

Haemangiopericytoma mimicking Glomus Jugulare tumour

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Citation

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Abstract

Haemangiopericytoma of the Skull Base is a rare entity. There are six cases of Haemangiopericytoma of the skull base reported in Literature, four of which were associated with Collet-Sicard syndrome. This case was not a part of this syndrome. Haemangiopericytoma greatly mimics glomus jugulare tumour if it involves the jugular fossa.

INTRODUCTION

Clinicians are inclined towards the diagnosis of glomus jugulare tumour whenever a lesion occupies the jugular fossa region particularly if there is an element of bone destruction. However, not every lesion occupying the jugular fossa and destroying it should be considered as Glomus Jugulare tumour. Haemangiopericytoma ; Extramedullary Plasmacytoma and Giant cell tumours should be seriously considered.

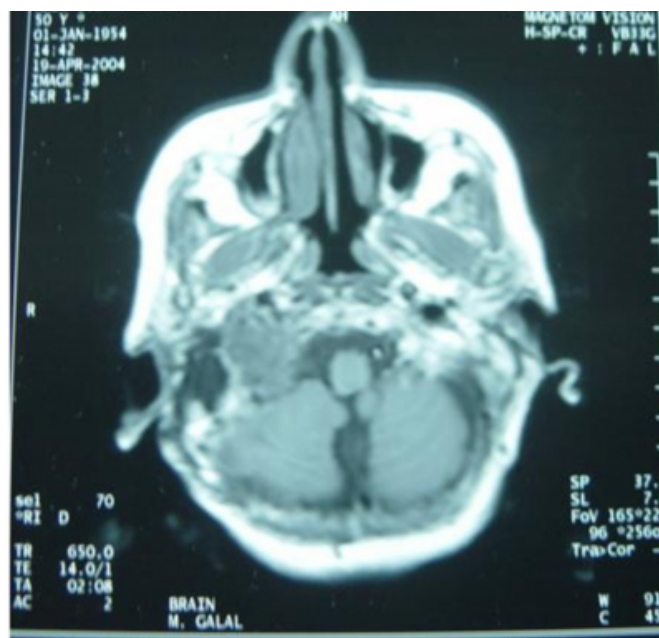
CLINICAL SCENARIO

A 50 year-old lady presented with right-sided pulsating tinnitus of one-year duration. The tinnitus was nearly constant all the time particularly noticeable at night. Three months after the onset of tinnitus, she started to notice deterioration of hearing of the ipsilateral side. Four months later, she noticed ipsilateral facial weakness.

When seen by myself, it was nearly 12 months after the onset of her tinnitus by which time she was totally deaf on the right side and has developed grade VI facial paralysis (House-Brackmann). The rest of cranial nerves and other neurological examinations showed no significant findings.

MRI temporal bone showed a 5x4x3 cm mass occupying the right jugular fossa and extending to the CPA (Figure 1).

Figure 1



Carotid Angiography was tried but the patient developed a serious reaction to the dye and it had to be abandoned. MRA has not been feasible.

Type A infratemporal fossa approach was undertaken to excise the mass. The majority of the tumour was taken out except the most medial part that is close to the brainstem (Figure 2). The tumour was found invading the lumen of the internal jugular vein down to the neck to the level of carotid bifurcation (figure 3).

Figure 2

Figure 2: postoperative T1 weighted MRI

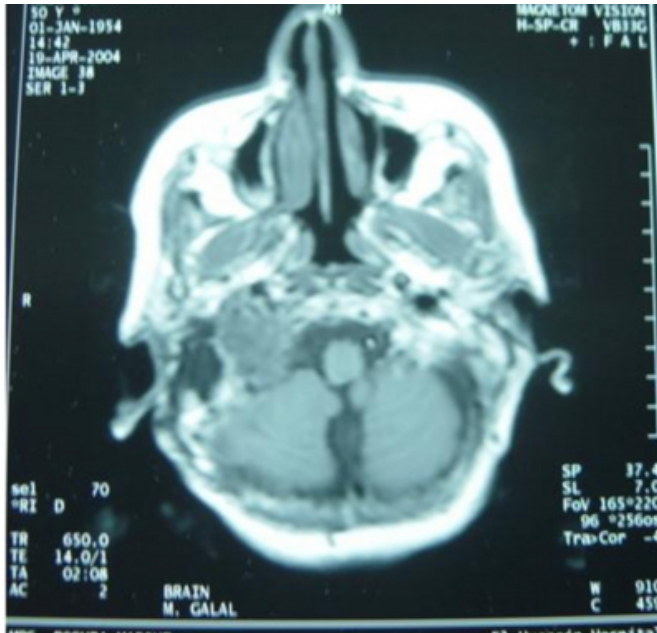
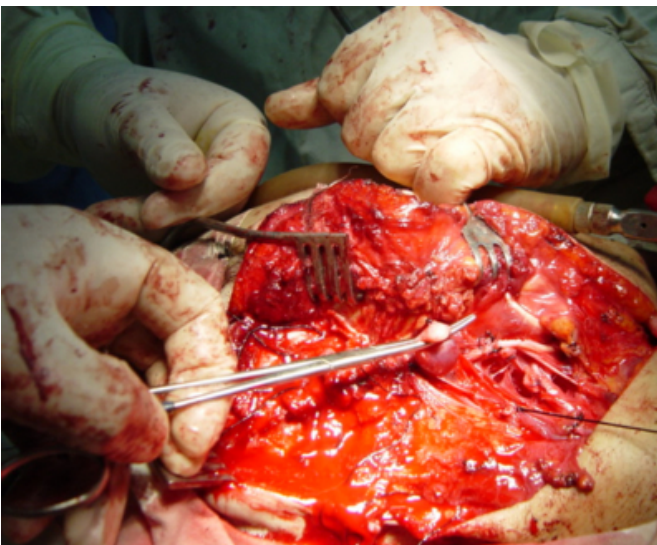


Figure 3

Figure 3: Intraop. Picture showing the tumour obliterating the lumen of IJV in the neck



DISCUSSION

Haemangiopericytoma is a malignant tumour arising from blood vessels (capillary pericyte of Zimmermann). The tumour appears in the form of capillaries surrounded by a connective tissue sheath. Outside this sheath are tumour cells that vary in appearance. It is very unpredictable i.e. non-mitotic tumours can metastasize. Also, distant metastasis is common but regional one has not been observed yet. Despite being pale and look avascular, these tumours bleed

vigorously when biopsied.

It has a single denominator: lack of uniformity in appearance and biological behavior. Distant metastasis has been reported but lymph node involvement hasn't been observed yet. Treatment is mainly surgical through local excision but recurrence is high.

There are six cases of Haemangiopericytoma of the skull base reported in the English Literature according to Medline search from 1966 to date (1,2,3,4,5).

Four out of the six cases were having Collet-Sicard syndrome.

This syndrome refers to having lower 4 cranial nerves palsy sparing the sympathetic chain. In the case of this study, it was only the facial nerve that was involved.

CONCLUSION

Lesions occupying the Jugular Fossa should not be taken for granted to mean Glomus Jugulare tumours. Other pathologies greatly resemble this and include Haemangiopericytoma, Extramedullary plasmacytoma and Giant cell tumours.

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