ISSN- O: 2458 - 868X, ISSN –P: 2458 - 8687

International Journal of Medical Science and Innovative Research (IJMSIR)

IJMSIR : A Medical Publication Hub

Available Online at: www.ijmsir.com

Volume – 2, Issue –5, September- October - 2017, Page No. : 156 - 159

Neurilemmoma: A Case Report And Review

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Conflicts of Interest: Nil

Abstract

Neurilemmomas are well-encapsulated, benign, slow-growing tumors originating from Schwann cells of the nerve sheath surrounding cranial, peripheral, or autonomic nerves. Intraoral neurilemmomas are relatively rare and have a wide variety of morphologic and radiologic features. This makes differential diagnosis difficult, and only histopathological features can lead to a definitive neurilemmoma diagnosis. In this report, we present the case of a 13-year-old patient whose chief complaint was a solitary, nodular mass on the ventral surface of the tongue. After ultrasonography, we performed an excisional biopsy that showed the typical characteristics of a neurilemmoma. The mass was removed completely with no signs or symptoms of recurrence.

Key words: Neurilemmoma, Tongue, Ultrasonography.

Introduction

Schwannomas, also commonly known as neurilemomas or neurinomas, are benign encapsulated nerve sheath tumours composed of Schwann cells. Schwannomas arise from proliferating Schwann cells which form a tumour mass of unknown etiology comprising motor and sensory peripheral nerves [1]. Although it occurs at any age, the peak incidence is between the third and sixth decades. There is no male and female predilection [2–4]. Extracranially, about 25% of all schwannomas are located in the head and neck, but only 1% occurs is intraoral origin. The intraoral lesions show a predilection for the tongue region. The palate, buccal mucosa, lip, and gingiva are also affected in decreasing order [5–7]. The tumour is normally solitary, smooth-surfaced, slow growing tumour which is generally asymptomatic. It may occasionally cause pain or discomfort. Schwannomas show no recurrence rate usually if completely excised [8,9]. Malignant transformation is rare. Das Gupta and Brasfield[10] found an incidence of 8% of malignant schwannomas in the head and neck, and Ghosh et al [11] reported an incidence of 13.9%. Here we report a case of schwannoma of the tongue.

Background{Case Report

A 13 year male patient came to our dental op with the chief complain of painless swelling in the ventral surface of the tongue for past 7 years. The swelling was of sudden onset which was small in size initially and has gradually progressed to present size in about 2 years after which there was no increase in size and has remained constant in

size. No history of pain, paraesthesia associated with the swelling and no history of trauma was elicited. On intra oral examination a single swelling was present on the ventral surface of the tongue of size 2*3 cm approximately, roughly oval in shape which extends superiorly 0.5cm away from tip of the tongue inferiorly 1cm away from lingual frenum medially extends to the midline and laterally 0.5cm from left border of the tongue (fig 1). It was firm in consistency, non compressible and non tender. According to the history of this lesion and clinical progression differential diagnosis were fibroma, Fibrosed mucocele, benign nerve tumor like schwannomas, neurofibromas, benign muscle tumors like leiomyomas, rhabdomyomas and benign minor salivary gland tumors were made. The hematological test result normal. Ultrasonography of the swelling revealed a well encapsulated lesion which was oval in shape and of size 0.8x0.9cm. The lesion was hypoechoic with no calcification and post acoustic enhancement was evident (fig 2). Colour doppler elicited no vascularity within the lesion. The lesion was surgically excised completely and subjected to histopathological examination which revealed interlacing bundles of spindle cells with hyperchromatic spindle nuclei and indistinct cytoplasm resembling Schwann cells. Antoni type B was predominant with nuclear palisading. Few verrucoy bodies were also seen throughout the section and these features were suggestive of neurilemmoma (fig 3).

Case Description

Schwannoma or neurilemmoma is a benign tumor developing from peripheral motor, sensory, sympathetic, and cranial nerve sheaths. Schwannomas in the head and neck region is relatively uncommon and should not be confused with neurofibromas. Both tumors are thought to arise from a common precursor; the Schwann cell,[1] which surrounds peripheral nerve tissue and is believed to have the neural crest origin [2]. However, some authors [8] believe that perineural fibroblast is the major cellular component, or at least, that cells participate equally with Schwann cells in their formation. Schwannomas are usually solitary, slow growing, smooth surfaced and usually asymptomatic. The lesion will be usually single, circumscribed, firm, painless, and of variable size. Ulceration of the overlying mucosa is rare and generally the result of trauma [6,9]. Masses are typically between 0.5 to 3 cm in size, rarely exceeding 5 cm,[12]. In our case also the lesion is solitary, well circumscribed painless, slow growing and firm in consistency. In the oral cavity, tongue is found to be the most common site for occurrence. Because of the mobility of the tongue, it is common to receive stimuli resulting in increase in the size of a tumour; also, it is comparatively easy to detect a lesion in the tongue. In this case USG reveals there is no vasularity and calcifications evident with an well encapsulated lesion. Histologically, schwannomas are composed of spindle type of cells which are arranged in two distinct patterns classified as Antoni Type A and B. In Type A, the cells are arranged in a palisade manner with the nuclei lying side by side in one strip and the cytoplasm of the cells in the adjacent strip (Verocay body) whilst in Type B there is no specific arrangement of the cells [4]. In this case also the histopathological examination revealed tissue of interlacing bundles of spindle cells with hyper chromatic spindle nuclei and indistinct cytoplasm resembling Schwann cells and Antoni type B were predominant with nuclear palisading. But in our case there was no pain and paraesthesia. All the cranial nerves in the head and neck region can give rise to schwannoma, except olfactory and optic nerves, which are not considered as true cranial nerves because of absence of Schwann cells(13). The features of schwannoma mimic many other lesions such as exophytic growths and other

benign tumors, which can confuse the doctor. Hence a comprehensive differential diagnosis for such lesions should always include neural tumor, though it is rare. The current treatment modality for neurilemmoma is complete surgical excision [14]. Incomplete surgical removal of a neurilemmoma may result in recurrence [15]. The choice of surgical approach is based on tumor size and location [16]. Once removed completely, neurilemmomas do not recur. The malignant transformation of neurilemmomas is rare [17]. Neurilemmomas are highly radio-resistant, and radiotherapy is not indicated for their management [18].

Conclusion

Neurilemmomas are rarest of the rare tumor which should always be included in the differential diagnosis of painless mass arising in and around the oral cavity, and careful attention should be directed towards reaching the diagnosis. In our cases wide excision of the tumours was done and showed no signs of recurrence after surgery.

Clinical Significance

In order to diagnose a painless mass arising in and around the oral cavity, with careful attention.

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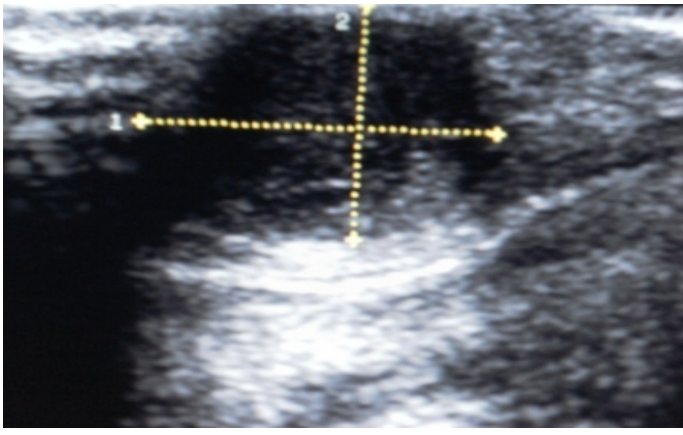

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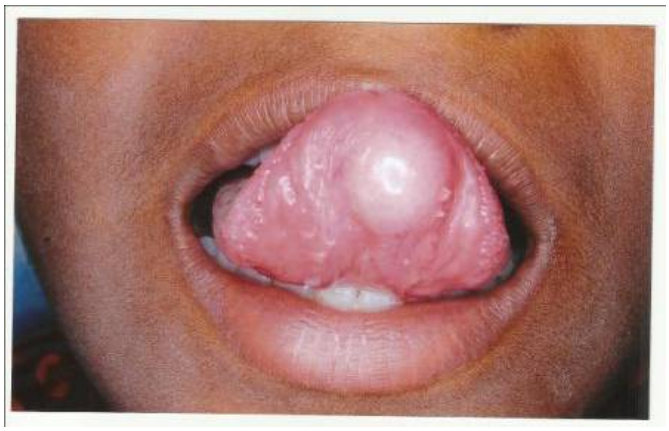
Figure 1:



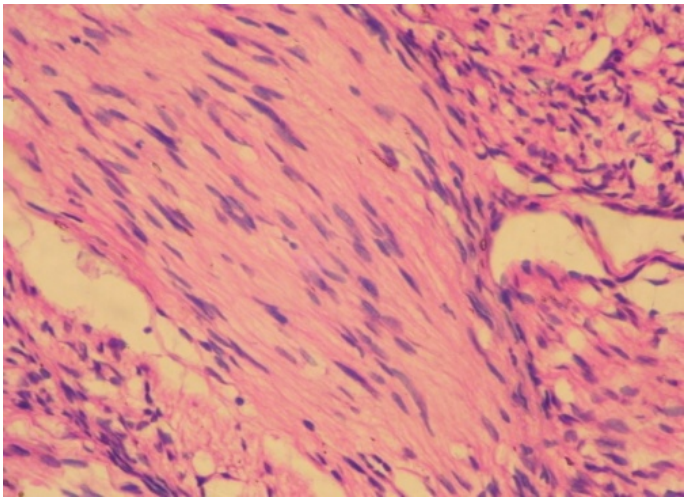


Figure 2:

Figure 3: