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A Rare Case of Submandibular Schwannoma

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KEYWORDS

Submandibular swelling, Schwannoma, Benign tumor, Peripheral nerve tumors.

ABSTRACT:

Introduction: Schwannoma is a benign tumour that typically grows slowly and is usually enclosed within a capsule. They arise from the Schwann cells of myelinated nerve fibres. These tumours can sometimes resemble primary or metastatic cancers in the head and neck area. Schwannomas in the salivary glands are especially uncommon and if present is usually found in the parotid gland originating from a peripheral branch of the facial nerve..

Case presentation: Here we report a case of submandibular schwannoma in an 85 year old male who presented with swelling in the jaw for one year. The mass was discrete and well demonstrated on MRI. Total excision of the swelling along with submandibular gland was done. Post-operatively there was complete resolution of symptoms with no cranial nerve deficits. The histopathological features have also been discussed in this study.

Conclusions: Most common differential diagnoses for submandibular swelling includes submandibular sialadenitis, sialolithiasis, pleomorphic adenoma of submandibular gland and lymphadenitis. But rare presentations such as submandibular schwannoma must also be kept in mind as total excision of the mass ensures complete recovery.

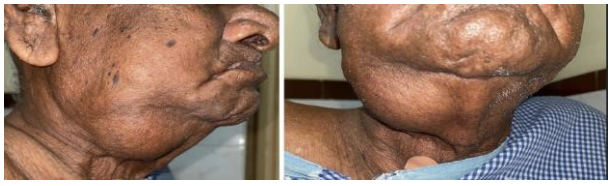
1. Introduction

Schwannomas are benign tumors with well-defined margins originating from Schwann cells of myelinated nerve fibers[1]. Nearly 45 percent occur in small peripheral nerves in caudal end and flexor surfaces of the limbs[2]. These are usually pain-free [3]. They are slow growing and usually have a delayed presentation[4]. The nerve from which the tumor is arising may not be found in about 40 percent of these tumors [5]. Here, we present a case of a schwannoma of the submandibular gland in an 85-year-old man.

2. Case presentation :

An 85-year-old male was admitted with complaints of swelling in right lower jaw for the past 1 year.

Initially, the swelling was small and gradually progressed to attain the current size of 6 x 5cms. It was initially painless, but as the size gradually progressed, it was associated with intermittent dull aching type of pain which aggravated while chewing. He gave no history of difficulty in swallowing, tooth ache, increased sensitivity to cold or hot substances, weight loss, irradiation to head and neck region. On examination, a swelling of size approximately 6x5cms noted in the right submandibular region - globular in shape, smooth surface. The swelling was soft to firm in consistency with well-defined borders. It was superiorly extending from the inferior ramus of the mandible, posteriorly to the anterior border of sternocleidomastoid muscle, anteriorly up to midline and inferiorly up to the level of superior border of thyroid cartilage. Skin over the swelling was pinchable. [Table/Figure 1] The swelling was bi-digitally palpable and mobile. Cervical group of lymph nodes not palpable.



[Table/Figure 1] Clinical presentation of the patient

Ultrasound examination of the neck showed a wellcircumscribed solid lesion with cystic areas in the

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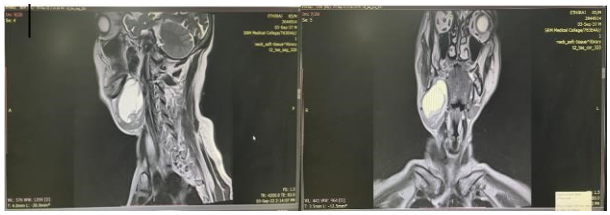
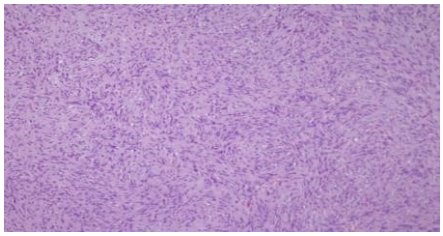
right submandibular gland. MRI Neck revealed :A welldefined multiloculated cystic lesion in the right submandibular gland region of size 4.2x4x0.4cms was noted. [Table/Figure 2]



[Table/Figure 2] MRI images showing the welldefined lesion

Fine needle aspiration cytology was done under ultrasound guidance which showed benign cystic lesion possibility of lymphoepithelial cyst of the right submandibular gland. The patient was planned for an excision biopsy under general anesthesia. On exploration of the submandibular region, a swelling attached to the submandibular gland was found and after careful dissection and identification of the adjacent anatomical structures, we proceeded with total excision of the swelling along with the submandibular gland. [Table/Figure 3]. Macroscopically the lesion was 5x4x2cms, capsulated, yellowish in color. The specimen was sent for histopathological examination. The wound was carefully closed in layers after securing hemostasis. Post-operative period was uneventful and wound healed well. Microscopic examination showed cells arranged in intersecting whorls and sheets with alternating hypercellular and hypocellular areas, lined by elongated spindle shaped nuclei, bland chromatin, absent atypia, moderate cytoplasm on a background of cystic degenerations, lymphoid aggregates and myxoid areas – features suggestive of schwannoma with cystic and myxoid degeneration in the right submandibular salivary gland. The patient was followed up once every 2 months for the next one year. He had no complaints and was symptom free.



[Table/Figure 4] Microscopic examination showed the capsulated benign tumor was composed of cells arranged in intersecting whorls and sheets with alternating hypercellular and hypocellular areas, lined by elongated spindle shaped nuclei

3. Discussion :

Schwannoma, also known as neurilemmoma, is a benign tumor that originates from Schwann cells [6]. Typically solitary, these tumors grow from the peripheral nerve sheath and are often connected by a stalk. They are welldefined and can be easily separated from nerve bundles, unlike neurofibromas, which grow within the nerves. Symptoms such as pain, tingling sensations, or numbness may occur, suggesting a compressive neuropathy or possible malignancy [2,6].

Schwannomas commonly arise from sensory nerve roots, with intracranial tumors frequently found on the vestibular branch of the eighth cranial nerve. Less frequently, they can develop from the trigeminal nerve or glossopharyngeal nerve, or in cases of neurofibromatosis type 2 (NF2), from lower cranial nerves [2,6]. Involvement of motor roots or the sympathetic chain is rare, and schwannomas seldom affect brain or spinal cord tissue. While many schwannomas are asymptomatic, some can cause pain or sensory disturbances, typically responding well to surgical removal. Unlike neurofibromas, they rarely undergo malignant transformation, which can sporadically occur or be associated with conditions like schwannomatosis or neurofibromatosis, or following therapeutic irradiation [7-15].

Diagnostic investigations include computed tomography (CT), magnetic resonance imaging (MRI), ultrasound, and fine-needle aspiration (FNA). MRI is preferred for evaluating tumor extent and correlates closely with surgical findings [16]. Biswas et al., in a decade-long study of extracranial head and neck schwannomas, found that only 6 percent of cases could be diagnosed preoperatively using clinical evaluation, CT, MRI, and FNAC [17]. Definitive diagnosis depends on histopathological examination of surgically excised specimens [17-19].

Due to its resistance to radiotherapy, surgical excision remains the mainstay of treatment [20-23]. Kang et al. reported that more than 50% of surgically managed cases experience postoperative neural deficits, primarily due to iatrogenic injury [19].

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Schwannomas typically present in middle-aged individuals, though it was identified in an 85-year-old male in this case. Diagnosis was confirmed postoperatively through histopathological examination, as schwannomas arising from the submandibular gland are rare. During surgery, the hypoglossal nerve, lingual nerve, and marginal mandibular branch of the facial nerve were identified and spared, resulting in no postoperative neurological deficits. There is speculation that the patient's schwannoma may have originated from an autonomic nerve supplying the gland.

4. Conclusion :

While schwannomas are relatively frequent in the head and neck region, their occurrence in the submandibular gland is rarely documented in the literature. Highlighting this diagnosis aims to emphasize the consideration of such rare possibility when encountering nodular lesions in the submandibular area as total excision of the mass ensures complete recovery as seen in our case.

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