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Case Report

When Glomus Strikes the Middle Ear: A Rare Case Presentation

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Abstract

Glomus tumor is a rare hamartoma, most often seen in young females, typically in the subungual region. Extradigital sites like the middle ear are uncommon and often misdiagnosed due to nonspecific presentation. Glomus tumor is a rare (7%) benign neoplasm arising from the glomus body, a neuromyoarterial structure involved in thermoregulation. We present a rare case of glomus tumor in the middle ear.

Keywords: Tumor, Ear, Neoplasm arising, Case report, Soft tissue.

INTRODUCTION

Glomus tumor is a rare (7%) benign vascular tumor arising from the glomus body, a neuromyoarterial structure involved in thermoregulation (controls blood flow to skin). Glomus tumors were first described by Hoyer in 1877 while the first complete clinical description was given by Masson in 1924. It accounts for <2% of soft tissue tumors and is most often seen in the dermis of fingers, toes, (especially subungual region) palms, soles, forearms, rarely visceral organs. Classical triad is severe pain, point tenderness, cold insensitivity. It is small, well circumscribed, bluish nodule. Combination of glomus cells, smooth muscle cells and vasculature. Atypical and malignant glomus tumors are more commonly deep seated than benign tumors.[1] Estimated incidence of 1.6% of soft tissue tumors. Most occur in adults aged 20 – 40. Equally common in both genders; subungual lesions have a female predominance of 3:1. Extradigital sites like the middle ear are rare and challenging to diagnose. Where strong clinical suspicion and expert diagnostic capability is required for diagnosing glomus tumor of ear. Here we present a rare case of glomus tumor in the middle ear.

CASE REPORT

A 67-year-old female presented with left ear pain since 1 year, associated with giddiness and tinnitus which at first sight was diagnosed as Cholesteatoma. Later as she further presented with otorrhea, pulsatile tinnitus and hearing loss in the left ear. Eventually, she was treated successfully with a canal wall down mastoidectomy. Grossly we Received 2 grey brown to grey black soft tissue bits each measuring 0.4cms which were all embedded

Microscopically it shows many tumor cells which are uniform, round with large nuclei and abundant, eosinophilic cytoplasm (Glomus cells) [Figure 1]. The glomus cells surround dilated, thin- walled blood vessels and the stroma may show areas of hyalinization [Figure 2].

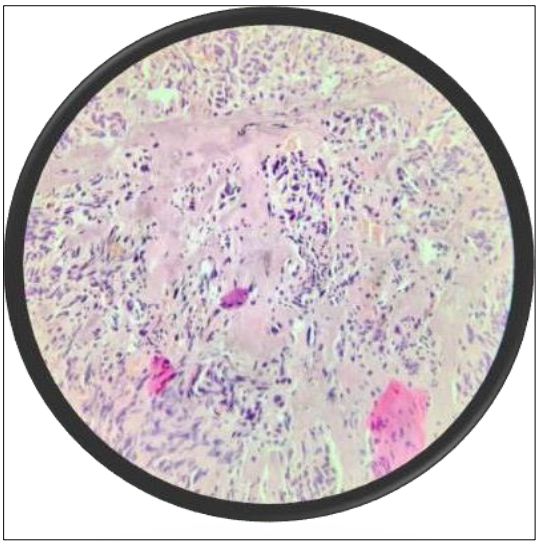

Figure 2: H&E, 200x, section studied shows focal areas of hyalinization and glomus cells

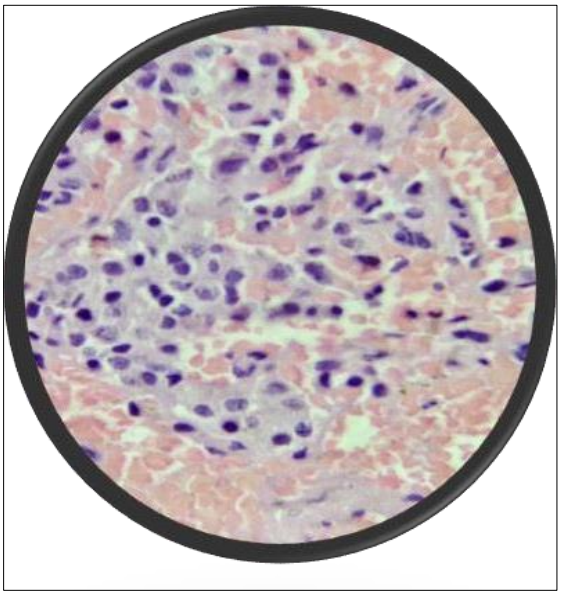

Figure 1: H&E, 400x section studied shows glomus cells are uniform, round cells with large nuclei and abundant eosinophilic cytoplasm

Further we suggested IHC with Desmin and SMA which show

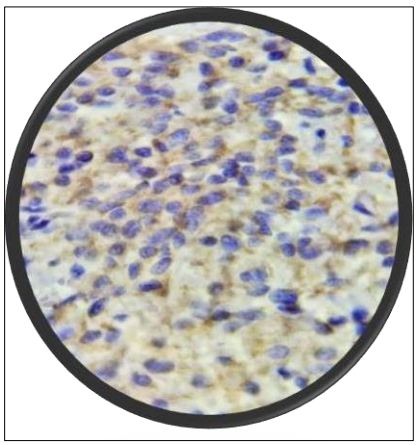

Figure 3: IHC: Desmin Negative

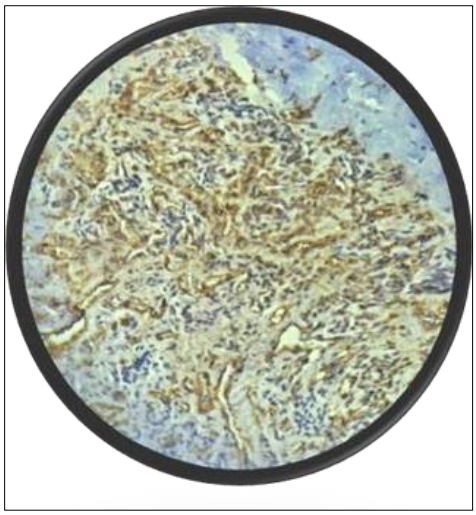

Figure 4: IHC: SMA Positive

DISCUSSION

Glomus tumor is a rare, usually benign neoplasm that arises from the glomus body, a specialized arteriovenous anastomosis involved in thermoregulation. It is best regarded as a hamartoma, meaning a tumor-like malformation composed of an abnormal mixture of tissue elements normally found at that site. The classic location of this tumour is the subungual region of the fingers or toes, especially beneath the nail bed, where it presents with excruciating pain, tenderness to touch, and sensitivity to temperature changes — a characteristic triad that strongly suggests the diagnosis. Although subungual lesions are most typical, extradigital glomus tumors can occur in unusual locations such as the middle ear, stomach, trachea, mediastinum, or soft tissue of the extremities.

These extradigital tumors are frequently misdiagnosed because of their nonspecific symptoms and variable presentations.

Clinically, glomus tumors are small, often less than 1 cm, and present as a solitary, bluish, dome-shaped papule or nodule. When present in the middle ear, they may mimic other neoplastic or vascular lesions, resulting in delayed recognition. The diagnosis is usually confirmed by high-resolution imaging techniques like MRI and CT, which reveal a hypervascular lesion. MRI typically shows a characteristic "salt and pepper" appearance, representing areas of hemorrhage and high-flow vessels interspersed with tumor cells. Surgical excision remains the definitive treatment, providing both diagnosis and cure. Histopathological Classification Histologically, glomus tumors are composed of uniform, round glomus cells arranged around dilated vascular channels.[3]

Based on their predominant histological component, they are classified into three subtypes:

1. Solid Glomus Tumor (75%) – This is the most common form, characterized by solid sheets and nests of glomus cells with scant vascularity and minimal smooth muscle component.
2. Glomangioma (20%) – In this type, vascular channels are more prominent, and the tumor resembles a hemangioma with intervening glomus cells.
3. Glomangiomyoma (5%) – The rarest variant, exhibiting significant smooth muscle differentiation along with glomus cells and vascular channels.

All these variants are typically benign; however, malignant transformation (glomangiosarcoma) is possible but extremely rare. Features suggesting malignancy include size >2 cm, deep location, high mitotic rate, and cytologic atypia.

Immunohistochemistry is useful in diagnosis — glomus cells are positive for smooth muscle actin (SMA) and vimentin, but negative for desmin and S-100 protein, helping to differentiate them from other soft tissue neoplasms. In contrast, middle ear paragangliomas — historically referred to as glomus tympanicum or glomus jugulare tumors — are neuroendocrine tumors arising from paraganglionic cells associated with cranial nerves IX (glossopharyngeal) and X (vagus). These are not true glomus tumors despite the historical name; they are paragangliomas, derived from chief cells of the paraganglia that function as chemoreceptors. These tumors are slow-growing but locally invasive, capable of eroding bone and invading surrounding structures. They are the most common primary tumor of the middle ear and temporal bone, most frequently seen in women between 40 and 69 years of age.

About 85% of these lesions arise from the jugular bulb (glomus jugulare), producing a mass in the middle ear or external auditory canal. Around 12% originate from the tympanic branch of the glossopharyngeal nerve (Jacobson's nerve), forming the

glomus tympanicum, and the remaining 3% arise from the auricular branch of the vagus nerve (Arnold's nerve).

CONCLUSION

Eventually, the glomus tumour is fairly a rare benign tumour that clinicians should keep in mind as a differential diagnosis when facing any nodule and don't rule out when the tumour is extradigital

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