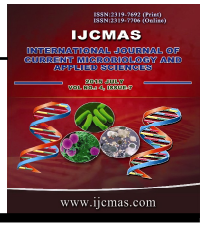
Int.J.Curr.Microbiol.App.Sci (2015) 4(7): 339-342

International Journal of Current Microbiology and Applied Sciences
ISSN: 2319-7706 **Volume 4 Number 7 (2015) pp. 339-342**
http://www.ijcmas.com



Case Study

Rapidly growing infiltrating *Glomus jugulare* tumor: An uncommon cause of bleeding aural polyp

Nadia Shirazi[1*] and SS Bist[2]

Department of Pathology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Jolly Grant, Dehradun, Uttarakhand, India
Department of ENT, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Jolly Grant, Dehradun, Uttarakhand, India
**Corresponding author*

Keywords

Glomus jugulare, Paraganglioma, Infiltrating

A B S T R A C T

Glomus jugulare tumors are rare slow growing tumors occurring within the jugular foramen. Although benign, these tumors are locally destructive because of their proximity to the petrous bone, lower cranial nerves and major vascular structures. We present the case of a 61 year old female presenting with sudden onset hearing loss and bleeding from ear.

Introduction

Glomus Jugulare Tumors (GJT) are rare vascular tumors included in the group of paragangliomas. They usually arise in the region of jugular bulb however can also be seen in a variety of sites like carotid body, glomus vagale and glomus tympanium. These hypervascular tumors are slow growing and arise within the jugular foramen of the temporal bone. GJTs have an annual incidence of 1 case per 1.3 million population (Forbes *et al.,* 2012). The tumors tend to be more common on the left side and the male:female ratio is 1:3–6. They are seen in the 5th to 6th decade of life and are rare in people younger than 18 years of age. Multicentricity is seen particularly in familial cases (Moffat and Hardy, 1989; Havekes *et al.,* 2008).

Case

A 61 year old hypertensive female presented to the ENT OPD with chief complains of ear fullness, otorrhea, hemorrhage and a left aural mass. There was no history of pain, facial palsy and hoarseness of voice or dysphagia. Otoscopic examination revealed a pulsatile reddish mass which appeared to be originating from the anterior part of auditory canal (Figure 1). CT scan showed an infiltrating hypervascularised tumor which was extending into the middle ear as well as inferiorly into the infratemporal fossa. Bony margins were eroded. A differential diagnosis of jugular paraganglioma or schwannoma were rendered radiologically. Patient was taken up for surgical excision.

Conservative jugulopetrosectomy was performed and mass was sent for histopathological examination (HPE).

On gross examination a reddish brown soft to firm tissue piece measuring 1.2 x 1 x 0.5 cm was received. Cut section showed a homogenous appearance. HPE described a benign tumor forming a "zell- ballen" pattern. Cells had round to mildly pleomorphic nuclei with granular salt and pepper type chromatin. Mitotic figures were sparse and necrosis was not seen. Tumor was involving the posterior surgical resected margin indicating an incomplete excision (Photomicrograph 1). Immunohistochemistry showed S-100 positivity in tumor cells. Diagnosis of glomus jugulare/ jugulotympanic paraganglioma was rendered. There were no post operative complications and the patient was sent for radiotherapy considering incomplete primary excision.

Discussion

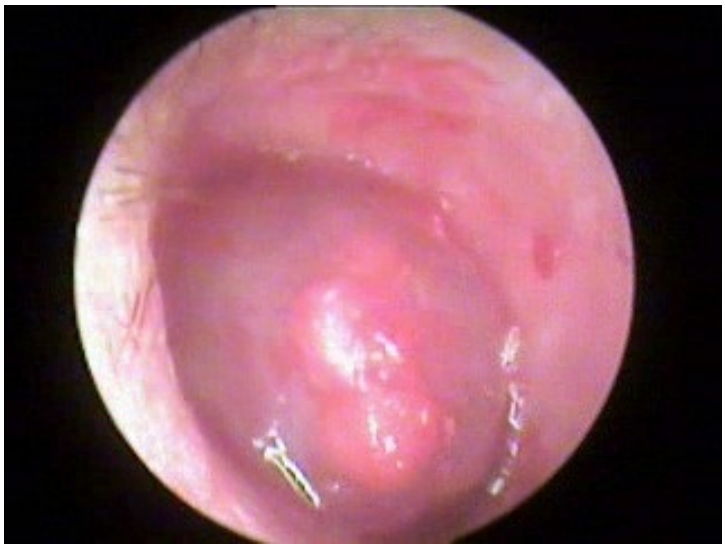
The GJT are small collections of paraganglionic tissue which are derived from embryonic neuroepithelium in close association with the autonomic nervous system and are found in the region of jugular bulb. Based on the presence of catecholamines and neuropeptides, paraganglia are included in the amine precursor uptake and decarboxylase (APUD) system, which has been referred to as diffuse neuroendocrine system (DNES). The first description of glomus tumors as hyperplastic glomus bodies was reported by Masson in 1924. These are rare tumors with benign characteristics and a slow growth rate of 1mm per year (Jansen *et al.,* 2000). The main blood supply is via the ascending pharyngeal artery from the external carotid artery and branches from the petrous portion of the internal carotid artery. The structures lying in close proximity to glomus jugulare are the jugular bulb including internal auditory canal, the posterior semicircular canal, the middle ear, medial external auditory canal, facial nerve (postero-laterally) and the internal carotid artery. On the basis of extension of tumor into the adjacent structures, Fisch classification of glomus tumors is widely used. The classification is detailed as follows:

Type A: limited to middle ear cleft (glomus tympanicus)
Type B: limited to tympanomastoid area with no infralabyrinthine compartment
Type C: Tumor involving infralabyrynthine compartment of the temporal bone and extending into the petrous apex
Type C1: Tumor with limited involvement of the vertical portion of the carotid canal
Type C2-Tumor invading the vertical portion of the carotid canal
Type C3:-Tumor invasion of horizontal portion of carotid canal
Type D1- Tumor with intracranial extension<2cm in diameter
Type D2-Tumor with intracranial extension>2cm in diameter

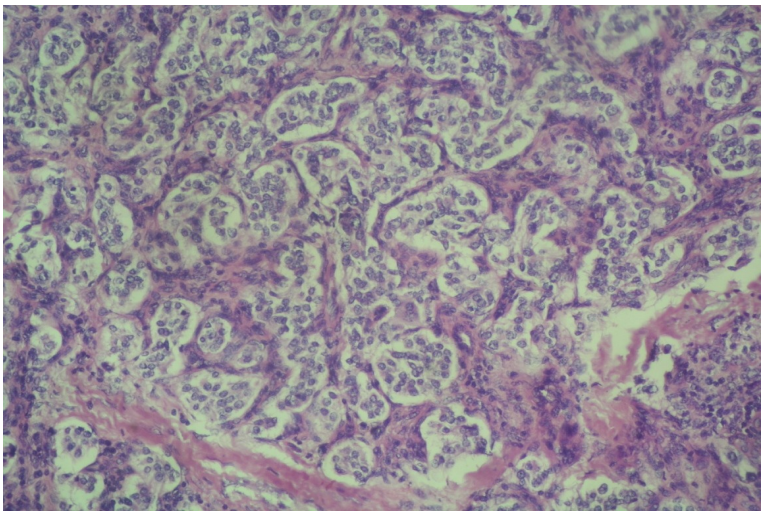
Classifying the tumor according to the degree of structures infiltrated closely relates to the mortality and morbidity associated with the tumor. Although no known risk factors have been recognized in GJT, a pathogenetic mutation in the gene for succinate dehydrogenase enzyme has been implicated in causing the disease (Pigny *et al.*, 2008).

Surgery (complete resection with conservation of cranial nerves) is the treatment of choice however other modalities of treatment are embolization, radiation, gamma knife radiosurgery, intratumoral injection of cyanoacrylate glue. Medical therapy may be indicated in tumors secreting catecholamines (α blockers, β blockers) (Sharma *et al.*, 2008).

Figure.1 Otoendoscopy showing a red vascular mass completely filling the Left external auditory canal



Photomicrograph.1 H&E: 200X: Zell ballen arrangement of glomus jugulare tumor



A study by Prabhu *et al.* (2004) showed that even complex glomus tumors can be managed surgically. Mortality rate is 6.2% among patients treated with radiation and 2.5% among those treated surgically. The overall mortality is 8.7% (Rosenwasser *et al.*, 1945).

Conclusion

Though the tumors are benign, they pose a significant diagnostic and management challenge because of the location and extent of involvement

References

Forbes, J.A., Brock, A.A., Ghiasi, M., Thompson, R.C., Haynes, D.S., Tsai, B.S. 2012. Jugulotympanic paragangliomas: 75 years of

evolution in understanding. *Neurosurg. Focus*, 33(2): E13

Havekes, B., van der Klaaw, A.A., Hoftijzer, H.C., Jansen, J.C., van der Mey, A.G., Vriends, A.H. 2008. Reduced quality of life in patients with head and neck paragangliomas. *Eur. J. Endocrinol.*, 158(2): 247–53.

Jansen, J.C., van den Berg, R., Kuiper, A., van der Mey, A.G., Zwinderman, A.H., Corne Lisse, C.J. 2000. Estimation of growth rate in patients with head and neck paragangliomas influences the treatment proposal. *Cancer*, 88: 2811–2816.

Masson, P. 1924. Le Glomus neuromyo-arterial des regions tactiles et ses tumours. *Lyon Chir.*, 16: 257–280.

Moffat, D.A., Hardy, D.G. 1989. Surgical management of large glomus jugulare tumors: infra- and trans-temporal approach. *J. Laryngol. Otol.*, 103(12): 1167–80.

Pigny, P., Vincent, A., Bouers, C.C. 2008. Pragangliomas after maternal transmission of a succinate dehydrogenase gene mutation. *J. Clin. Endocrinol. Metabol.*, 93(5): 1609–1615.

Prabhu, S.S., Demonte, F. 2004. Complete resection of a complex glomus jugulare tumor with extensive venous involvement. Case report. *Neurosurg. Focus*, 17(2): E12.

Rosenwasser, H.1945. Carotid body tumor of the middle ear and mastoid. *Arch. Otolaryngol.*, 41: 64–67.

Sharma, M.S., Gupta, A., Kale, S.S., Agrawal, D., Mahapatra, A.K., Sharma, B.S. 2008. Gamma knife radiosurgery for Glomus jugulare tumors: Therapeutic advantages of minimalism in the skull base. *Neurol. India*, 56(1): 57–61.