CASE REPORT

The Submandibular Tumor: Unmasking a Rare Hypoglossal Schwannoma

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ABSTRACT

Hypoglossal schwannoma(HS) is a rare benign tumor of the twelfth cranial nerve, and its extracranial presentation can closely resemble a submandibular gland mass. We report a patient with a painless, gradually enlarging left submandibular swelling initially thought to arise from the submandibular gland. Imaging revealed an enlarged left submandibular gland containing a well-defined, heterogeneous lesion and fine-needle aspiration(FNAC) suggested pleomorphic adenoma(PA). Surgical excision via a transcervical approach was performed, and histopathological examination confirmed the diagnosis of schwannoma. Extracranial HS are exceedingly rare and may be misdiagnosed preoperatively. Complete surgical excision remains the treatment of choice, with nerve preservation crucial to prevent postoperative tongue weakness.

Keywords: Hypoglossal schwannoma, hypoglossal nerve, submandibular gland, pleomorphic adenoma

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INTRODUCTION

Schwannomas are benign, slow-growing neoplasms originating from Schwann cells, which form the myelin sheath of peripheral nerves[1,2]. They may arise from either peripheral or cranial nerve. They are typically encapsulated and exhibit an indolent course, making them clinically challenging to diagnose when arising in uncommon locations. Schwannomas arising from hypoglossal nerve are exceptionally rare, accounting for less than 5% of all schwannomas[1-3]. The majority of documented cases involve the intracranial or skull base region, while purely extracranial hypoglossal schwannomas are extremely rare[4,5]. Due to their anatomical course, extracranial hypoglossal schwannomas often present as neck masses, and closely mimic parapharyngeal or submandibular gland lesions[6-8]. This overlap in anatomical location and clinical features often complicates preoperative diagnosis. Therefore, heightened awareness of this entity is essential for otorhinolaryngologists and head and neck surgeons to ensure accurate diagnosis and management.

CASE SUMMARY

A 35-year-old lady with no medical illness, presented with a left submandibular swelling which was progressively increasing in size over the past five years. The swelling began as small, painless mass, with no associated history of trauma, infection, difficulty in chewing or swallowing, or systemic symptoms. On physical examination, a firm, oval, and smooth swelling measuring approximately 3 × 4 cm was noted in

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the left submandibular region. It extended from the angle of the mandible to the hyoid bone, medially reaching the midline and laterally up to the anterior border of the sternocleidomastoid muscle. The mass was non-tender, mobile, and non-fluctuant, with normal overlying skin and no evidence of regional lymphadenopathy (Fig. 1). Intraoral assessment revealed a ballotable mass, a patent submandibular duct, and absence of calculi. Nasal endoscopy findings were unremarkable. The initial fine needle aspiration cytology (FNAC) revealed pleomorphic adenoma (PA). Ultrasound neck showed a well-circumscribed hypoechoic lesion with cystic components (2.2 × 2.8 × 3.1 cm) arising from the left submandibular gland. The patient was scheduled for a left submandibulectomy; however, the procedure was postponed for approximately one year due to personal circumstances. At follow-up, the swelling continued to enlarge without associated pain or obstructive symptoms. Repeat FNAC indicated a salivary gland neoplasm of uncertain malignant potential (SUMP), with differential diagnoses including schwannoma, cellular pleomorphic adenoma, and myoepithelioma.

Contrast-enhanced computed tomography (CECT) neck revealed an enlarged left submandibular gland containing a well-defined, heterogeneous lesion (3.4 × 3.1 × 4.5 cm) with multiple subcentimeter bilateral cervical lymph nodes. The impression favored a benign lesion, though malignancy could not be excluded. Intraoperatively, a lobulated, firm, well-encapsulated, whitish mass measuring 3 × 5 cm was noted arising from the deep lobe of the submandibular gland, while the superficial lobe appears normal (fig. 2). Both superficial and deep lobes were excised. The facial vessels were carefully identified and managed. The lingual nerve was preserved, and neither the hypoglossal and marginal mandibular nerves were identified intraoperatively (Fig. 3). Postoperatively, the patient developed immediate left marginal mandibular nerve paresis and left hypoglossal nerve palsy. At one-month follow-up, tongue deviation to the left persisted. The histopathological examination revealed a well-encapsulated tumor exhibiting biphasic Antoni A and Antoni B areas with Verocay body formation (Fig. 4 & 5). The spindle-shaped tumor cells showed diffuse S-100 positivity, confirming the diagnosis of schwannoma (Fig. 6).

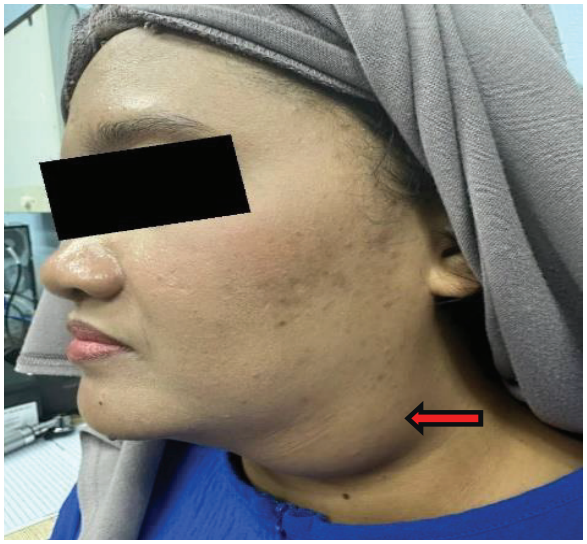

Figure 1 : Preoperative view of the left lateral neck showed a well defined and firm lesion in the left submandibular region (red arrow).

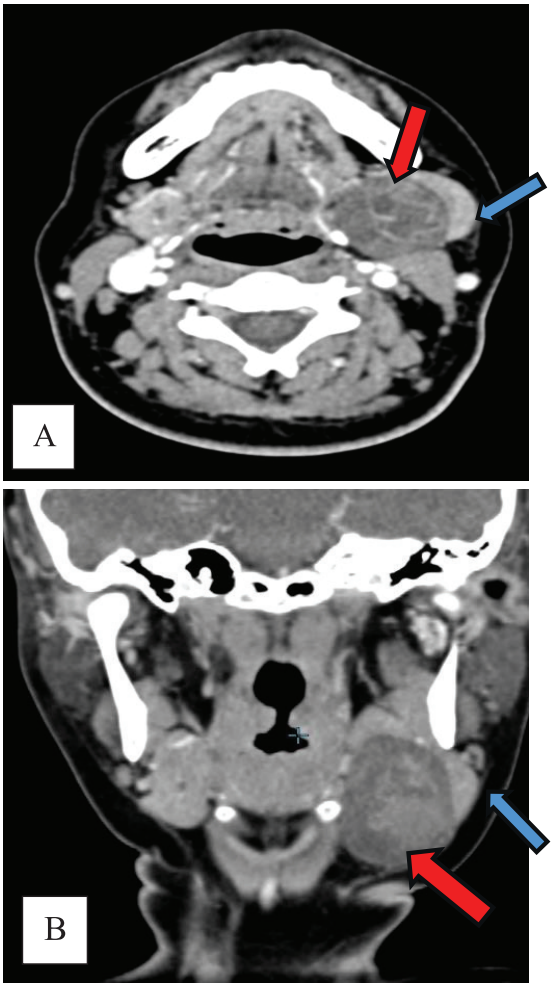

Figure 2: Contrast-enhanced CT of head and neck (A: axial view, B: coronal view): red arrow showing a well-defined heterogenous lesion measuring 3.4 cm × 3.1 cm × 4.5 cm arising from left submandibular gland (blue arrow).

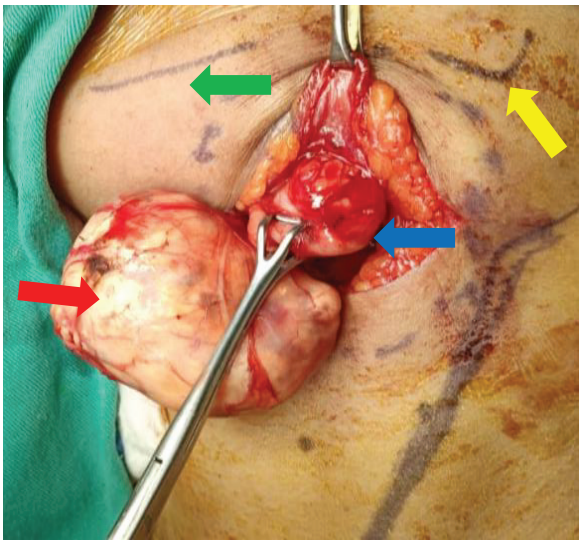

Figure 3: Intraoperative picture of left neck showing swelling of hypoglossal nerve adjacent submandibular gland (red arrow), reflected normal left submandibular gland (blue arrow), body of mandible (green arrow), tip of mastoid (yellow arrow).

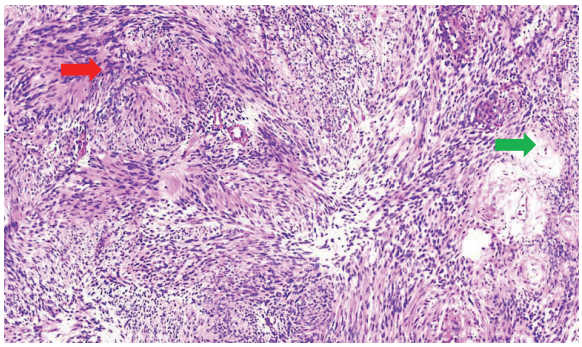

Figure 4: Photomicrograph showing a biphasic spindle cell tumor with alternating Antoni A (cellular) (red arrow) and Antoni B (loosely textured) areas (green arrow), consistent with a benign schwannoma (H&E; × 200).

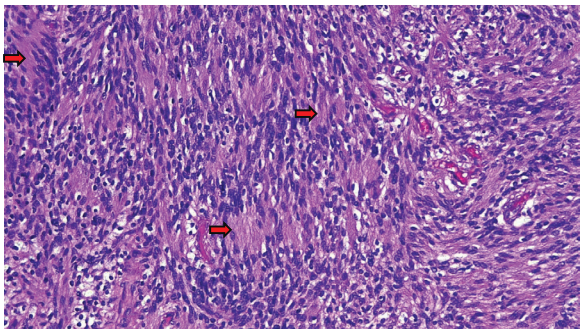

Figure 5: High-power view demonstrates a hypercellular proliferation of bland spindle cells exhibiting nuclear palisading (Verocay bodies), (red arrow)(H&E; × 200).

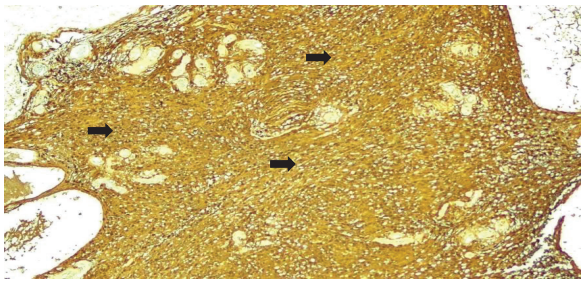

Figure 6: Immunohistochemical staining for S-100 shows diffuse strong positivity in the tumor cells (black arrow), supporting Schwann cell differentiation (S-100; × 200).

DISCUSSION

Neurilemmomas, commonly referred to as schwannomas, are benign, encapsulated tumors of the nerve sheath originating from Schwann cells, which produce the myelin sheath of peripheral nerves[1,2,4]. These lesions are typically slow-growing and well-circumscribed, and they may arise along the course of peripheral, cranial, or autonomic nerves[1-6]. Hypoglossal nerve involvement is exceedingly rare[1-8], accounting for approximately 5% of all non-vestibular head and neck schwannomas[1,5]. Although 25%–45% of schwannomas occur in the head and neck region, localization within the submandibular area is distinctly uncommon[3,5,7,8]. Extracranial hypoglossal schwannomas that mimic a benign submandibular gland mass are exceptionally rare[4,6]. These tumors are most frequently observed in middle-aged individuals, with a female predominance[1,3,4]. The rarity of submandibular schwannoma frequently results in diagnostic uncertainty, as its clinical presentation closely mimics more common salivary gland lesions. Patients with hypoglossal schwannoma typically exhibit a painless, slowly enlarging submandibular swelling, often without neurological deficits[1-4]. The absence of sensory or motor dysfunction aligns with the indolent, non-invasive nature of schwannomas. These tumors are generally solitary, encapsulated, and slow-growing, appearing attached to or surrounded by a nerve[5]. When a submandibular schwannoma exceeds a certain size approximately 2 cm or more, it may produce symptoms such as neck discomfort, pain, or paresthesia[8].

Radiological assessment is essential for diagnosis and preoperative planning, although imaging findings are often nonspecific[3,4].

The radiologic appearance of hypoglossal schwannomas frequently overlaps with that of submandibular gland tumors or paragangliomas[3,5]. Ultrasonography typically demonstrates a well-circumscribed, heterogeneous mass[2]. CECT neck may reveal a well-encapsulated soft tissue lesion with homogeneous or heterogeneous enhancement[1,4,8]. Magnetic resonance imaging (MRI) offers superior soft tissue contrast and may display the characteristic "target sign"[3]. This sign consists of a central low-intensity area (fibrous component) surrounded by a hyperintense rim (myxomatous component) on T2-weighted images, corresponding to dense Antoni A and loose Antoni B regions[2,6,7]. In many reported cases, imaging results have been inconclusive, often favoring a benign submandibular gland neoplasm rather than a neurogenic tumor[1,2,4,6].

FNAC although routinely employed for evaluating submandibular masses, offers limited diagnostic accuracy in schwannomas[1-8]. FNAC is generally unreliable for identifying these tumors and often produces inconclusive or misleading results. Several authors have reported that FNAC frequently suggests a benign salivary gland lesion, underscoring the limitations of cytology in such cases[1-8]. Histologically, submandibular schwannomas exhibit a biphasic growth pattern characterized by alternating Antoni A and Antoni B areas. Antoni A regions are hypercellular, composed of spindle-shaped Schwann cells arranged in compact fascicles with nuclear palisading and Verocay body formation, whereas Antoni B regions are hypocellular, featuring loosely textured, myxoid stroma with microcystic and vascular changes[1-8]. A strong and diffuse S-100 immunoreactivity is a hallmark feature supporting Schwann cell differentiation[1-8]. Immunohistochemically, diffuse and strong S-100 expression supports Schwann cell differentiation[1-8]. In contrast, neurofibroma typically lacks encapsulation and the distinctive Antoni pattern, perineurioma exhibits epithelial membrane antigen (EMA) positivity with absent or weak S-100 staining, and malignant peripheral nerve sheath tumor (MPNST) demonstrates cytologic atypia, increased mitotic activity, and areas of necrosis, features that aid in differentiating these lesions from schwannoma[9].

Complete surgical excision with preservation of the involved nerve, whenever feasible, remains the primary treatment for submandibular schwannoma[1-8]. As the tumour is encapsulated and typically displaces rather than infiltrates the parent nerve, meticulous dissection often permits enucleation without significant neurological compromise[1-7]. Postoperative recovery is usually uneventful, and recurrence is exceedingly rare[1-8]. Although malignant transformation of schwannoma has been documented in the literature, it remains exceedingly rare. The long-term prognosis following complete surgical excision is excellent. Regular postoperative follow-up is advised to monitor for recurrence or delayed nerve dysfunction.

CONCLUSION

Submandibular schwannoma is a rare clinical entity that poses a diagnostic challenge due to its resemblance to benign submandibular gland lesions. Preoperative diagnosis is difficult, as clinical, radiological, and cytological findings are often inconclusive. Histopathological and immunohistochemical evaluation remains the cornerstone of diagnosis. Awareness of this rare presentation among clinicians, radiologists, and pathologists is essential to include schwannoma in the differential diagnosis of submandibular swellings. Early identification and precise surgical management ensure excellent outcomes with minimal morbidity.

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Authors' contribution: All authors were involved in the patient selection and clinical data collection, core biopsy collection, cytopathology and histopathological examination as well as manuscript preparation, review, editing and final submission.

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