Case Report

Neurilemmoma Masquerading as Tonsillitis: A Case Report

Pathology Section

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ABSTRACT

Neurilemmoma, also known as schwannoma, is a benign neoplasm which originates from the Schwann cells which cover the myelinated nerve fibres. The most common site is the tongue while palatal schwannomas are even rarer. A case of schwannoma which was clinically diagnosed as a tonsillar mass (tonsillar hyperplasia) which caused obstructive symptoms in a young male child has been reported here. The patient presented with repeated episodes of sore throat, fever and difficulty in and pain on swallowing, of 6 months duration, along with a recent episode of high grade fever, which subsided with antibiotics.

On local examination, it was observed that there was bilateral hypertrophy of the tonsils with congested pillars and that the right tonsil appeared to be more enlarged in size than the left one. The right side anterior pillar could not be separately differentiated from the tonsil. The tonsilolingual sulcus on the right side was obliterated and it was assumed that the palatine tonsil was enlarged and intermingled with the lingual tonsil. Bilateral tonsillectomy was perfomed and the tissue was sent for histopathological examination. The right sided mass turned out to be Neurilemmoma.

This case was worth reporting as it highlighted the importance of including schwanomma as an important differential diagnosis whenever a treating physician encountered a tonsillar mass or a mass of unexplained aetiology in the oral cavity, which caused a diagnostic dilemma. A detailed clinical history, physical examination, cytology and radiological assessment helps in differentiating schwanomma in such settings.

Key Words: Neurilemmoma, Schwannoma, Tonsillar mass

INTRODUCTION

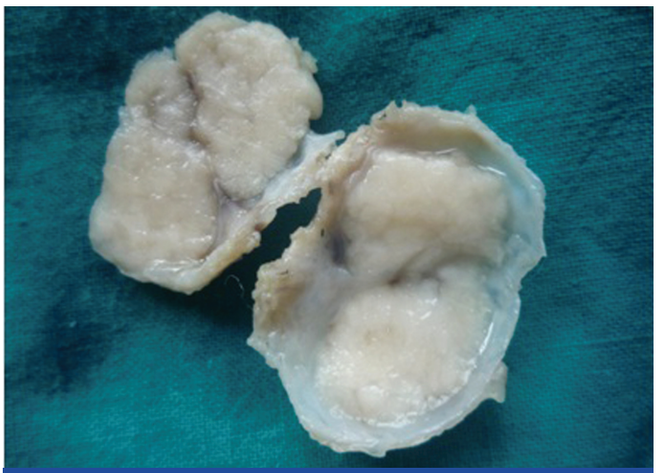
Neurilemmoma, also called as schwannoma or neurilimoma, is a benign neoplasm which originates from the peripheral neural sheath of any myelinated nerve [1]. Schwannoma arises from the Schwann cells which cover the myelinated nerve fibres [2]. Only 1% of the schwannomas are intraoral in location [3, 4], with the tongue being the commonest site, while palatal schwannomas are even rarer [5]. By physical examination, sometimes it is very difficult to differentiate a schwanomma from other reactive and neoplastic swellings; which can sometimes be misleading for a treating physician. Ancient Schwannomas with the longest duration of 18 years in the floor of the mouth and of 20 years in the vestibule of the left maxilla have also been seen [6, 7]. A case of schwannoma which was clinically diagnosed as tonsillar hyperplasia, which caused obstructive symptoms in a young male child, has been reported here.

CASE REPORT

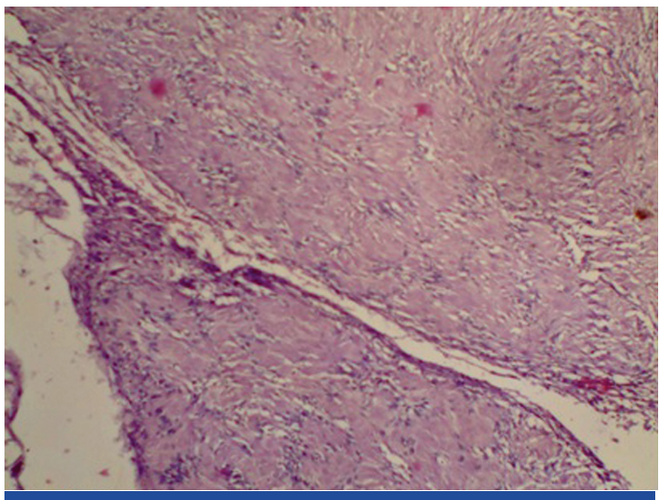
A 14 year old male presented to the outpatients department of the Otolaryngology unit of a tertiary care centre with the chief complaint of repeated sore throat for the past 5 years, along with fever once every two months. He also reported difficulty in and pain on swallowing for the past 6 months, with a recent episode of high grade fever, with a swelling on the neck on the right side, which subsided with antibiotics.

On local examination, it was found that there was bilateral hypertrophy of the tonsils, with congested pillars. The right tonsil appeared to be more enlarged in size than the left one and the right sided anterior as well as the posterior pillar could not be appreciated throughout its extent. The vascular markings on it were quite prominent. On palpation, the right tonsillar mass was found to be firm to soft in consistency, while the left tonsil was found to be soft. The tonsilolingual sulcus on the right side was obliterated and it was assumed that the palatine tonsil was enlarged and intermingled with the lingual tonsil.

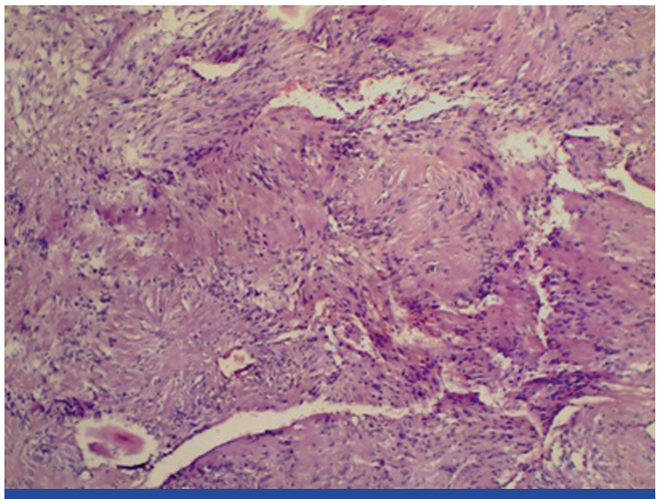
On the basis of the history, a number of episodes of sore throat and fever and the positive right jugulodigastric lymph node, a diagnosis of tonsillitis was made and bilateral tonsillectomy was done. Peri-operatively, a separate mass in the right tonsillar fossa was noted, which was thought to be the rudimentary right tonsil and it was removed by using a classical dissection method and was sent for histopathological examination.



[Table/Fig-1]: Gross appearance of schwannoma: The tumour is well encapsulated and is grevish white in colour



[Table/Fig-2]: Photpmicrograph showing area of Antoni type - B tissue in schwannoma



[Table/Fig-3]: Photpmicrograph showing area of Antoni type - A tissue with palisading of cells in schwannoma

Grossly, the larger mass which was labelled as the "right side", was grey, completely encapsulated, 3.5×2.5×2.0 cm in size and was firm in consistency, with the cut surface being greyish white. The smaller second mass in the same container was 1×1 cm in size and soft in consistency. The pieces were processed from both the masses and also from the "left sided tonsil". The histopathology from the larger mass on the right side revealed a tumour which showed hypo and hypercellular areas with the presence of elongated cells with cytoplasmic processes which were arranged in fascicles, with the presence of verocay bodies in areas of moderate to high cellularity with little stromal matrix and hypocellular areas with a loose meshwork of cells along with microcysts and myxoid changes. Thus, a diagnosis of schwanomma was suggested and imunohistochemical (IHC) studies were advised.

The light microscopic findings from the rest of the masses revealed tonsillar hyperplasia with chronic inflammatory changes. On IHC, the cells demonstrated a characteristic immunoreactivity for S-100. The light microscopic findings from the rest of the masses revealed tonsillar hyperplasia with chronic inflammatory changes. Thus, a final diagnosis of palatine schwanomma was made. It was thus proposed that the presence of schwanomma in the anatomical region led to atrophy of the tonsil of that side and to the various obstructive and other symptoms. Till the last follow up, the child was symptom free and had no other complaints.

DISCUSSION

Nerve sheath tumours which originate from the peripheral nerves are of two types: neurofibromas and schwannomas. Neurofibromas are benign neoplasms which are composed of neurites, Schwann's cells, and fibroblasts within a collagenous or myxoid matrix, whereas schwannomas originate from the Schwann cells of the nerve sheath which cover the myelinated nerve fibres [2].

Shwannomas can reach upto considerable sizes, although they usually remain small. They account for only 1% of all the benign tumours in the oropharynx and in the oral cavity [3] with the tongue, palate, cheek mucosa, lip and gingiva being the most frequent locations in the oral cavity [8]. They are often seen in the 2nd and 3rd decades of life, and are very rare below 10 years of age [9] , with no gender predilection.

Palatal schwannoma is generally asymptomatic and it usually presents as a slow growing painless nodule. The invasion of the submucosal area leads to pain and discomfort. Dysphagia and dyspnoea are present at times.

Apart from a diagnostic possibility, other ancillary investigations such as fine needle aspiration cytology (FNAC) and a radiological opinion can help to a certain extent. FNAC, though it is tedious in the oral region, can be attempted, though the information yield is often inadequate. Magnetic Resonant Imaging (MRI) can show not only the tumour and its capsule, but also in certain cases, the nerve from which it had developed. On imaging, schwannoma appears to be smooth and well-demarcated. This tumour is isointense to the muscle on the T1-weighted images and homogeneously hyperintense on the T2-weighted images [10]. Radiology however has its limitations, as Schwanomma at times, is indistinguishable from other ecapsulated benign tumours on the basis of the imaging findings, and therefore, a definite diagnosis requires a histological examination. The histopathological features are classified into two patterns: densely packed spindle cells with a palisading arrangement (Verocay bodies) as Antoni A type, and a loose hypocellular arrangement with hyalinized blood vessels and no definite architecture as Antoni B type [11]. A positive reactivity to the S-100 protein supports the Schwann-cell nature of this tumour on immunohistochemistry [12-14].

Other differentials which should be kept in mind with a mass in the palatal or in other locations in the oral cavity, include malignant lesions such as squamous cell carcinomas and sarcomas and benign lesions such as salivary gland tumours, leiomyomas, rhabdomyomas, lymphangiomas, hemangiomas, dermoid cysts, lipomas and inflammatory lesions. [15] Shah et al reported a rare case of schwannoma with secondary erosion of the zygomatic arch. Here, the tumour may have originated from a branch of the infra orbital nerve and it may have extended into the zygomatic arch and caused bone destruction. [16] Verma etal also reported a case which was close to the tip of the tongue. [17]

A detailed clinical history, physical examination, cytology and radiological assessment helps in differentiating schwanomma in such settings. Surgical excision or enucleation is the treatment of choice. The prognosis is excellent, as malignant transformation and recurrence are rare after the complete resection.

This case was worth reporting, as it highlighted the importance of including schwanomma as an important differential diagnosis whenever a treating physician encountered a tonsillar mass or a mass of unexplained aetiology in oral cavity and described a schwanomma which arose at a rare site and caused a diagnostic dilemma.

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