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**Review Article**

# HISTOPATHOLOGICAL AND IMMUNOHISTOCHEMICAL UNRAVELLING THE MYSTERY OF ANCIENT SCHWANNOMA- A CASE REPORT WITH BRIEF REVIEW

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**ABSTRACT**

Schwannoma or neurilemmoma is an uncommon benign tumour in the oral cavity that originates from the Schwann cells on the neural sheath of the peripheral nerves. We report a case of 65year old male patient who presented to our college with a swelling in the preauricular area. Computed tomography (CT) imaging was done to know the extent of lesion. Excision biopsy of the lesion was done. The histologic examination and the immunohistochemistry confirmation with S100 proved it to be ancient schwannoma of the preauricular region.

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## INTRODUCTION

Schwannoma or neurilemmoma is an uncommon benign tumor in the oral cavity that originates from the Schwann cells on the neural sheath of the peripheral nerves.[1] Verocay first described the entity schwannoma in 1908.[2] They occur most often in head and neck region and the flexor surfaces of the upper and lower extremities. Enzinger and Weiss have described four variants of schwannomas, viz. the common schwannoma, the plexiform schwannoma, the cellular schwannoma and the ancient schwannoma, all of which are benign.[2] 1-12% of all the schwannomas are located in the oral cavity.[1] Intra orally schwannomas can arise from both soft tissue and bone. Generally schwannomas manifest clinically as slow growing, asymptomatic solitary nodules, with no gender predilection and with a typical patient age of onset between 20-50 years.[4]

Ancient schwannoma is a rare variant, which is long standing schwannoma exhibiting degenerative changes. It was first described by Ackerman and Taylor in 1951. It rarely affects the head and neck area and its intraoral occurrence is all the more rare. Eversole and Howell(1971) reported the first ancient schwannoma of head and neck region.[3] Here we report a rare case of schwannoma occuring in the preauricular region.

## CASE REPORT

A 65 year male patient reported to our college with chief complaint of swelling in the right facial region in front of the ear since 2½ years. The swelling was firm in consistency, freely mobile, measuring 5cm x 4cm in size, tender on percussion with no discharge and rise in local temperature. On palpation submandibular lymphnodes were palpable and measuring 2cm x 2cm in size.

Radiographic examination revealed a well circumscribed radio opaque lesion superficial to the parotid gland. Computed tomography(CT) shows a hyperintense area with few hypointense areas within it[Figure:1].

A clinical diagnosis of benign histiocytic fibroma was given. Complete excision of the mass was done.

On gross examination the excised specimen revealed a well encapsulated mass measuring 3.8cm x 3cm in size, firm in consistency, whitish brown in colour[Figure:2]. The specimen was subjected for histopathological examination for H and E stain. The section was also sent for immunohistochemical (IHC) study using S100 marker to confirm the nerve tissue origin.

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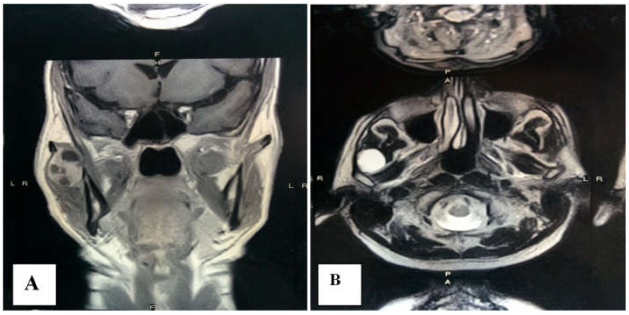


**Figure 1** CT scan demonstrating the well circumscribed hyperintense area superficial to the parotid gland.

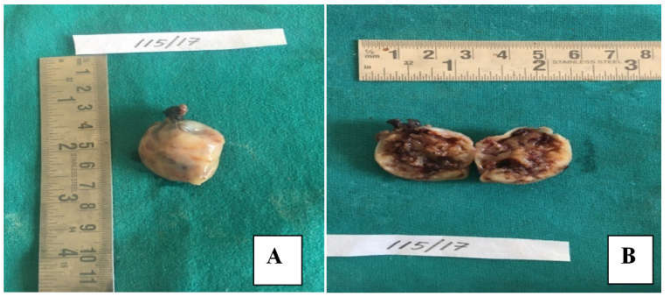


**Figure 2** The excised specimen grossly measuring about 3.8cmx3cm and cut surface showing whitish brown solid and cystic areas.

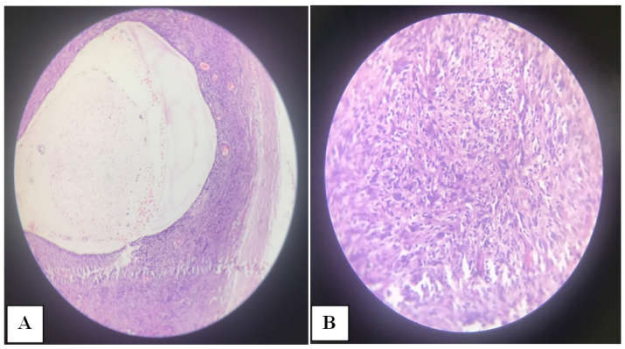


**Figure 3** Photomicrographs showing

A: lesional tissue surrounded by capsule, microcystic space lined by Schwann cells.
B: haphazardly arranged spindle cells in the loosely arranged connective tissue stroma giving Antoni B pattern.

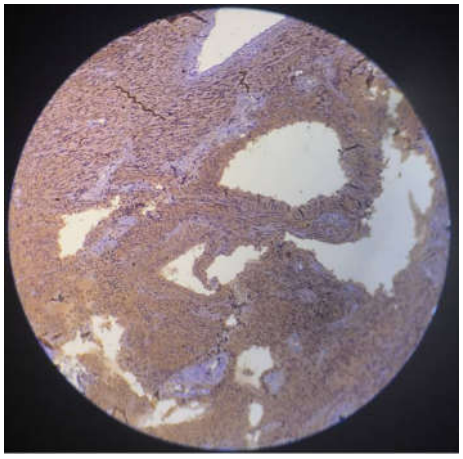

**Figure 4** Intense positive staining for S-100 protein in the spindle-shaped cells (Under 10x magnification).

***Histopathological Findings***: The biopsy showed a well circumscribed and encapsulated tumour mass surrounded by a thick fibrous capsule, composed of numerous spindle cells with large irregular wavy nucleus which are arranged haphazardly in the loosely textured matrix giving Antoni B pattern. The section also shows few microcystic spaces lined by Schwann cells. Presence of hyalinized blood vessels, mild chronic inflammatory cell infiltrates and degenerative changes are also evident in the section [Figure3]. Based on these histopathological findings the diagnosis of Ancient schwannoma was established. Immunohistochemistry for protein S-100 Schwann cells was strongly positive and the diagnosis of ancient schwannoma was confirmed [Figure 4].

## DISCUSSION

Schwannoma is an uncommon benign tumour in the oral cavity which is believed to arise from the Schwann cells. The entity was first described by Verocay in 1908. Most frequently found in the head and neck region and the flexor surfaces of upper and lower extremities, generally manifest as asymptomatic nodules. Schwannomas most often occur in third to sixth decades of life and seems to have a slight female predilection.[2] Schwannomas can be divided into central or intraosseous and peripheral lesions. Variants such as ancient schwannoma and plexiform intra osseous schwannoma are also described in the literature. Intraoral peripheral schwannomas are most frequently found in the tongue, followed by the palate, floor of the mouth, cheek mucosa and gums, whereas the intraosseous schwannomas are most often located in the mandible, though there have been descriptions of cases in the intramasseteric region, zygomatic arch or parotid zone, affecting the facial nerve. The definitive diagnosis of schwannoma is established only by the histopathological study of the lesion, with the presence of the two typical tissue patterns referred to as Antoni A and Antoni B. These patterns are characterized by the presence of cells with elongated or fusiform nuclei adopting a palisade distribution. Schwannomas shows positivity for the protein S-100(a specific neural tissue marker), Leu-7 and myelin basic protein (MBP) immunohistochemically to confirm the neural origin.[7]

The differential diagnosis of schwannoma includes tumors such as neuromas, neurofibromas, granular cell myoblastoma, neuroepitheliomas, fibromas or adenomas. Surgical excision of the tumour with preservation of the neighbouring structures is the most advocated treatment for schwannoma.[4]

Ancient schwannoma is a rare variant which is longstanding with degenerative changes. The entity was first described by Ackerman and Taylor in 1951. It rarely affects the head and neck area and intraoral lesions are even rare. Eversole and Howell in 1971 reported the first ancient schwannoma of head and neck region. Ancient schwannomas are benign, slow-growing tumours with rare malignant transformation but nuclear atypia and hyperchromasia may lead to perplex this tumour with malignancy and also misdiagnosed with myxoid neurofibroma and nerve sheath myxoma.[3] Till date 16 cases of ancient schwannomas of head and neck have been reported by various authors[Table 1].

Histopathologically schwannoma shows numerous spindle cells with eosinophilic cytoplasm and oval nucleus on hypercellular areas (Antoni A) myxoid and hypocellular areas (Antoni B), characteristic eosinophilic formations can be seen in the Antoni A areas, the Verocay bodies.

**Table 1** Review of literature of reported cases of ancient schwanomma

| Authors | Year | Age | Sex | Location |
|---|---|---|---|---|
| Eversole & Howell[11] | 1971 | 58 | F | Floor of the mouth and ventral tongue |
| Marks *et al*[17] | 1976 | 65 | F | Floor of the mouth(right) |
| McCoy *et al*[13] | 1983 | 36 | F | Maxillary vestibule (left) |
| Dayan *et al*[15] | 1989 | 52 | F | Maxillary vestibule (left) |
| Nakayama *et al*[14] | 1996 | 40 | F | Floor of mouth and ventral tongue |
| Ledesma *et al*[18] | 1999 | 21 | F | Floor of mouth and ventral tongue |
| Chen *et al*[16] | 2006 | 34 | M | Floor of the mouth (left) |
| Krishnaraj, Subhash raj *et al*[12] | 2008 | 18 | M | Posterior vestibule (left) |
| Shilpa B[5] | 2009 | 40 | F | Lower lip |
| Amirchagm *et al*[19] | 2010 | 14 | M | Gingiva |
| Shetty *et al*[10] | 2011 | 42 | M | Mandible(left) |
| Sayed *et al*[6] | 2012 | 46 | M | Submandibular region (right) |
| Muruganandhan *et al*[7] | 2013 | 22 | M | Right back tooth region |
| Wanjari *et al*[8] | 2013 | 68 | F | Lower jaw(left) |
| Balasubramaniam *et al*[9] | 2014 | 40 | F | Cheek (left) |
| Present case | 2017 | 65 | M | Preauricular region(right) |

The presence of extensive degenerative changes such as nuclear atypia, hemorrhagic foci or the formation of microcysts and calcification can be the result of a long standing nature of the lesions, and suggest the diagnosis of the "ancient" variety of the tumour. No malignant transformation has been described for the Ancient variant of Schwannoma and local recurrence is rare.[2]

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