



JUGULO-TYMPANIC PARAGANGLIOMA, A RARE CASE REPORT WITH REVIEW OF LITERATURE

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<p>Article Info <i>Received 15/01/2015</i> <i>Revised 27/01/2015</i> <i>Accepted 22/02/2015</i></p> <p>Key words: Paraganglioma, Jugulotympanic glomus tumour.</p>	<p>ABSTRACT Jugulotympanic paragangliomas (JTPs) are neoplasms of variable Invasiveness arising from the Para ganglia situated around the jugular bulb or middle ear. Here we report a case of JTP in 48 years, female with pre-operative diagnosis of mastoiditis Radiological (CT) examination and biopsy were inconclusive. Pre operatively it was highly vascular with severe bleeding. Histopathologic finding revealed cell nests with many dilated small blood vessels in between. The nuclei are round with uniform, granular chromatin; prompting a diagnosis of JTP. As these tumours are very rare and easily misdiagnosed, the awareness of peculiar histological features may help the clinician to avoid such misinterpretation.</p>
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INTRODUCTION

The term paraganglion was first described by the histologist Kohn in the early part of this century to describe the carotid body [1]. The first case of jugulotympanic paraganglioma (JTP) was reported by Rosenwasser in 1945 [2].

Head and neck paragangliomas are relatively uncommon tumors, representing 0.6% of all neoplasms of the head and neck. they are classified into four types based on anatomic origin as carotid body, jugulotympanic, vagal and laryngeal. Among them, JTPs arise from anatomically dispersed paraganglia near the base of the skull and middle ear. JTPs tend to be more vascular and cell nests are less uniform and frequently smaller compared with other paragangliomas [3].

Jugulotympanic paraganglioma are the most common tumor among the middle ear tumors though it is rare. with an incidence of 1:1,30,000 [4]. They affect women more often than men and they are common in 5th - 7th decade of life. They also have been reported in other unusual sites, such as the gallbladder, the biliary duct system, and the urinary bladder.

CASE SUMMARY

A 48 year old lady presented with earache and discharge since 3 days. She noticed sudden appearance of mass in the ear. She is diabetic, hypertensive since 10 years and on medical treatment for the same. Otoscopic finding revealed reddish fleshy mass occupying the ear canal completely and unhealthy tympanic membrane was visualised. Systemic examination was not significant. CT scan was reported as right sided mastoiditis with soft tissue density lesion in right external auditory canal suggestive of polyp (Fig-01). Antral polypectomy was done. Per operatively, the mass was highly vascular with severe bleeding. Excision of the mass was done, Submitted for histopathological examination,

Microscopy showed keratinized stratified squamous epithelium beneath which is seen tumor mass composed of cells arranged in nests (Fig-02). These nests are separated by slender fibrovascular septae. Tumor cells are round to oval having irregular cell border with granular chromatin and prominent nucleoli (Fig-03). Also seen are dilated and congested blood vessels.



Immunohistochemistry: Peripheral Sustentacular cells were positive for S-100. A diagnosis of jugulotympanic paraganglioma (JTP) was made.

Figure 1. CT scan image showing right sided mastoiditis with soft tissue density lesion in right external auditory canal suggestive of polyp

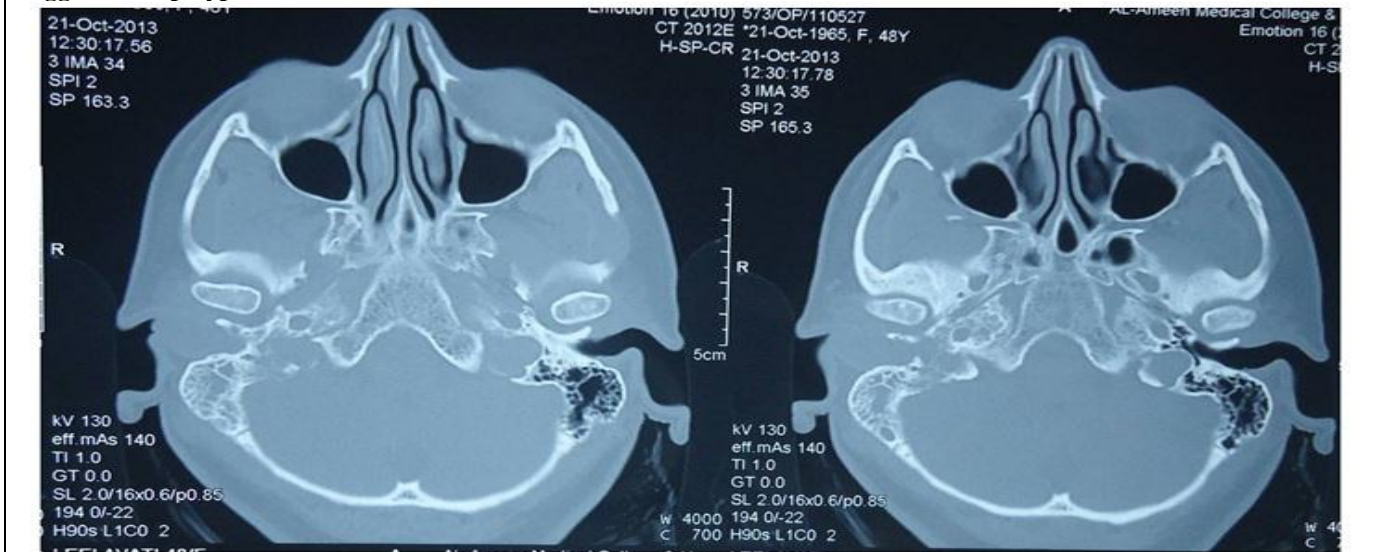


Figure 2. Microscopy showing keratinized stratified squamous epithelium beneath which is seen tumor mass composed of cells arranged in nests [H&E 100x]

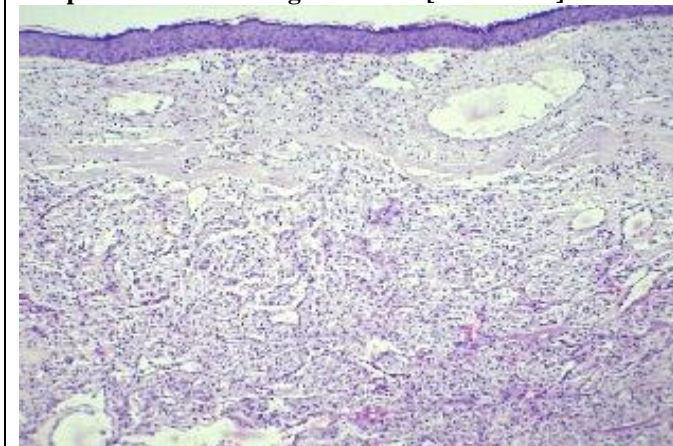
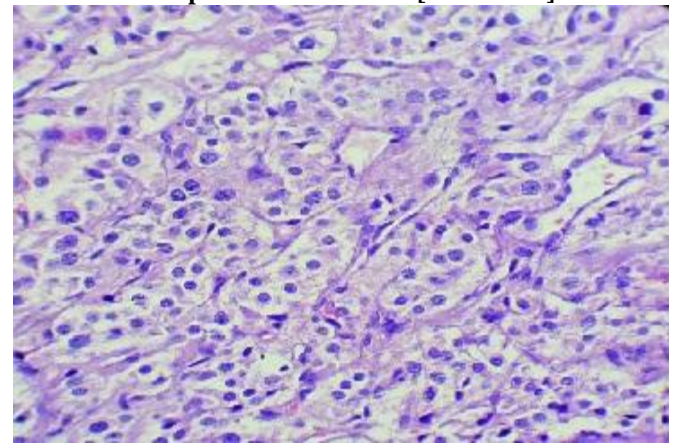


Figure 3. Higher magnification showing round to oval cells having irregular cell border with granular chromatin and prominent nucleoli. [H&E 400x]



DISCUSSION

Jugulotympanic paragangliomas (JTP) can be confused with vascular tumor, middle ear adenoma and even with a chronic otomastoiditis. The two entities – glomus tumors and JTPs that are called ‘glomus tumors’, are definitely different lesion. JTPs are tumors arising from paraganglia whereas glomus tumors arise from a modified smooth muscle cell located in the walls of specialized arteriovenous anastomoses involved in temperature regulation [1]. Clinically, the typical presentation is that of a red mass protruding behind the tympanic membrane or extending in the canal. Profuse bleeding may be encountered at the time of biopsy [5]. Head and neck paragangliomas can be functional and show

hyperadrenergic syndrome despite being less frequent than pheochromocytoma or abdominal/pelvic paraganglioma. So, in the patient showing hyperadrenergic manifestations without an adrenal, abdominal or pelvic mass, head and neck paraganglioma must be considered [3].

The incidence of clinically functioning paraganglioma with symptoms and signs of norepinephrine excess, particularly hypertension, is only 1–3% [6]. Only 15 reports in the English literature are about malignant head and neck paragangliomas including JTPs [3].

JTP usually arise laterally in the temporal bones and, through erosion of the floor of the hypotympanum, present as a mass in the middle ear or the external auditory canal [5].

JTPs are slow-growing tumors but may be locally invasive, with extension into and destruction of adjacent structures, including the temporal bone and mastoid. Intracranial extension may occur in up to 15% of cases [7]. The paragangliomas appear grossly as sharply circumscribed polypoid masses and they have a firm to rubbery consistency. They are highly vascular tumors and may have a deep red colour. Paragangliomas are composed predominantly of chief cells that are round to oval with uniform nuclei, a dispersed chromatin pattern and abundant eosinophilic granular cytoplasm. The sustentacular cells are located at the periphery of the cell nests. The hallmark histological feature is presence of cell nests or Zellballen pattern. The stroma surrounding and separating the nests is composed of prominent fibrovascular tissue. Although this pattern is characteristic of paragangliomas, it can also be seen in other tumors such as any neuroendocrine tumors including carcinoids, atypical carcinoids and melanomas

The diagnosis of JTP is facilitated by immunohistochemical stains. The immunohistochemical antigenic profile of paragangliomas includes chromogranin and synaptophysin positivity in the chief cells and S-100 protein staining localized to the peripheral sustentacular cells.² Some studies have suggested that malignant paragangliomas will have either absent or decreased S-100 staining sustentacular cells [8]. The differential diagnosis of JTP includes middle ear adenoma, meningioma, melanoma, carcinoids tumors and acoustic neuroma. If histology is not definitive in separating JTP from these tumors, immunohistochemical reactivity can differentiate them [2]. Middle ear adenoma and carcinoids patients comes to clinical attention with unilateral conductive hearing loss and otoscopic findings shows usually intact tympanic membrane with fluid level or mass noted behind

the ear drum. Histological nuclear features shows fine dispersed chromatin which differs from nuclear features of JTP. Immunohistochemical profile is positive for pancytokeratin marker, in contrast to JTP [9]. Patients of Meningiomas presents with progressive hearing loss, headache and loss of equilibrium. In radiographic findings, the pathognomonic feature of meningioma is presence of speckled calcification in the soft tissue mass.⁹ Immunohistochemistry for meningioma shows positivity for epithelial membrane antigen and vimentin. In contrast to JTP, Meningiomas are negative for neuroendocrine markers [2].

Complete surgical excision is the treatment of choice; however, the location and invasive nature of these lesions often preclude complete resection. In such cases, radiotherapy is a useful adjunct to surgery. Preoperative embolization is useful in decreasing the tumor's vascularity and facilitating surgical resection. Local recurrence is seen in as many as 50% of cases. The histologic appearance of paragangliomas does not correlate to their biologic behavior [9]. JTP is a neoplasm of slow growth. The jugular variety infiltrates the petrous bone, but distant metastasis is rare. Radiation therapy, and in some cases surgery, offers a high rate of cure for these neoplasms.

CONCLUSION

Jugulotympanic paragangliomas can have varied presentations in the temporal region. Hence, suspicion of paragangliomas should be kept in any case of middle ear mass to avoid intra-operative complications. As they are rare and easily misdiagnosed clinically, the existence of such a lesion warrants the clinician to avoid intraoperative complications and profuse bleeding.

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