Paraganglioma of External Auditory Canal: A Rare Entity Case Report

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Abstract

Paragangliomas are benign tumours originating from the paraganglia. In the ear paragangliomas are found commonly in middle ear or jugular bulb. Paragangliomas of external auditory canal are extremely rare. Our case describes a woman in her 5th decade presenting with difficulty in hearing since 6 months and swelling in the right ear. Physical examination revealed a red to pink swelling, non friable, bleeding on touch and obscuring the external auditory canal completely. Differential diagnosis of granuloma/exostosis was considered. Patient underwent excision biopsy. Hemorrhage during surgery was copious and suspicion of vascular lesion was raised. Histopathology and immunohistochemistry confirmed the diagnosis of paraganglioma. The aim of this article is to make clinicians aware of the entity for diagnosis and appropriate treatment.

Key Words: Paraganglioma, external auditory canal, granuloma

Introduction

Paragangliomas are rare benign neoplasms originating from the neuroectodermal tissue derived from paraganglion cells of the autonomic nervous system. Paragangliomas are most common in the adrenal glands and if found elsewhere they are labeled as extra-adrenal pheochromocytomas.[1]

In head and neck region anatomically they are classified as carotid body tumours and glomus vagale(cervical) and jugulotympanic(glomus jugulare and glomus tympanicum) with incidence of carotid body tumours make upto 60%, glomus jugulare 13% and glomus tympanicum 6%.

In ear paragangliomas are represented as glomus bodies which are in close association with Jacobsons nerve(tympanic branch of glossopharyngeal nerve), Arnolds nerve(auricular branch of vagus nerve) or the adventitia of the jugular bulb.[2]

There are very few reported cases of paraganglioma restricted to external auditory canal as is our case without recurrence following surgical excision.

Case Presentation

44 years old female presented with a mass in the right external auditory canal with difficulty in hearing in right ear since 6 months with occasional bleeding from the ear. She was a known case of hypertension under control with anti-hypertensives. Physical examination of her right ear revealed a normal appearing pinna with a soft to firm mass,pink in colour appearing like a granuloma covered with skin in the right external auditory canal fully obscuring

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the external auditory canal. The tympanic membrane was not visible. On probing,probe could not be passed in the posterosuperior region.The mass was attached superiorly. All other otologic examinations were normal.

An excisional biopsy was performed under general anaesthesia through endaural approach. The tumour was attached to postero-superior bony meatal wall. There was brisk and copious bleeding encountered during the surgery. The tympanic membrane was intact. The tumour was vascular and was completely excised. Hemostasis was achieved with the use of bipolar cautery and ear pack.

Upon histopathological examination was done. The tumour was located under the epidermis with size 1X0.5cms. Classic organoid zell ballen pattern of paraganglioma was noted with central round chief cells containing eosinophilic granular and at places vacuolated cytoplasm. Prominent fibrovascular stroma separated the nests. There was no evidence of necrosis or mitosis. Immunohistochemistry was positive for synaptophysin and chromogranin and negative for S100. At 6 months follow up postoperative the canal was clear with no recurrence.

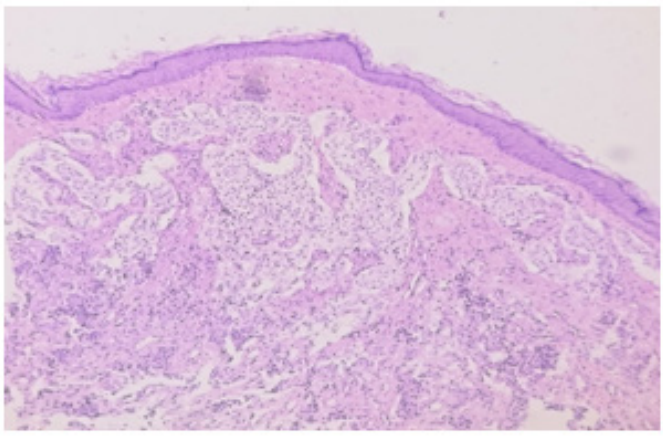

Figure 1

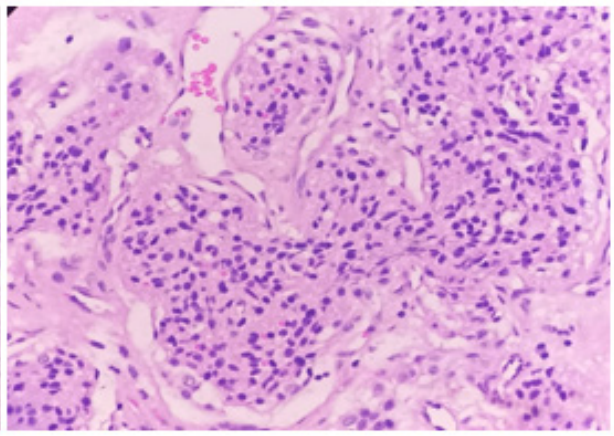

Figure 2

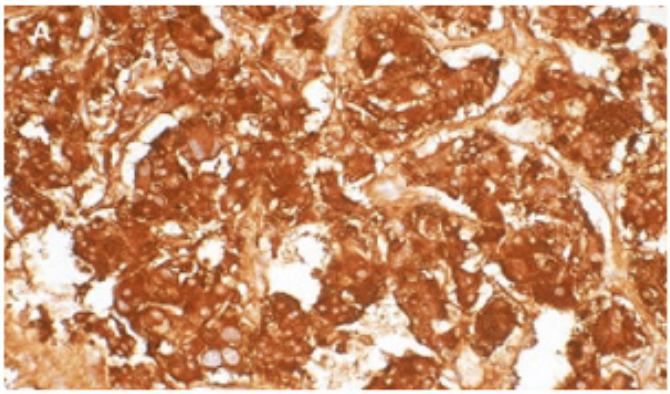

Figure 3

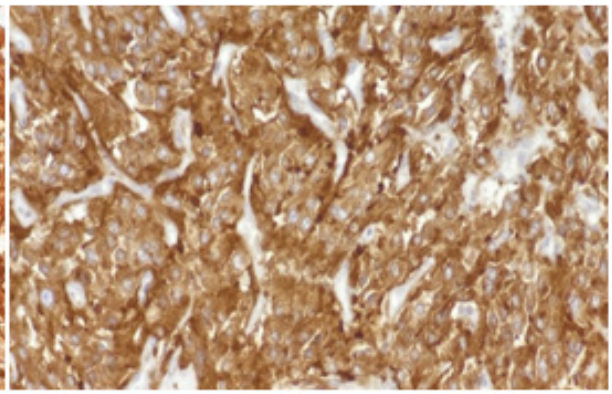

Figure 4

Figure 1: Tumour located under the epidermis with organoid zell ballen pattern and prominent fibrovascular stroma. (100X) Figure 2: Classic organoid zell ballen pattern of paraganglioma with central round chief cells containing eosinophilic granular cytoplasm. (400X). Figure 3 and 4: Immunohistochemistry positivity for chromogrannin and synaptophysin respectively.

Discussion

Glomus jugulare tumours are rare benign paragangliomas arising from the adventitia of jugular bulb and are embryologically derived from the chromaffin cells. These tumors represent 0.6% of all the head and neck tumours. Paraganglioma

confined to the external auditory canal is very rare and is closely related to Arnold's nerve and the tumour in our case also originated from the Arnold's nerve.[3] These are usually solitary tumours occurring in middle aged persons predominantly women.

The patient most often presents with pulsatile tinnitus, hearing loss, vertigo, otalgia and otorrhoea. Neuropathies of cranial nerves VII,IX,X, XI and XII are common.[4] On otoscopic examination one usually sees a red blue vascular mass behind the tympanic membrane. It is described as a rising sun from the hypotympanum. In our case it appeared like a granuloma in the external auditory canal, the traditional salmon pink appearance of a glomus tympanicum was not seen. The diagnosis was confirmed on histopathological examination.

Paragangliomas are generally benign or low grade malignancy but they can be locally invasive with destruction of bones and adjacent structures. the potential routes of spread to middle ear includes the foramen of various nerves, blood vessels that connect the external auditory canal skin to the middle ear such as the auricular branch of the vagus nerve or the chordate tympani nerve.[5]The tumours have a median growth rate of 1mm per year and median doubling time of 4.2 years.

Imaging with contrast aids in the diagnosis of paragangliomas. HRCT is a useful tool to delineate the adjacent bony details. MRI can be used as a imaging modality for soft tissue lesion when suspicion for bony erosion is low to access such rare pathology. In our case since the pathology of granulomas was considered radiological intervention was not done and may not be essential for such diagnosis.[6]

In general the differential diagnosis of external auditory canal polyps includes exostosis, osteoma, fibrous dysplasia, granuloma, ceruminous gland tumour, epidermoid cyst, cholesteatoma, papilloma and malignancy. Paraganglioma was not considered as differential diagnosis.[7]

The diagnosis of paraganglioma requires histopathological examination and immunohistochemistry. Classic organoid (zellballen) pattern and IHC analysis positive for chromogranin and synaptophysin confirms the tumour. Surgical removal is the first line of treatment. Conventional radiation therapy can be used if surgery is not possible. Paraganglioma confined to the external auditory canal is rare and recurrence has to be evaluated. Also mutation in succinate dehydrogenase gene is seen in 40% of patients of head and neck paragangliomas making them vulnerable to develop multiple paragangliomas requiring regular follow up of the patient.[8]

Conclusion

Paraganglioma of the external auditory canal is rare and can be misdiagnosed as granuloma,polyp,exostosis. Paraganglioma should be considered in the differential diagnosis of external auditory canal mass, to make a clinical diagnosis and initiate appropriate treatment.

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Conflict of Interest - Nil.

References

1. Suárez C, Sevilla MA, Llorente JL. Temporal paragangliomas. Eur Arch Otorhinolaryngol 2007;264:71931.
2. Skinner LJ, Curran AJ, Barnes C, Timon CI. Paraganglioma of the external auditory canal: An unusual case. J Laryngol Otol 2000;114:3702.
3. Lack EE, Cubilla AL, Woodruff JM, Farr HW. Paragangliomas of the head and neck region: A clinical study of 69 patients. Cancer. 1977; 39: 397-409.
4. Scott Brown's Otolaryngology (1997) : Glomus and other tumors of the ear. (6 'h Edn.); Vol.3, Chapter 23.
5. Brown J. Glomus jugulare tumours revisited: a ten year statistical follow-up of 231 cases. Laryngoscope 1985;95(3):284-8..
6. Guild SR. The glomus jugulare: A non-chromaffin paraganglioma in man. Ann Otol Rhinol Laryngol 1953;62:1045-71.
7. Kundu I, Goswami S, Barman D, Biswas S. Parganglioma of external auditory canal. Indian J Otolaryngol Head Neck Surg 2001;53:304-6.
8. World Health Organization. Pathology and Genetics: Tumours of Endocrine Organs. Lyon, France: International Agency for Research on Cancer; 2004.