Case Report

Glomus Jugulare Tumor Presented as Aural Polyp With Intermittent Ear Discharge

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INTRODUCTION:

Glomus jugulare tumors (Paraganglioma) are very slowly growing tumors; arise from non-chromaffin paraganglionic tissue situated in close relationship to the middle ear. It may originate from Glomus bodies in the adventitia of the dome of the jugular bulb (Glomus Jugulare). The ganglionic tissue of glomus bodies is derived from the neural crest and is widely distributed in the autonomic nervous system. Guild (1953) discovered the presence of glomus ganglionic tissue within the temporal bone, in close relation to the jugular bulb in 50%, and on the mucosa of the promontory of the middle ear 25%. [1] Rosenwasser (1945) was the first surgeon to recognize the relationship between these tumors and the normal glomus jugulare. [2] These tumors have well defined thin fibrous capsule. Histological examination shows similar appearance to the normal glomus jugulare. The cells of the tumor usually have no endocrine function. They are not very active with only rare mitotic figures & usually of low malignancy, very rarely have nodal or distant metastasis. However, clinically they are invasive and destructive to the temporal bone and the facial nerve. They readily infiltrate the cellular system of the mastoid. Glomus jugulare shows predominance in females of 6: 1 and tends to be more common in the middle age group with familial and hereditary patterns [3]. Jugular bulb tumors may present a syndrome of paralysis of 7th, 9th, 10th and/or perhaps 12th cranial nerves, deafness, tinnitus and vertigo. [4]

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CASE REPORT:

On the 4th of March 2007, a 28 years old male patient brought to the casualty department with multiple clinical & neurological disorders; he was feverish & semiconscious, he had difficulty in swallowing & left facial nerve paralysis associated with swollen discharging left ear. He was suspected to have an intracranial complication due to chronic suppurative otitis media & was first referred to the neurological department, where he was treated conservatively as a case of meningitis. A brain CT scan was performed & was normal, then referred to our department to deal with his chronic ear disease. When we received him, he was looking ill, drowsy with multiple cranial nerves paralysis (7th, 9th, 10th and 12th) and otalgia.On ear examination, a large aural polyp was found, filling the left external auditory meatus and extruding outside the canal.

The history of his illness was dated back for more than two years, when he complained of repeated attacks of an ear discharge associated with deafness & tinnitus treated by his local doctors with antibiotics & other conservative measures which showed some response. On one of his visits to that clinic, he was found to have an aural polyp and advised to have it to be removed by surgery; he refused the advice & went home.

One year later, he visited his local ENT clinic complaining of tinnitus which was getting worse & recurrent bleeding from the affected ear. On examination, it was found that the polyp had increased in size filling the whole ear canal and preventing the introduction of the speculum of the aurioscope. He was also advised to have surgical intervention & he also refused that perhaps due to some personal or social problems. After six months, he was admitted urgently to the local Neurological Unit with a suspicion of meningitis on the basis of clinical findings of neck stiffness, fever and night sweating and he was treated by antibiotics. Apparently he responded well to that

conservative treatment and discharged home after one week. At home, his condition deteriorated and developed unsteadiness in addition to the symptoms mentioned above.

In our ward a new CT scan of the mastoid air cells and brain was taken which showed a picture of a large tumor infiltrating the mastoid process & obliterating the external ear canal. An MRI with venography was taken which showed narrowing & diminished blood flow of the lateral sinus & confirmed the intracranial extension of the tumor.

On microscopic examination under general anesthesia, the lesion within the external ear was examined properly, it was not a simple inflammatory polyp but it was a firm mass of tumor tissue obliterating the canal & preventing the introduction of the aural speculum. Biopsy was taken from the lesion which has caused severe bleeding controlled by firm BIPP pack inserted in the ear canal & a bandage applied over the head. Several small pieces were taken and sent for histopathological examination.

Histopathological result showed nests of uniform epithelial cells separated by network of compressed blood vessels. The picture goes with Paragangilioma (Glomua Jugulare). By this report the diagnosis of advanced glomus jugulare tumor of grade D was confirmed. Unfortunately, our surgical experience to excise such a large tumor is limited and it was decided to refer him for treatment by radiotherapy.

DISCUSSION:

Age & sex:

Glomus Jugulare tumour shows a predominance in females of 6: 1. Glomus tumors tend to be more common in the middle age groups [3]. Our patient was male and in his twenties which is rather uncommon incidence of this tumor.

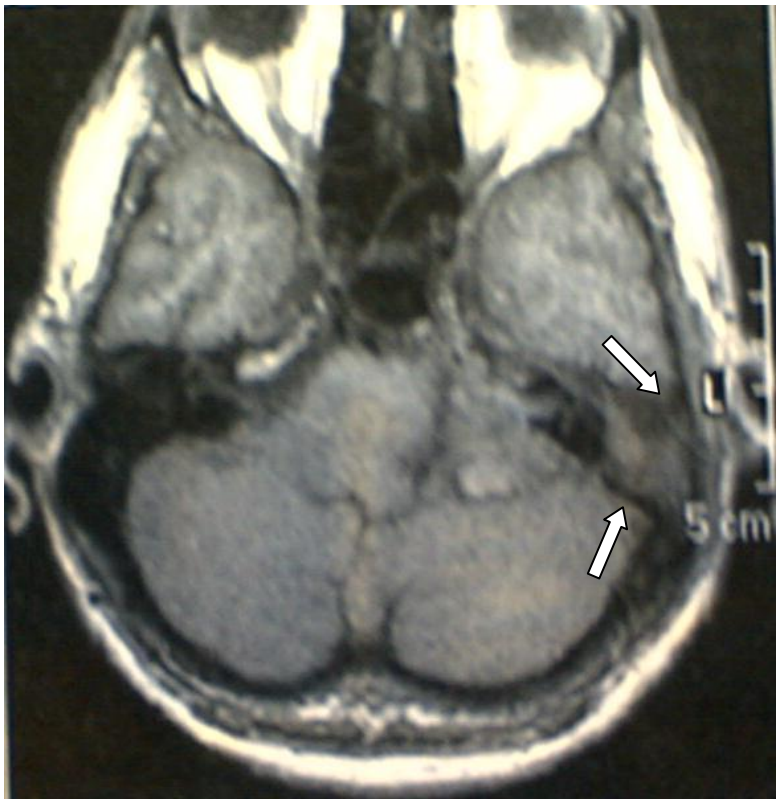
Presenting features:

Glomus jugulare is slowly growing tumor, its diagnosis is often missed until the tumor is very extensive. Alford & Guilford (1962) found the average delay is six years. [5] Symptoms generally follow the middle ear. Deafness & tinnitus presented in 75% of patients. Development of polyps arising from the wall of the external ear canal was unreported in the review of 61 case of glomus jugulare by Watkins et al (1994). [6]

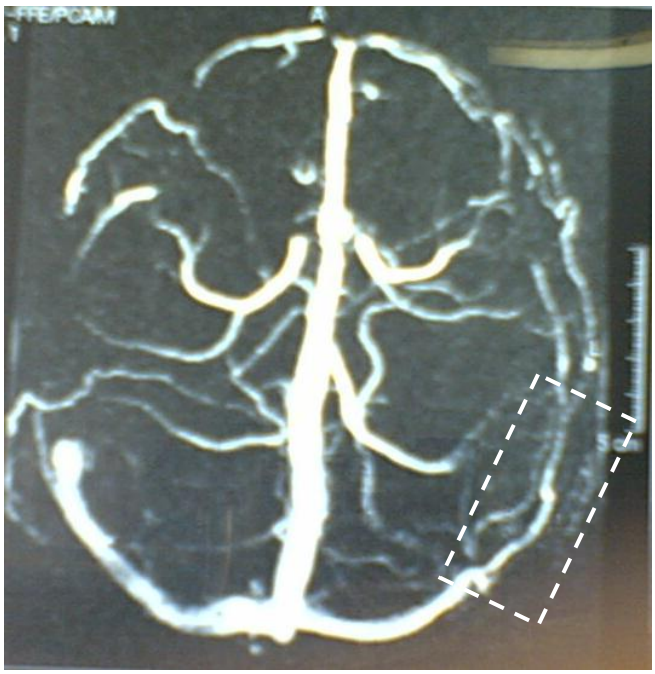
Our patient presented with deafness, tinnitus associated with ear discharge as usual. The development of an enlarging polyp in the external ear canal is rather different. The tumor has a rapid growth rate; within two to three years period, infiltrated the mastoid air cells and invaded the external auditory meatus. Also the quick deterioration in the clinical condition of the patient due to the intracranial extension may be another feature of its rapid growth which is rather unusual for glomus jugulare tumors in general. The associated infection of the middle ear has responded quickly to antibiotic administration perhaps due to the vascularity of the tumor which raises the antibiotic level at the site of infection.

Prognosis & outcome of glomus tumors:

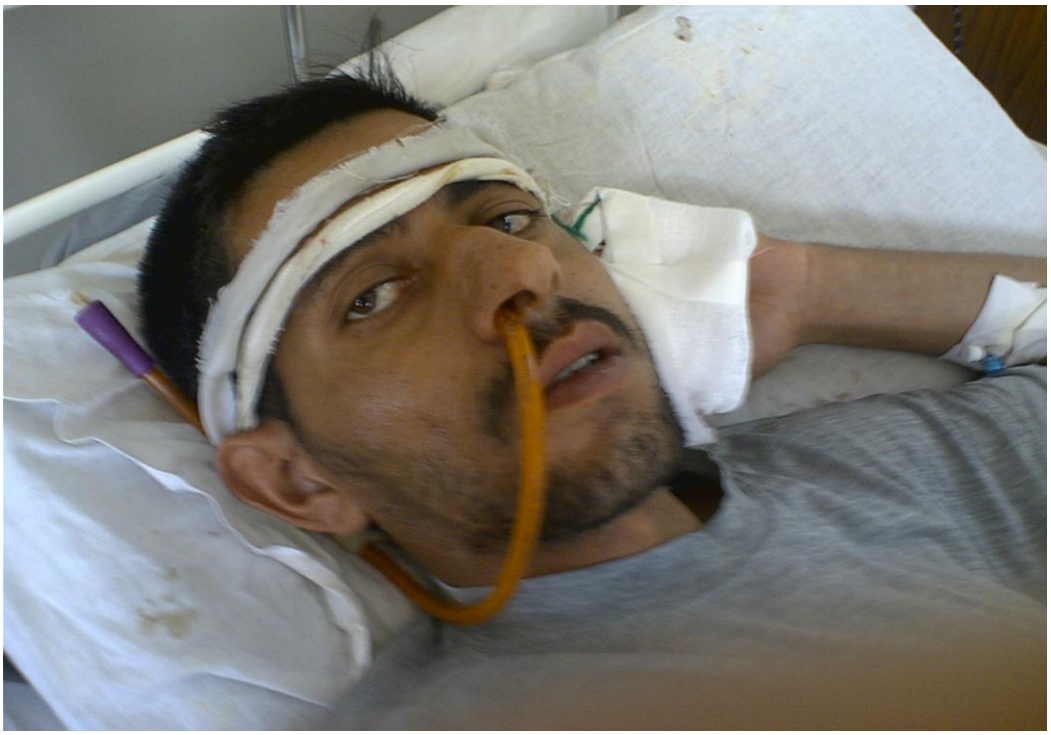
The prognosis of untreated patient with glomus jugulare tumor is serious and the tumor can be lethal especially in its late stages, and early diagnosis & treatment gives better prognosis. Therefore an increase in suspicion of the diagnosis by the doctor in charge and by ordering imaging investigations of the affected area is of great importance in order to receive appropriate management of this condition during its early stages.



(Fig. 1) :MRI (axial view) showing mass invading left E.A.C. (upper arrow) with intracranial extension (lower arrow).



(Fig.2):MRI venography shows compression and narrowing of left lateral sinus.



(Fig.3): Patient in the late stage of Glomus Jugulare tumor.

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