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CASE REPORT

Schwannomas of the head and neck: a case series

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Abstract Schwannomas are relatively slow growing benign tumors of the nerve sheath with a well developed capsule. They can reach a considerable size. Typically, symptoms of schwannomas are based on the affected nerve. We present four cases of this tumor that occurred in the head and neck. The differential diagnosis of small painless nodules in head and neck must include schwannomas as it is likely that nerve sheath neoplasms are more common than previously reported. However, due to benign nature and low recurrence prognosis is excellent.

Keywords Neurilemoma · Nerve sheath · Antoni A variants · Antoni B variants

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Introduction

A schwann cell is a type of glial cell of the peripheral nervous system that helps separate and insulate nerve cells. A schwannoma is a tumor of these cells that form the nerve sheath. Formerly known as neurilemoma, it is slow-growing and nearly always benign [1]. Schwannoma has a 25–48% predilection for nerves of the head and neck [2] hence it has a predilection for sites in the vicinity of nerves: in the parotid gland along facial nerve, in the neck along the cervical sympathetic chain, in the infratemporal fossa along mandibular division of trigeminal nerve, palate, buccal mucosa and parapharyngeal space. The commonest oral site is said to be the tongue [3]. The tumor occurs more often in females. An intraoral schwannoma is a smooth, submucosal swelling bearing an uncanny resemblance to mucocele, fibroepithelial polyp, fibroma, lipoma or benign salivary gland tumor [4]. Usually a soft tissue neoplasm, it is rarely intrabony too.

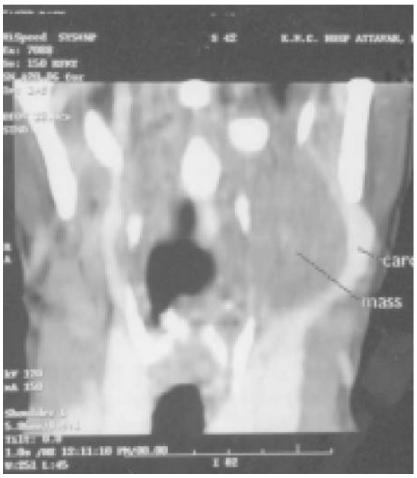
Case reports

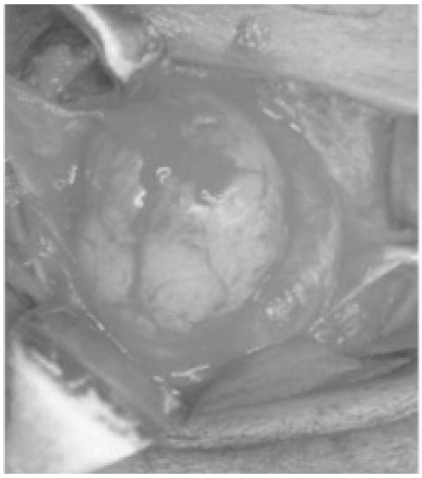
Case 1: A 40-year-old healthy female with no co-morbidities was referred to us for evaluation and treatment of a mobile, pulsatile swelling in the left side of the neck. According to the patient, the swelling had started growing gradually without any significant symptoms over a period of 8 months. There were no paresthesias, no muscular weakness and no dysphagia or pain. When we examined we discovered a solitary swelling that was firm to palpation, circumscribed, non-tender and mobile. A transmitted pulsation was also noted. As fine needle aspiration cytology was inconclusive, a CT scan of the neck was obtained. The swelling was a well-defined mass isodense in relation to surrounding soft tissue, close to the external carotid artery and displacing it outwards. The close proximity to a major artery explained the visible pulsatility of the lump. It appeared to be well-encapsulated and we decided to excise the tumor under general anesthesia. Access was via a submandibular approach, and the tumor separated without difficulty as there was no infiltration into adjacent vessels. Histopathology confirmed a diagnosis of schwannoma. The patient recovered uneventfully.

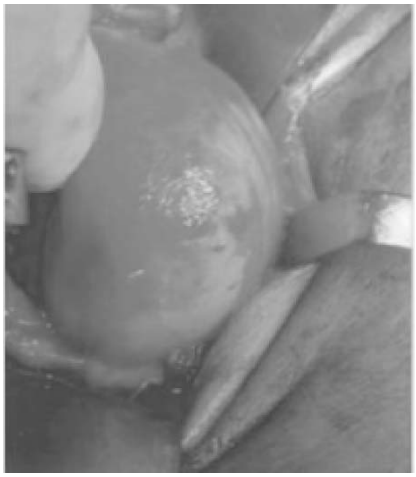
Case 2: A 46-year-old female reported to us with a lump measuring 5x5 cm in the left side of the neck. On examination we noted a solitary, circumscribed swelling which was firm in consistency, non-tender and mobile in the transverse plane. Aspiration cytology did not provide diagnostic information. A CT scan revealed displacement of the internal jugular vein by a homogenous, circumscribed lesion. The patient was subsequently taken up for excision of the lesion under general anesthesia. A submandibular incision was placed and after dissection through the deep cervical fascia an encapsulated mass was encountered. The tumor was carefully separated from adjacent structures with a combination of blunt and sharp dissection and the specimen submitted for histopathology. A vacuum drain was inserted and the surgical wound was closed in layers. The oral pathologist reported the excised specimen as schwannoma. The patient is on follow-up and is free of tumor recurrence 5 years later.

Case 3: A 15-year-old female was brought to our centre by her parents for a swelling in the left soft palate that had been discovered only recently by the patient. The swelling was present at the junction of the soft and hard palates and was painless and non-tender. There were no sensory deficits and overlying mucosa was normal, without erythematous or ulcerative changes. FNAC was attempted but was negative for aspirate. A decision to obtain CT scan was made to evaluate whether the mass had involved nerve or destroyed bone: the ominous signs of malignancy. Axial sections demonstrated a circumscribed, isodense, homogenous lesion on the left posterior palate with no evidence of infiltration into the greater palatine foramina. With a provisional diagnosis of a minor salivary gland tumor, under general anesthesia the lesion was excised with a margin of normal tissue upto but not including palatal bone. We sutured a collagen sheet to the defect and the wound healed secondarily by granulation. The specimen was submitted for histopathology

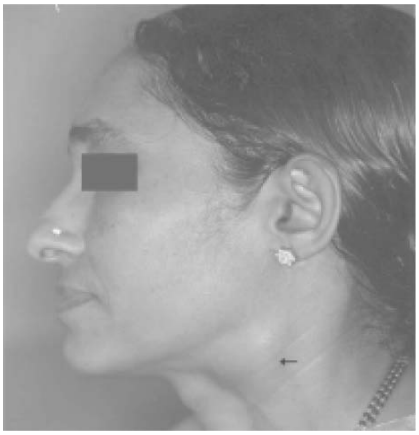
Case - 1

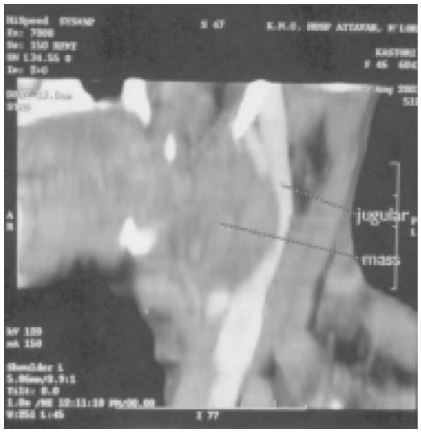


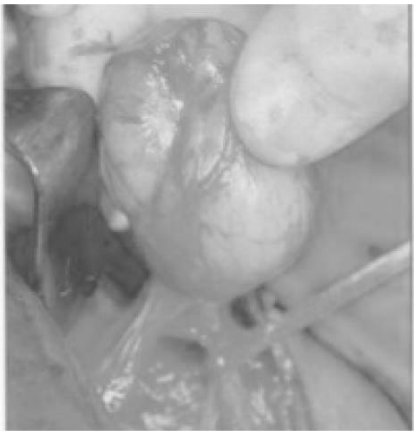




Case - 2







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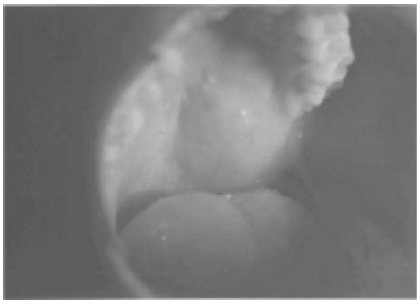
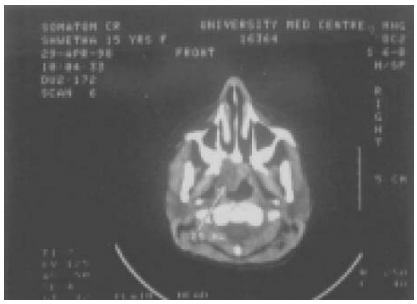




which revealed a schwannoma. The patient had an uneventful recovery and a decade later is free from recurrence.

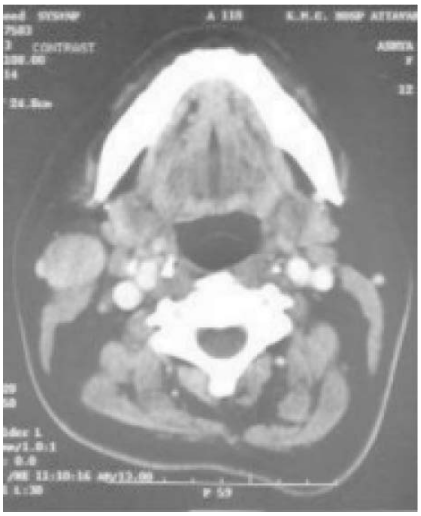
Case 4: A 50-year-old diabetic male presented with a lump of the right preauricular region of considerable duration and insidious onset. The swelling had elevated the ipsilateral earlobe, was non-tender and mobile. There was no apparent facial weakness and with a provisional diagnosis of pleomorphic adenoma of the parotid gland he was advised surgical excision of the same. Under general anesthesia, the operation was begun with a modified Blair's preauricular incision and a layer-by-layer dissection was carried out. Once the lower buccal branch of the facial nerve was identified careful dissection was done in a retrograde fashion, gently separating tumor from nerve. The encapsulated specimen was then sent for histopathology and was diagnosed to be a schwannoma of the facial nerve. Postoperatively, the patient had grade 2 palsy of the facial nerve with noticeable weakness of the depressor muscles of lower lip on the right side. Ultrasound therapy and Transcutaneous Electrical Nerve Stimulation (TENS) were provided by the physiotherapist and the patient made a complete recovery with no residual neurological deficit at the end of 4 weeks.

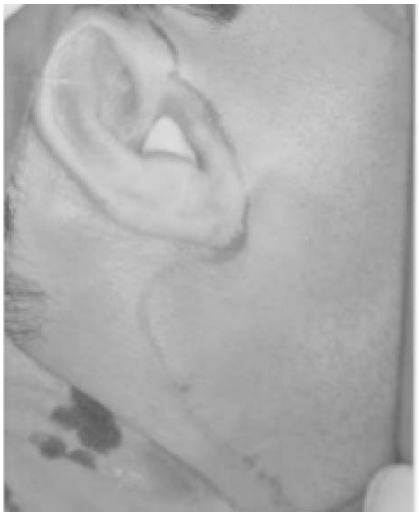
Discussion

Schwannomas are said to occur most frequently in the 25–55 year age group, but can occur at any age. Although said to occur more frequently in women than in men, there is no definite gender predilection. Clinically, these benign tumors are easily mistaken for other entities such as lipoma [4] and pleomorphic adenoma [5] on account of their slow growth and absence of neural symptoms.

These benign tumors of the nerve sheath are well encapsulated and can reach considerable size since their slow rate of growth over several years goes unnoticed by patients especially when intraoral. When the tumor is located intraorally, the lesion typically presents as a slowly enlarging [5], painless, submucosal nodule which is slightly mobile beneath the surface, rarely enlarging beyond 2 cm in greatest

Case - 4





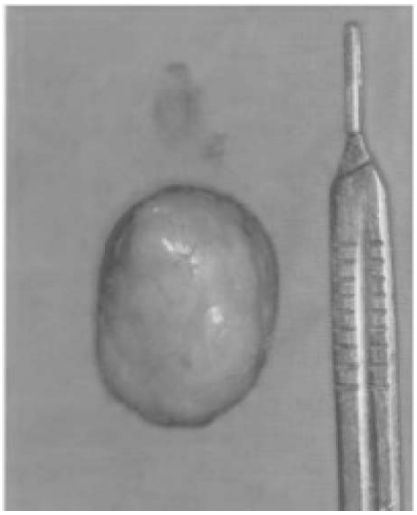


Table 1 Differential diagnosis of swellings of submandibular fossa and lateral neck [12]

Lesion	Characteristic feature
Metastatic lymph node	Shorter duration; indurated; fixity
Lymphangitis due to infectious mononucleosis	Malaise and fatigue; sore throat; shorter duration than schwannoma; tender
Lipoma	Soft consistency
Pleomorphic adenoma	Deceptively similar to schwannoma
Carotid body tumor	Pulsatile but immobile
Branchial cleft cyst	Soft and compressible; young adults
Reactive lymphadenopathy	Tender on palpation

Table 2 Differential diagnosis of swellings of the hard and soft palate

Lesion	Characteristic feature
Mucous retention cyst	Bluish hue; mucinous aspirate; fluctuant
Lipoma	Soft consistency
Pleomorphic adenoma	Deceptively similar to schwannoma
Minor salivary gland tumor	Neural involvement; rapid growth
Palatal abscess	Associated infected tooth; pain; tenderness
Fibroma/papilloma	Lesion corresponds to ill-fitting denture
Squamous cell carcinoma	Ulceroproliferative growth with irregular surface
Lymphoma	Boggy, edematous consistency

dimension. The tongue is the most frequent site of occurrence, but other oral sites are also susceptible and the palate is the second most common oral subsite to be involved [6]. Schwannomas are actually a proliferation of the nerve sheath with the nerve of origin remaining easily identifiable and hence readily separable from the tumor by careful dissection during surgical removal.

Schwannomas are relatively slow growing and push the nerve aside, only occasionally producing pain or paresthesia. Typically, presenting symptoms of schwannomas relate to the affected nerve [7]. Hence, trigeminal nerve schwannomas can present with facial pain and/or atrophy of the muscles of mastication; seventh cranial nerve lesions might manifest as facial weakness while lesions of the inferior alveolar nerve may cause pain/paresthesia. However, these neural symptoms are the exception rather than the rule. These are benign tumors and almost never undergo malignant transformation [8]. Therefore, recurrence is not expected. Even in the rare event of a recurrence and repeated surgical intervention, schwannoma does not undergo malignant transformation unlike a neurofibroma [6].

In the evaluation of such masses, Magnetic Resonance Imaging (MRI) scans can provide useful information regarding not only the location, but also the nature of the lesion. MRI is of some use in eliminating certain differential diagnoses namely, rhabdomyosarcoma, neurofibroma, vascular lesions such as hemangioma, lesions with semisolid contents such as dermoid cyst and malignant salivary gland tumors. However, to rule out bone resorption due to a malignant tumor, Computed Tomography (CT) scans are useful. Although soft tissue schwannomas have no useful radiographic findings [9], in the rare case of intrabony (central) schwannoma the role of plain film radiography in verifying location and determining extent should be appreciated.

In our series of 4 cases, site of the lesion, history and clinical features were important in arriving at a provisional diagnosis. Though not exhaustive, a useful guide to the formulation of a clinical differential diagnosis follows (Table 1 and 2).

Histopathologically, schwannomas are unilobular masses surrounded by a capsule of epineurium and residual nerve fibers, often with the edge of the neoplasm attached to the peripheral nerve. The substance of the

tumor is composed of a mixture of two cellular patterns: Antoni A and Antoni B [10]. Antoni A areas are composed of compact spindle cells with twisted nuclei arranged in bundles or fascicles. In highly differentiated areas there may be nuclear palisading and formation of Verocay bodies, which are formed by alignment of two rows of nuclei and cell processes which assume oval shape. A low power microscopic view of this tissue is reminiscent of an aerial view of soldiers aligned against each other across multiple battlefields.

Antoni B variant is less cellular and less organized, representing degenerated Antoni A areas composed of haphazardly arranged spindle or oval cells within a myxoid, loosely-textured, hypocellular matrix punctuated by micro cysts, inflammatory cells and delicate collagen fibers. However, in spite of nuclear pleomorphism and atypia, schwannomas are benign. It is vital that the surgeon and pathologist not mistake these alterations for sarcoma. The so-called ancient neurilemoma is benign and must be differentiated from neurofibrosarcoma and malignant neurilemoma [11].

In view of the clinical presentation and histopathological features, it is important that surgeon does not mistake this lesion for malignancy. In contrast to malignant tumors, schwannoma is treated by conservative surgical excision. Dissecting the tumor off the nerve is usually possible with minimal disturbance to nerve function [12], as we encountered in our fourth case. Malignant transformation in treated lesions has been reported but is very uncommon [10]. Patients with multiple neural tumors should be evaluated for von Recklinghausen's neurofibromatosis.

Conclusion

An important conclusion that has evolved from this case series is that the differential diagnosis of small painless nodules in head and neck must include schwannomas. It is likely that nerve sheath neoplasms are more common than previously reported. Although slow-growing, the schwannoma pushes the nerve aside. When this happens in an intrabony lesion associated with the inferior alveolar nerve, it will produce paresthesia; in the preauricular region it may produce facial weakness, thus raising a suspicion of malignancy. However, schwannomas are entirely benign. Prognosis is excellent as the tumor is benign, and recurrence is nearly unknown, so it is possible–and indeed recommended – to preserve nerve integrity with careful dissection. An important differential diagnosis of swellings of the head and neck including the oral cavity, with excellent prognosis, is the schwannoma.

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