



**JUGULOTYMPANIC PARAGANGLIOMA PRESENTING AS EAC MASS, A CASE REPORT**

**Nitika Chawla<sup>1</sup>, Saral Ahuja<sup>2</sup>, Meenu Gilotra<sup>3</sup>, Kulwant Singh<sup>4</sup> and Swaran Kaur<sup>5</sup>**

<sup>1,4,5</sup>BPS Govt. Medical College (W), Khanpur Kalan, Sonipat, Haryana

<sup>2</sup>Consultant (Otorhinolaryngology), Rohtak City

<sup>3</sup>Demonstrator, BPS Govt. Medical College (W), Khanpur Kalan, Sonipat, Haryana

**ARTICLE INFO**

**Article History:**

Received 5<sup>th</sup> January, 2018

Received in revised form 20<sup>th</sup>

February, 2018 Accepted 8<sup>th</sup> March, 2018

Published online 28<sup>th</sup> April, 2018

**ABSTRACT**

Glomus jugulare tumour is type of extra adrenal chemodectoma or paraganglioma. Glomus jugulare tumors are rare and locally aggressive. Tumors may progress to cause lower cranial nerve palsies and involve the major vascular structures in skull base. We report a case of a 60 years old female who presented with impaired hearing and middle ear soft tissue mass. Appropriate use of imaging and histopathological examination of mass of ear led to early diagnosis.

**Key words:**

Chemodectoma, Paraganglioma, Skull base tumor

*Copyright©2018 Nitika Chawla et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.*

**INTRODUCTION**

**Background**

Glomus jugulare tumors originate from paraganglia called as the glomus bodies, located within the wall of the jugular bulb.<sup>1</sup> Paragangliomas accounting for 0.012% of all tumors, and 0.5% of all head and neck tumors.<sup>2</sup> The common presenting symptoms are tinnitus, decreased hearing, ear pain and vertigo.<sup>3</sup> Patients present over a wide range (10-85 years), with a mean in 6<sup>th</sup> decade. Sporadic tumors show a strong female predilection, whereas inherited or familial tumors are more common in men. If familial or syndrome associated, this is an autosomal dominant trait. Biopsy of the tumor is contraindicated due to bleeding risk.<sup>4</sup>

**Case presentation**

A 60 years old female presented to department of ENT, with symptoms of impaired hearing, pain and ear discharge from left ear for last 2 years. No history of dizziness was present. On examination a pulsatile reddish mass was seen in ear.

Radiological evaluation by CT showed a large expansile mass in medial part of external auditory canal with extension into the middle ear cleft, non visualisation of tympanic membrane and bony erosion involving the left external auditory canal. The possibilities of mitotic etiology and cholesteatoma were given. Exision biopsy of lesion was done and received in department of pathology.

*\*Corresponding author: Nitika Chawla*

BPS Govt. Medical College (W), Khanpur Kalan, Sonipat, Haryana

Histopathological examination of the slides showed round to oval cells arranged in nesting pattern, separated by spindle cells and fibrovascular stroma. The cells had abundant granular eosinophilic cytoplasm with uniform nuclei. Due to the abundant dilated blood vessels and artefact, Zellballen pattern was difficult to recognize. Differential diagnosis considered were hemangioma and jugulotympanic paraganglioma. IHC was performed and sustentacular cells became prominent on S-100 immunohistochemical stain. Chief cells were positive for chromogranin and synaptophysin. CD31 positivity in endothelial cells separated the interspersed blood vessels. The tumor cells were negative for cytokeratin (CK) and Desmin. Thus, immunohistochemistry helped in making the diagnosis by separating the chief cells, sustentacular cells and blood vessels.

**DISCUSSION**

Glomus tumors were first described by Rosenwasser in 1945.<sup>5</sup> In the head and neck, 2 anatomic groups of glomus tumor can be divided into 2 main categories-Cervical and Temporal bone paraganglioma. The cervical group includes carotid body tumor and glomus vagale. While jugulotympanic group comprises-glomus jugulare and glomus tympanicum.<sup>6</sup>

They are slow growing, benign but locally destructive tumors.<sup>7</sup> Patients are mostly middle aged and present with tinnitus, hearing loss and lower cranial nerve palsies.<sup>8</sup>

Histologically, the tumors are usually infiltrative and arranged in a characteristic clustered or zellballen architecture. These balls of paraganglia are surrounded by a layer of supporting or sustentacular cells and invested by a rich vascular stroma.

Tumor cells are small to intermediate and contain ample granular to basophilic cytoplasm and nuclei are monotonous. Mitoses are uncommon to rare.<sup>4</sup>

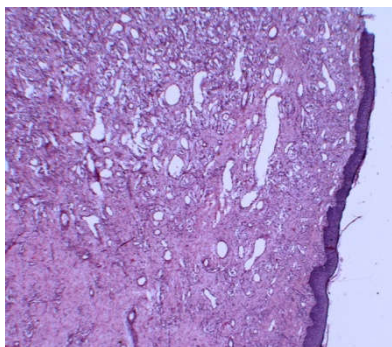
Malignant transformation has been reported in 6% glomus carotid body tumor and 2-4% in glomus tympanicum tumor.<sup>9</sup> Malignant tumor may show histological capsular invasion.<sup>10</sup>

The small size of biopsies from middle ear is the reason for diagnostic difficulties.<sup>4</sup> A variety of tumors enter the differential diagnosis, including schwannoma, neurofibroma, meningioma, metastatic and primitive neuroectodermal tumor. Immunoreactivity with synaptophysin, chromogranin and NSE along with sustentacular cells positivity with S-100 and lack of reactivity with keratin confirms the diagnosis of paraganglioma and exclude other tumors included in differential diagnosis.

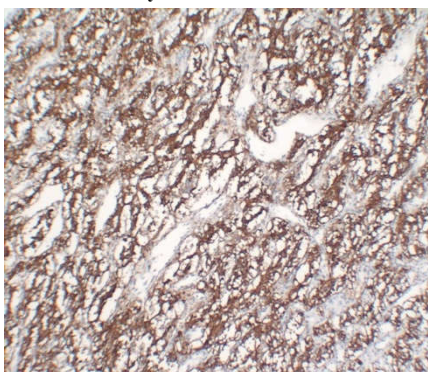
Treatment modality should be individualized for each patient. For smaller tumors with few symptoms, treatment aims to prevent progression of lesion. Surgical excision is standard procedure along with radiation therapy and stereotactic radiation are other options. Surgery and all therapies have complications like worsening of cranial nerve function. So it is appropriate to withhold the treatment for older patients with asymptomatic tumors. In these patients, wait and scan (with interval MR scanning) policy is advised.<sup>11</sup>

## CONCLUSION

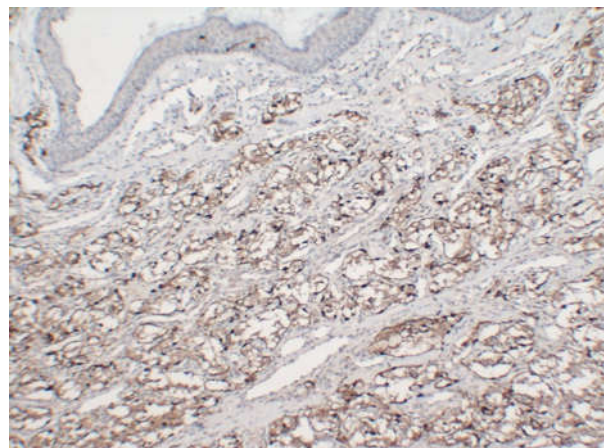
Glomus jugulare tumors are rare and can resemble hemangiomas on histology. However, loss of organoid growth pattern induced by surgical manipulation during removal of tumor, dense fibrous stroma and infiltrative growth pattern, result in wrong interpretation of growth. Judicious use of immunohistochemical markers comprising of S-100 and Neuroendocrine markers can help in differentiating these tumors. Thus, these tumors should always be kept in mind in differential diagnosis of vascular lesions of EAC and middle ear.



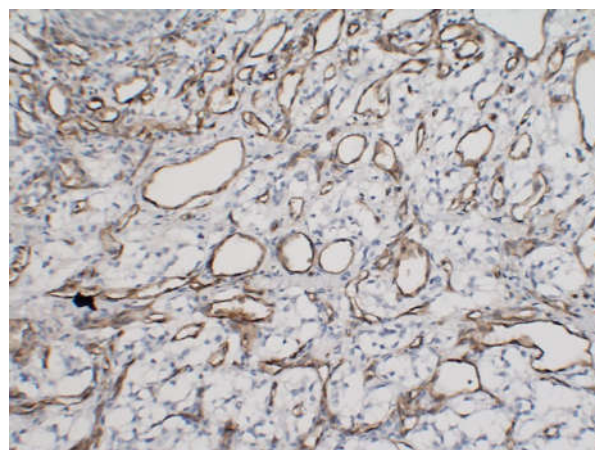
Hemotoxylin and Eosin stain 40x



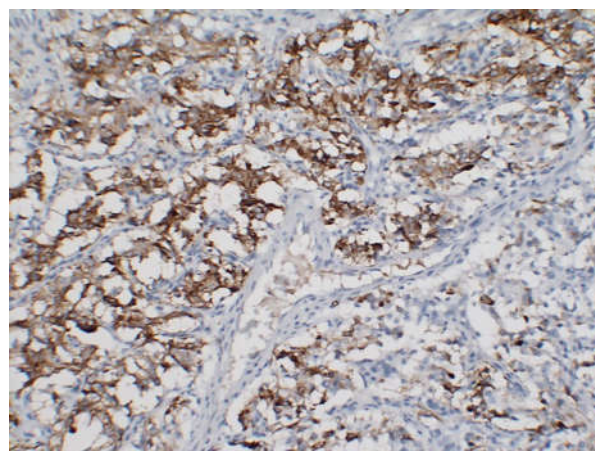
IHC:- Synaptophysin (100x)



IHC:- S-100 (100x)



IHC:- CD-31 (100x)



IHC:- CHROMOGRANIN (100x)

## References

1. Ashraf M, Rehman A, Akhter M, *et al.* Glomus jugulare: a case of secretory glomus jugulare with review of literature. Webmed Central Otorhinolaryngol 2010;2013:WMC00552
2. Lee JH, Barich F, Karnell LH, *et al.* National Cancer Data Base report on malignant paragangliomas of head and neck. Cancer. 2002; 94:730-737.
3. Guss ZD, Batra S, Li G *et al.* Radiosurgery for glomus jugulare: history and recent progress. Neurosurg focus 2009; 2013:1-6
4. Fletcher DM, In: Diagnostic Histopathology of Tumors, 4<sup>th</sup> Ed. New York; Elsevier. Tumors of The Ear. P 2132-34.

5. Rosenwasser H. Carotid body tumor of middle ear and mastoid. *Arch otolaryngol.* 1945; 41:64-67
6. O'Leary MJ, Shelton C, Giddings NA, *et al.* Glomus tympanicum tumors: a clinical perspective. *Laryngoscope* 1991;2013:1038-43
7. Kuhweide R, Lanser MJ, Fisch U. Catecholamine secreting paragangliomas at the skull base. *Skull Base Surg.* 1996; 6(1):35-45.
8. Woods C, Strasnick B, Jackson C. Surgery for glomus tumors: the Otolaryngology Group experience. *Laryngoscope.* 1993; 103 (11 Suppl 60):65-72.
9. Manolidis S, Shohet JA, Jackson CG, Glasscock ME., 3<sup>rd</sup> malignant glomus tumors. *Laryngoscope.* 1999; 109(1):30-34.
10. Karasov RS, Sheps SG, Carney JA, Van Heerden JA, De Quattro V. Paragangliomatosis with numerous catecholamine producing tumors. *Mayo Clin Proc.* 1982; 57:590-595.
11. Kumar S., Thakar A. Complex Glomus Jugulare Tumor: Management issues. *Indian J Otolaryngol Head Neck Surg.* 2013 Dec; 65(Suppl 3): 676-81.

**How to cite this article:**

Nitika Chawla *et al* (2018) 'Jugulotympanic Paraganglioma Presenting As Eac Mass, A Case Report', *International Journal of Current Advanced Research*, 07(4), pp. 11834-11836. DOI: <http://dx.doi.org/10.24327/ijcar.2018.11836.2063>

\*\*\*\*\*