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An unusual presentation of Jugulotympanic Paraganglioma –a rare case report

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Abstract: Paraganglioma or glomus tumor are named according to their origin. Jugulotympanic paraganglioma(JTP) are rare, slow growing benign tumors. This case report describes an unusual presentation of JTP in a 17 years old female. Clinical diagnosis was chronic suppurative otitis media with aural polyp. Microscopic examination revealed darkly stained basophilic cuboidal to epitheloid cell arranged in nesting, organoid and classical Zell- Ballen pattern. These clusters were surrounded by fibrous capsule of vascular core. The scant stroma between tumor island show increased vascularity and the diagnosis of JTP was made. This case report helps to avoid misinterpretation of JTPs.

Key Words: Jugulotympanic, paraganglioma, glomus tumor

Introduction: The carotid body was first described by Kohn a histologist in the early part of this century and he used the term paraganglioma. Paraganglioma in the head and neck are relatively rare tumors representing 0.6% of all neoplasms of the head and neck region. These tumors arise at four sites: carotid body, middle ear, jugular bulb and vagus nerve. They are also found in the orbit, larynx . Among them jugulotympanic paraganglioma (JTP) arise from anatomically dispersed paraganglia near the base of the skull and middle ear[1] . Guild first described vascularised tissue in the dome of the jugular bulb and on the promontory of the middle ear and named as glomic tissue in 1941[2]. Actually, jugulotympanic paraganglioma are those arising from paraganglia situated in the vicinity of the jugular bulb and tympanic paraganglia. They are usually located along the course of Jacobson and Arnold's nerve in the middle ear cavity. However it is difficult to distinguish between these two entities and the term JTP is favoured. The incidence of JTP is approximately 1: 1,300,000[3]. The tumor is typically vascular and grows from capillary and pre-capillary vessels in between epithelial cells. The cell nests are less uniform and frequently smaller compared with other paragangliomas. Dense sclerotic matrix is another characteristic of JTP[4].This case report describes an unusual presentation of Jugulotympanic paraganglioma in a 17 years old female.

Case Report: A 17 years old female presented to ENT out patient department with the history of swelling in the ear since 8 months. She had also left sided earache and hearing disturbance since 6 months. Her past history, family history, medical history was unremarkable. On physical examination the left external auditory canal was narrowed and was filled with a protruding mass with an invisible tympanic membrane. Trozer's sign was present. Fistula test and tenderness was negative. Audiometry findings revealed

conductive type of deafness. Radiological findings were not available. Right ear was completely normal, nasal and oral cavity examination was normal. Clinical diagnosis was chronic supportive otitis media with aural polyp. Routine laboratory investigations was performed and was within normal limits. Initially patient was treated with a course of antibiotics. Later on patient was posted for left canal wall mastoidectomy. Intra-operative findings was an extensive granulomatous polyp, firm in consistency and bleeds on touch. There was subtotal perforation of tympanic membrane. The mass was removed and sent for histopathological examination.

The tissue was processed routinely in the histopathology section and the sections were stained with haematoxylin and eosin (H/E).Microscopic examination revealed a polypoidal structure lined with a stratified squamous epithelium. The subepithelial stroma showed clusters of darkly stained basophilic cuboidal to epitheloid cell arranged in nesting, organoid and classical Zell- Ballen pattern. These clusters were surrounded by fibrous capsule of vascular core. The scant stroma between tumor island show increased vascularity giving homogenous appearance and occasional clusters of lymphocytes as shown in figure 1, 2, 3 . This was diagnosed as Jugulotympanic Paraganglioma.

Figure 1: Shows low power view of jugulotympanic paragnglioma having clusters of darkly stained basophilic cuboidal to epitheloid cell arranged in nesting, organoid and classical Zell- Ballen pattern (H/E stain, 10X)

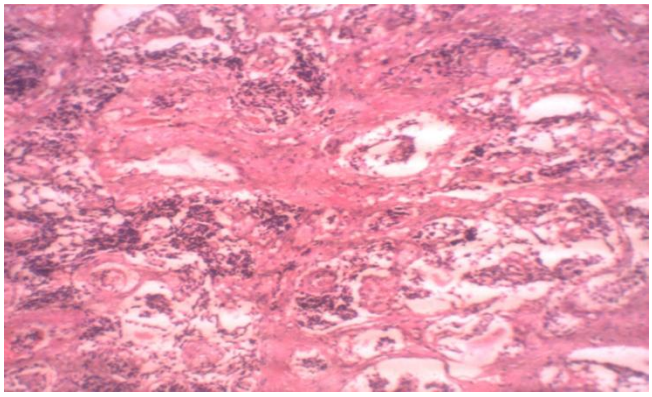


Figure 2 : Shows high power view of jugulotympanic paraganglioma having clusters of darkly stained basophilic cuboidal to epitheloid cell arranged in nesting, organoid and classical Zell- Ballen pattern (H/E stain, 40X)

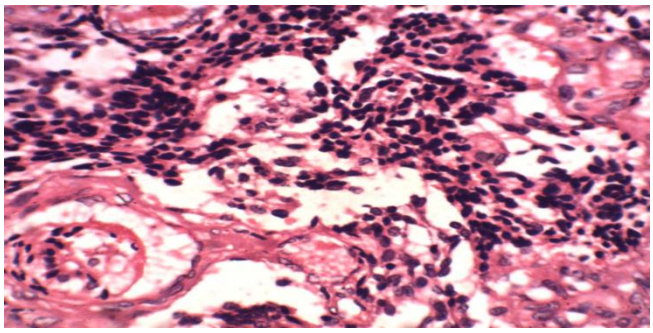
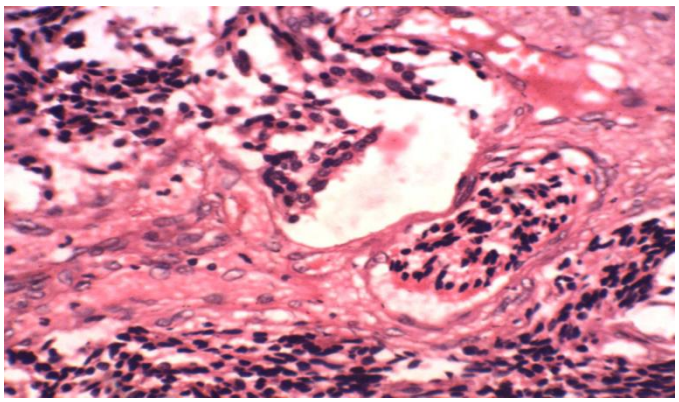


Figure 3 : Shows high power view of jugulotympanic paraganglioma having clusters of darkly stained basophilic cuboidal to epitheloid cell arranged in nesting, organoid and classical Zell- Ballen pattern (H/E stain, 40X)



Discussion: Paraganglioma or glomur tumor are named according to their origin. JTP originates in the middle ear. Because JTPs are rare tumor and mimick other vascular tumor, middle ear adenoma, chronic suppurative otitis media and are easy to misdiagnose[5] .

JTPs usually develop in adults in the 5th to 6th decade of life and there is a definite female preponderance with a female: male ratio of 4-6:1. In this case, the patient was female aged 17 years which is an unusual presentation. But Ji – Youn Surg et al reported a case in 18 years old boy[4]. The difference between familial and sporadiac

cases of JTPs are not known. Kliewer et al reported that familial paragangliomas were found in younger patient (mean age: 25.8 yrs) than in the rest of paraganglioma patient (mean age: 42.5yrs) [6] .

But Maria Eugenla et al reported right ear involvement more common[7] . In the present case the left ear was affected which is again unusual. However, we are unable to find data in the literature corroborating this incidence.

Clinically, the first symptom of JTP is pulsating tinnitus, due to slow growing mass, the symptoms will often occur late. The second most common symptom is hearing loss, conductive at first . When the inner ear is affected in case of central tumor growth, sensorineural hearing loss may occur[8] . In the present case the first symptom of the patient was swelling in the ear , later on she complained of earache and conductive hearing loss unilaterally.

The hearing disturbances can be identified by audiometry and impedanciometry helps to diagnose during the examination, the tumor pulsation will shift the balance-meter needle synchronically to patient's pulse. Diagnosis is confirmed by CT scan and MRI studies are recommended when there is evidence of intracranial invasion, as well as neck and large vessels involvement. Digital angiography also helps to study the features of vascular tumor and its vascular supply[7] .

The choice of treatment is surgical removal, if surgery is contraindicated radiotherapy is indicated. In the present case open cavity mastoidectomy.

Histologically JTPs can be confused with glomus tumors which are tumors of perivascular cells. These two entities are definitely different diseases. JTPs are tumor arising from paraganglia whereas glomus tumor arise from a modified smooth muscle cell located in the walls of specialized arteriovenous anastomoses involved in temporal region. Hence, it is better to use the term JTPs rather than glomus tumor to avoid confusion. The second differential diagnosis is epitheloid hemangioendothelioma (EHE) because of the high vascularity of JTPs. EHE have moderate amount of eosinophilic cytoplasm and immunoreactive for vascular markers such as CD31, CD34 and factor VIII. However, careful microscopic examination revealed that, the mass was shown to be neuroendocrine tumor. Another differential diagnosis is middle ear adenoma (MEA) , which is thought to arise from pluripotent cells in the middle ear mucosa. As these tumors are less vascular and have mixed patterns of differentiation, ranging from glandular to neuroendocrine may be helpful to differentiate it from JTPs[9] . The last differential diagnosis is CSOM, as this has chronic inflammatory cells in the mucosa.

In conclusion, This case report describes about JTP in an unusual age, unusual side and clinical presentation. This entity has to be kept in mind when we interpret middle ear or temporal bone masses.

References:

1. Kohn A. Dieparaganglien Arch Mihr. Anat. 1908;62:263.

2. Guild SR. A hitherto unrecognised structure, the glomus jugularis, in man. Anat. Rec. 1941;88:3-40.

3. Fisch U, Mattox D. Classification of glomus temporate tumors. In:fish U, Mattox D Eds. Microsurgery of the skull base. Stuttgart and New York, Georg Thieme. 1988;149-153.

4. Ji-Young Sung, Chang II Cha, Young-Koo Park. Jugulotympanic paraganglioma, mimicking a vascular tumor. The Korean Journal of Pathology. 2010;44:543-6.

5. Sharmin Ferdousi, SM Badruddoza. Jugulotympanic paraganglioma mimicking chronic suppurative otitis media. AKMMC J. 2012;3(2):37-38.

6. Kliewer KE, Wen DR, Cancilla pa, Cochran AJ. Paragangliomas: assessment of prognosis by histologic, immunohistochemical and ultrastructural techniques. Hum Pathol. 1989;20:29-39.

7. Maria Eugenia LRB de V Neto, Isabela M de Vuono, Luiz RO Souza, Jose RG Testa. Tympanic paragangliomas. Rev. Bras. Otorrinolaringol. 2005;1:71.

8. Anutunes ML, Testa JRG, Frazatto R, Freitas CE. Paraganglioma tympanico. Acta WHO. 1999;18(3):154-8.

9. Berns S, Pearl G. Middle ear adenoma. Arch Pathol Lab Med. 2006;130:1067-9.sss

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