Clinical Case Report

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# "Ancient" schwannoma of the submandibular gland

## A case report and literature review

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**Abstract**

**Rationale:** Schwannomas are solitary neurogenic tumors that arise from cells of the neural sheath. Ancient schwannoma is a relatively rare variant of schwannoma, characterized by increased cellularity and atypia. These cellular changes could be confusing and make the accurate pathologic diagnosis difficult.

**Patient concerns and diagnoses:** A 36-year-old man presented with painless swelling in left submandibular region for more than 2 years. The computed tomography confirmed a well-defined cystic lesion in the left submandibular space, which caused superior and posterior displacement of the left submandibular gland. Surgical excision was performed and the pathology confirmed the diagnosis of ancient schwannoma. To our knowledge, this patient is the second case of primary submandibular ancient schwannoma reported in the literatures.

**Interventions and outcomes:** The patient underwent tumor resection and postoperative recovery was uneventful. There were no nerve deficits after the operation. There was no recurrence within 1 year of follow-up.

**Lessons:** Schwannoma originated from the submandibular gland is extremely rare and only a few cases have been reported. Ancient schwannoma is an even more rare tumor. The increased cellularity and atypia of ancient schwannoma can resemble features of malignancy. Great care must be taken to make differential diagnosis with fibrosarcomas and malignant schwannoma.

**Keywords:** ancient schwannoma, cystic mass, submandibular gland

### 1. Introduction

Schwannomas are solitary neurogenic tumors that arise from cells of the neural sheath. Although approximately one-third of the reported cases occur in the head and neck region,[1] extracranial forms located in salivary glands are uncommon with most presenting in the parotid gland originating from a peripheral branch of the facial nerve.[2] Schwannoma originated from the submandibular gland is extremely rare and only a few cases have been reported.[3] Ancient schwannoma is an even more rare tumor.

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### 2. Case report

A 36-year-old male presented to our outpatient department with painless swelling in left submandibular region for more than 2 years. Swelling was insidious in onset and slowly progressive in nature. On physical examination, a single swelling of approximate size 4 x 3 cm was noted in the left submandibular region. Surface was smooth, well-defined lower margin but poorly defined upper margin, elastic in consistency, nontender, and slightly mobile. There was no palpable lymphadenopathy and all cranial nerve functions were within normal limit. The computed tomography confirmed a well-defined cystic lesion in the left submandibular space, which caused superior and posterior displacement of the left submandibular gland (Fig. 1).

We prepared the patient for surgery under general anesthesia after obtaining informed consent. On exploration, a cystic lesion attached to the submandibular gland was found and total excision of the lesion without injury to facial and hypoglossal nerves was performed carefully. Postoperative recovery was uneventful. Histopathologic examination showed a well-defined, encapsulated tumor consisting of spindle cells with Antoni type A and type B areas (Fig. 2A). A great portion of cystic change and nuclear atypia were also noted (Fig. 2B). Immunostaining consistent with schwannoma by strongly positive for S-100 protein (Fig. 2C). These findings confirm the diagnosis of ancient schwannoma. There was no recurrence within 1 year of follow-up.

### 3. Discussion

Schwannomas can be divided into assortment subtypes based on the histological findings, such as ancient, microcystic-reticular, epithelioid, cellular, psammomatous, and melanotic types.[4]

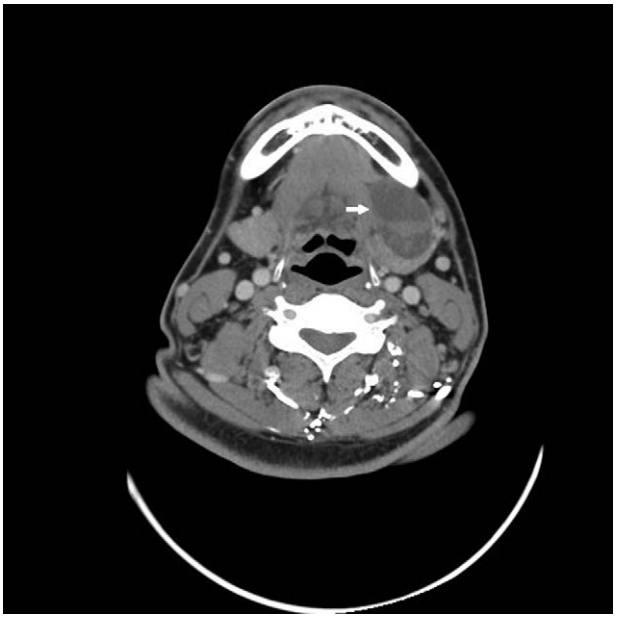

**Figure 1.** Axial view of the computed tomography revealed a well-defined cystic lesion in the left submandibular space (arrow), which caused superior and posterior displacement of the left submandibular gland.

Ancient schwannoma is a relatively rare variant. It was first described by Ackerman and Taylor in 1951,[5] characterized by increased cellularity and atypia. Additional histologic characteristics included areas of diffuse fibrosis and hyalinization with nuclear pleomorphism and hyperchromasia but without atypia.[5] It is believed to have these changes as the result of long-term tumor growth and aging. These cellular changes could be confusing and make the accurate pathologic diagnosis difficult. The increased cellularity and atypia can resemble features of malignancy. Great care must be taken to make differential diagnosis with fibrosarcomas and malignant schwannoma.

Up to date, there was only 1 case reported in the literature in 1996, which presented a 25-year-old female patient with a 6-month history, firm, tender, mobile, 3 x 2 cm mass in the left submandibular region.[6] The patient was treated with antimicrobial therapy initially with the impression of inflammatory disease. Surgical excision was considered because of the limited improvement. Herein, we reported the second case of primary submandibular ancient schwannoma with more typical clinical course of schwannoma. With schwannoma developing within a salivary gland, patients usually describe a painless and mobile mass in the region of the affected gland.[4] These benign tumors tend to push the associated nerve aside, which accounts for their usually asymptomatic presentation. Generally, patients seek medical advice until the tumors become large to make cosmetic concerns or cause compression to their adjacent structures.

In the present case, the cystic change of the tumor made the preoperative differential diagnosis more confusing. Because of the rarity of ancient schwannoma, the gold standard of diagnosis remains histopathological examination. Management of schwannomas in head and neck is complete surgical excision, which usually yields excellent results.[3]

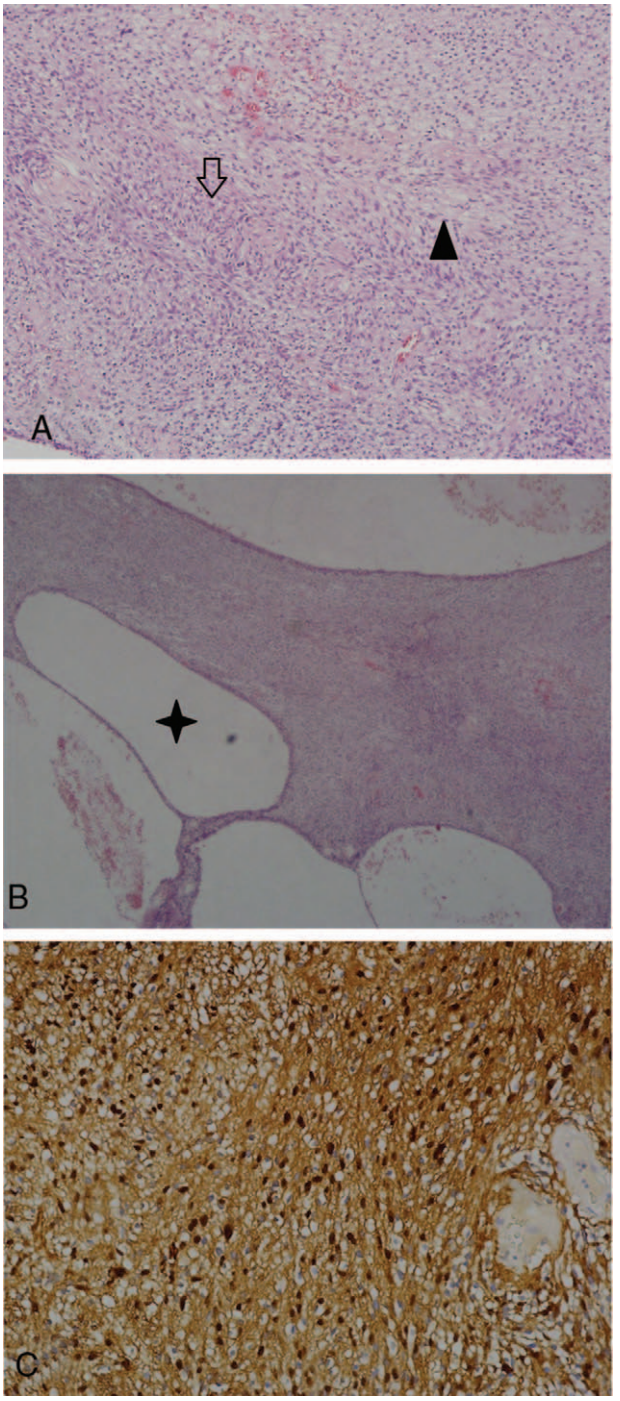


**Figure 2.** Histopathological examination of ancient schwannoma. (A) The tumor is composed of hypercellular Antoni A areas with classic spindle shaped cells (arrow) and hypocellular Antoni B areas (arrowhead) (x200). (B) Marked degenerative cystic change (asterisk) (x100). (C) Tumor cells present uniformly intense immunopositivity for S-100 protein (x200).

## 4. Conclusion

Primary submandibular ancient schwannoma is an extremely rare tumor with only 1 case reported in the literature. However, it must be included in the differential diagnosis, along with other primary salivary gland tumors. The characteristic features of ancient schwannoma are increased cellularity and atypia, which can resemble malignant tumors. Great care must be taken to make differential diagnosis with fibrosarcomas and malignant schwannoma.

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