generically labelled 'nonchromaffin paraganglia'.[10, 14] They may become tumorous at any site at which they usually occur, the commoner sites being the middle ear and in close association with the jugular bulb. Variation in the clinical presentation of these tumours is attributed to the difference in anatomical sites at which glomus bodies exist. Their diagnosis is presumptive on clinical and radiological grounds and is confirmed on biopsy.

CASE REPORT

A 47-year old European female was admitted to the Johannesburg General Hospital on 21 September 1957 on account of a sudden 'collapse' followed immediately by a left-sided hemiplegia.

Ten years before this admission she began to complain of right-sided earache and 'whistling' of the voice. The pain was lancinating in nature and radiated to the right eye. She was seen by a surgeon who told her that she had a 'blood bag' in the right ear. Removal of this 'bag' was attempted on three separate occasions in 1947, 1948 and 1949. Each operation was accompanied by profuse haemorrhage from the ear, necessitating blood transfusion on each occasion. The earache persisted unchanged.

About 3 months after the third operation, in 1949, she collapsed after getting off a tram. She was 3½ months pregnant at the time, and was admitted to the Johannesburg General Hospital. She regained consciousness after 10 hours. A diagnosis of left basal pneumonia was made. During convalescence she developed epileptiform seizures, which, she was told, were grand mal epilepsy. She cannot remember how long these continued, and was allowed to go home, where she stayed in bed until the end of her pregnancy. A normal child was delivered when she was 40 years old. She had had rheumatic fever and chorea at the ages of 13 and 20 years respectively, and during the last trimesters of each of her 6 pregnancies she complained of some degree of dyspnoea on effort, palpitations and oedema of the ankles. She was told that she had rheumatic heart disease, and accepted the advice to be sterilized after the birth of her last child in 1949.

A month after delivery she was sterilized. On the fourth post-operative day she suddenly became 'paralysed' in all four limbs, but she cannot recollect any sphincteric or sensory abnormality. She was discharged in this state and remained in bed at home for 9 months. Recovery was gradual but complete.

In 1951, due to the persistence of the original earache, she consulted an otorhinolaryngologist, who advised mastoidectomy The surgeon described the findings of a highly vascular polyp in the middle ear on the right side. Attempted removal of the polyp was met by profuse haemorrhage which persisted for 2 days. For the 4th time she was given a blood transfusion. Biopsy of the growth was reported on histologically as being an 'haemangio-endothelioma'. Immediately after the operation she noticed complete deafness in the right ear and loss of use of the facial muscles on the same side. During the intervening years, between 1951 and 1956, she regained partial use of the muscles of the face, but remained completely deaf. During this same period, she complained of attacks of dizziness and 'black-outs'. She noticed these particularly whenever she was excited, breathless and on bending forward. It was the latter observation that prevented her from carrying out the household chores, for, although fully ambulant, and with no notable degree of cardiac disability, she was afraid to stoop forward for fear of blacking out.

In August 1956 she developed a sudden left-sided hemiplegia and was bedridden for a month. Recovery was spontaneous and complete. Again in August 1957 there was a recurrence of the hemiplegia, lasting only four days this time and leaving no residua.

On the day of the present admission (21 September 1957) while climbing some stairs. she suddenly experienced dizziness, palpitations and flushing of the face. She 'went lame' down the left side of the body, experienced momentary lancinating pain down the left side of the body and the right side of the face and for some minutes was unable to speak. She did not lose consciousness. On examination there was a flaccid paralysis of the left side of the body with hoarseness of the voice. Palsies of the 5th, 6th, 7th, 8th, 10th, 11th and 12th cranial nerves on the right were noted. There was wasting of the sternomastoid and trapezius muscles on the right side, and hemiatrophy of the right side of the tongue. The right ear drum was replaced by red granular tissue. Abdominal reflexes were absent on both sides and both plantar responses were flexor. The heart was slightly enlarged, in sinus rhythm, and displayed signs of a mitral stenosis. There was no evidence of congestive cardiac failure. The blood pressure was 145/110 mm. Hg. The following day the 6th-nerve paresis returned to normal.

During the next 2 weeks there was considerable improvement in the power of the left leg and arm. She however, began to complain of increasing difficulty in swallowing and was unable to protrude the tongue out beyond the teeth. On the 24th day after admission, when the weakness of the left arm and leg were much improved, she developed a paralysis on the right side of the body, the right arm being more affected than the leg. At about this time she had begun complaining of an oppressive feeling in the chest and difficulty with breathing, most notable during the night. There were no signs or accompaniments of congestive cardiac failure and her symptoms were considered clinically to be due to some form of central respiratory involvement. This view seemed to be justified clinically, since she gained immense relief from the use of a cuirasse respirator.

Special investigations, including full blood counts, serum electrolytes and blood urea estimations and radiology of the chest were all normal, except for a doubtful increase in the protein content of the cerebrospinal fluid (45 mg. %). However, X-rays of the skull were most informative, and a copy of Dr. R. Glyn Thomas' report is included:

> 'There is destruction of the right jugular foramen, extending anteriorly towards the temporomandibular joint, and posteriorly towards the condylar fossa. The right petrous bone tip (Fig. 1) is completely destroyed and there is irregular moth-eaten destruction, leaving small islands of bone intact, extending laterally into the middle ear region. Auditory ossicles are visible on the left but not on the right. Both vertebral artery foramina in the atlas are large. A very large mastoid emissary vein foramen is demonstrated on the Stenver's view of the right temporal bone. This suggests and extremely vascular lesion. A clear-cut area of operative removal of the temporal air cells is present behind the right acoustic meatus.

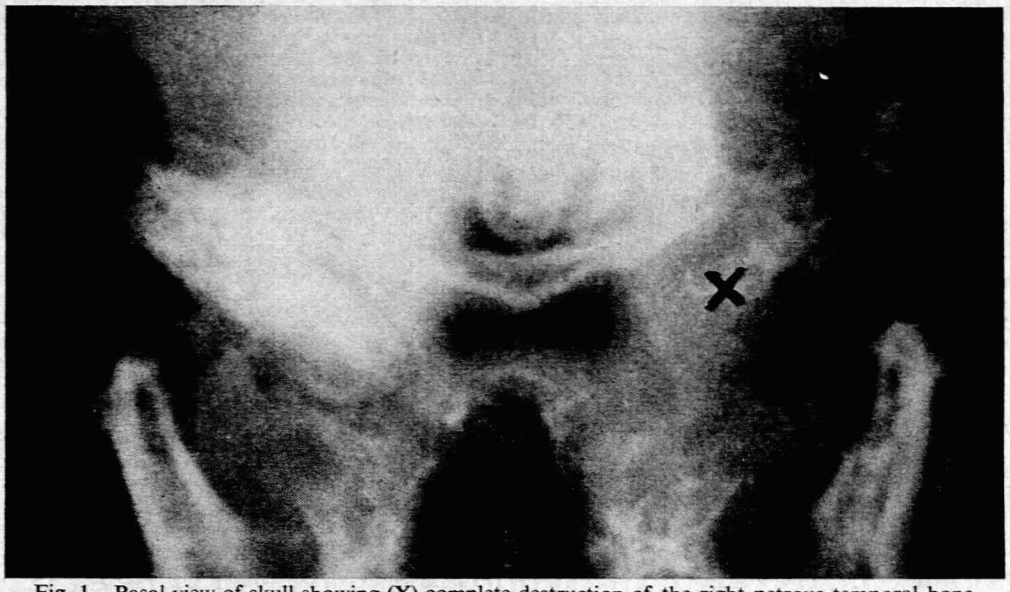

Fig. 1. Basal view of skull showing (X) complete destruction of the right petrous temporal bone. Compare the density of the left petrous temporal bone.

Interpretation: The radiological appearances are those of a glomus jugulare tumour arising in the right jugular fossa. There is, in addition, evidence of operative removal of temporal air cells. The following is the differential diagnosis:

1. Extension upwards to the base of the skull of a naso-pharyngeal tumour.
2. Metastatic tumour involvement.
3. Carcinoma of the right middle ear with extension chiefly medially. This seems unlikely.
4. Glomus jugulare tumour.

Conclusion: The appearances favour the diagnosis of a glomus jugulare tumour (of the right temporal bone) arising from the jugular fossa area'.

In view of the X-ray report and the clinical picture, a review of the biopsy slides of 1951 was requested and an ammended diagnosis of glomus jugulare tumour was made. On the evidence, a course of deep X-ray therapy was instituted. Under radiotherapy, the improvement previously noted, continued.

The use of a respirator was discontinued when it was found that she no longer required it. A review on 1 December showed that there was still present a gross weakness of the right arm with increased jerks, together with some residual weakness of the other limbs. The 7th, 10th, 11th and 12th cranial nerves remained paralysed, and the deafness was unchanged. Sensation of the tongue on the right side was found to be lost, and there was hyperaesthesia to all modalities of sensation on the right side of the body. Hyperventilation of 4 breaths at this stage produced a very notable feeling of intense dizziness and faintness, with a bradycardia.

The course of deep X-ray therapy was completed on 11 December, a total tumour dose of 3,800 r having been given. Enquiries revealed that in 1951 she had received a dose of 2,350 r privately. In view of this, further treatment was abandoned in favour of an expectant approach. In the course of the next week or two, the weakness of her limbs improved enough for her to get out of bed and walk. At the time of her discharge on 21 December, she was walking normally, the limbs were of normal power, and the residual neurological signs were confined only to the cranial nerves. The 7th, 8th, 9th, 10th, 11th and 12th nerves on the right remained paralysed. It required about 20 breaths to produce the hyperventilatory symptoms previously produced by 4.

DISCUSSION

The diagnosis of glomus jugulare tumour in this patient was confirmed by biopsy. Not infrequently a glomus jugulare tumour is histologically mistaken for a vascular endothelioma, as well as for a variety of other tumours,[6, 8-10] Poor site of section and imperfect technique in the preparation of the histological specimen are the main causes of this error.[9, 10] As the tumour was first described in 1945,[15] it is not unlikely that pathologists are still unfamiliar with its histological appearance.

Numerous cases of glomus jugulare tumour with middle ear symptoms and with or without cranial nerve palsies, have survived over 20 years. Survival for 10 years, as in the present case, is not unusual, provided the tumour does not spread towards the lower cranial nerves to involve the vital centres.[12] The 7th nerve paralysis and the deafness in this patient could well be accounted for by the local tumour, or ascribed to a post-operative complication of mastoidectomy.

The other neurological manifestations require an explanation. Extension of the tumour through the petrous temporal bone, to involve the lower cranial nerves, is not unusual.[8] A less common feature is the numerous attacks of functional loss of the pyramidal tract, involving mainly the left side in this case. All these attacks were severe and incapacitating, but transient. The involvement of respiratory function is also unusual, only one case being reported with a history of 'suffocation'[13] The difficulty with breathing appeared to be due to respiratory-centre involvement since it had none of the qualities of cardiac dyspnoea and was dramatically relieved by the respirator. The mechanism whereby the tract functions were affected temporarily, yet severely, remains speculative. A single glomus jugulare tumour, originating in the middle ear, eroding the petrous temporal bone, and eventually surrounding (and in places compressing) the upper medulla, could explain the clinical picture except for the transient nature of some of the symptoms. Changes in the vascularity of the tumour, which in themselves would be dependent upon carotid blood flow, could cause these transient features. Changes in the vascularity could be affected by the relative hypoxia of exercise in a person with a rheumatic heart, by changes in posture or pressure and by hyperventilation from any cause, thereby implicating chemo-receptor activity of the tumour.[5] In

this physiological response, glomus jugulare tumours appear to behave in the same way as carotid body tumours.

Opinions vary in regard to the radiosensitivity of the glomus jugulare tumours. Surgical intervention is fraught with the danger of massive haemorrhage, but seems to be the treatment of choice when the tumour is localized to the middle ear.[6] In any case there are good grounds for advising radiotherapy with or without surgical intervention.[3, 6, 7, 11] In the present case it is not possible to come to any conclusions in regard to the sensitivity of the tumour, since previous attacks had cleared up spontaneously and the present attack was improving at the time that deep X-ray therapy was started.

SUMMARY

A case of glomus jugulare tumour (with evidence of intracranial extension) is described, with some usual and some unusual neurological complications. The unusual complications were recurrent hemiplegias, involvement of the respiratory function and central nervous symptoms which could be readily produced on hyperventilation.

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REFERENCES

1. Guild, S. R. (1941): Anat. Rec., **7**, 29.
2. *Idem* (1953): Ann. Otol. (St. Louis), **52**, 1045.
3. Williams, H. L., Childs, D. S., Parkhill, E. M. and Pugh, D. G. (1955): *Ibid.*, **64**, 546.
4. Dockerty, M. B., Love, J. G. and Patton, M. M. (1951): Proc. Mayo Clin., **26**, 26.
5. Huppler, E. G., McBean, J. B. and Parkhill, E. M. (1955): *Ibid.*, **30**, 53.
6. Capps, F. C. W. (1957): J. Fac. Radiol. (Lond.), **8**, 312.
7. Williams, I. G. (1957): *Ibid.*, **8**, 335.
8. Beckerstaff, E. R. and Howell, J. S. (1953): Brain, **76**, 576.
9. Lattes, R. (1950): Cancer, **3**, 667.
10. Lattes, R. and Waltner, J. G. (1949): *Ibid.*, **2**, 447.
11. Henson, R. A., Crawford, J. V. and Cavanagh, J. B. (1953): J. Neurol. Psychiat., **16**, 127.
12. Efron, G. (1957): S. Afr. Med. J., **31**, 164.
13. Hooper, R. S. (1955): J. Fac. Radiol. (Lond.), **7**, 77.
14. Watzka, M. in Mollendorff, W. V., ed. (1943): *Handbuch der Mikroskopischen Anatomies des Menschen*, T. 4, p. 262. Berlin: Springer-Verlag.
15. Rosenwasser, H. (1945): Arch. Otolaryng. (Chicago), **41**, 64.