International Journal of Medicine and Medical Sciences Vol. 1(1), pp. 001-003, 27 November, 2011 © International Scholars Journals (http://internationalscholarsjournals.org)

Case Report

Sublingual Neurilemmoma: A Case Report

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Received October 31, 2011; Accepted November 27, 2011

Neurilemmoma or schwannoma is a benign, slow growing and encapsulated tumour originating from Schwann cells. Neurilemmoma in the oral cavity are rather uncommon and merits to be differentiated from neurofibroma. We here present a case of neurilemmoma in the floor of mouth with typical histopathological appearance.

Key words: Neurilemmoma, floor of mouth, verocay body, case report.

INTRODUCTION

A neurilemmoma or schwannoma is a benign, slow growing and encapsulated tumour originating from Schwann cells[1-4]. Approximately 25% to 40% of all neurilemmomas are seen in the head and neck region, and only 1% exhibits an intraoral localisation[5,6]. Neurilemmomas of the hypoglossal nerve are uncommon, mostly originating from the intracranial portion or having a dumb-bell shape with intracranial and extracranial components[7, 8]. Neurilemmomas of the most peripheral segment of the hypoglossal nerve, in the lower oral cavity, are very rare and only few cases have been reported till date in the literature[4, 8-12]. Here we report a case of neurilemmoma in the floor of the mouth.

Case Report

35 year female presented with swelling in the upper central part of the neck for 6 years and difficulty in deglutition for 1 year (Figure. 1). Swelling was progressively increasing in the size up to the present size and was initially seen in the upper part of the neck but now being seen in the floor of the mouth as well. There was no other swelling in body (relevant in reference to Neurofibromatosis).

Examination revealed a swelling of about 6 cm x 4 cm size in the floor of the mouth and submandibular region, non tender, firm to cystic in consistency, cross fluctuant and transillumination test negative. Tongue movements were normal though slightly restricted. There was no cervical lymphadenopathy.

Ultrasound Neck showed 6x3 cm, well defined, heterogeneous, multiloculated swelling in the floor of the mouth without any cervical lymphadenopathy.

FNAC suggested possibility of a mucous cyst.

The patient was operated by oral approach. The swelling was encapsulated and free from surroundings (Figure. 2). It was 12x8 cm in size, multinodular (Figure. 3) and on cutting, was found having fibrolipomatous tissue without any cystic area. Cut surface was uniformly grey, lobulated and solid with firm areas (Figure. 4).

Microscopically- It showed predominantly Antoni type I picture and organoid structure (Figure. 5). Palisade of cells noted alongwith *verocay bodies*. Findings suggested *Neurilemmoma.*

DISCUSSION

The schwannoma or neurilemmoma is a benign neoplasm with no stimulus or cause[13]. They are derived from the proliferation of Schwann cell (neural crest origin) that surrounds peripheral nerves. Schwann cells are

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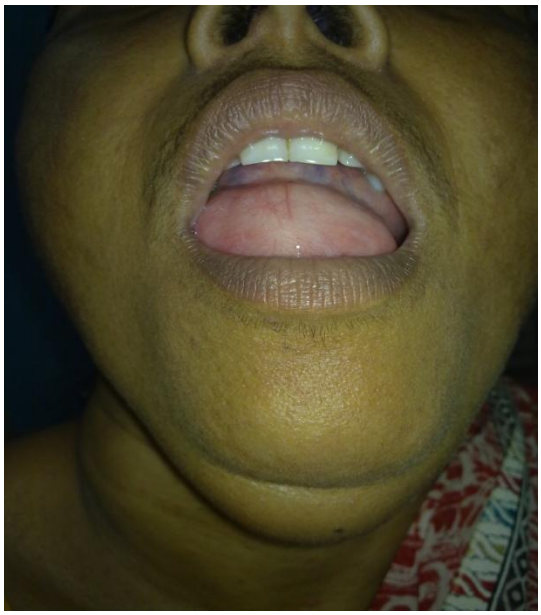

Figure 1. Clinical photograph

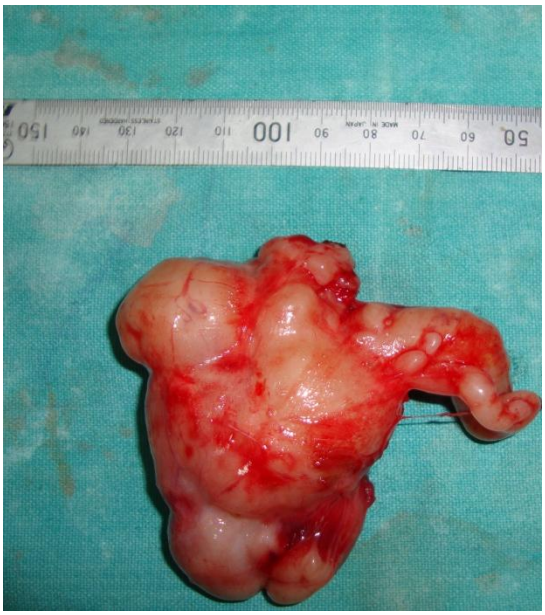

Figure 3- Lobulated specimen

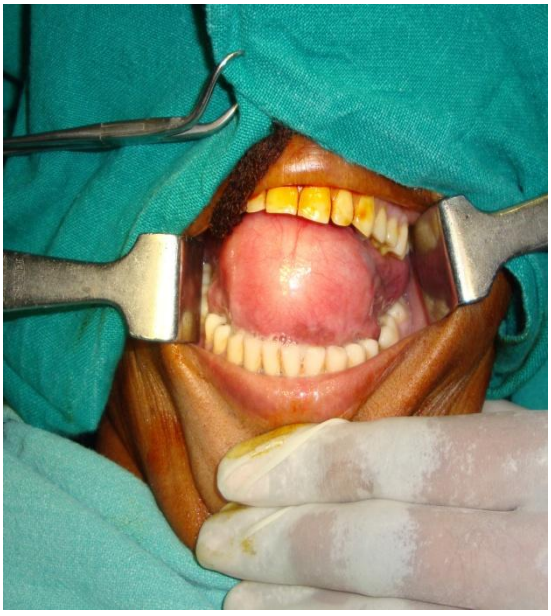

Figure 2. Intraoral lesion underneath the tongue

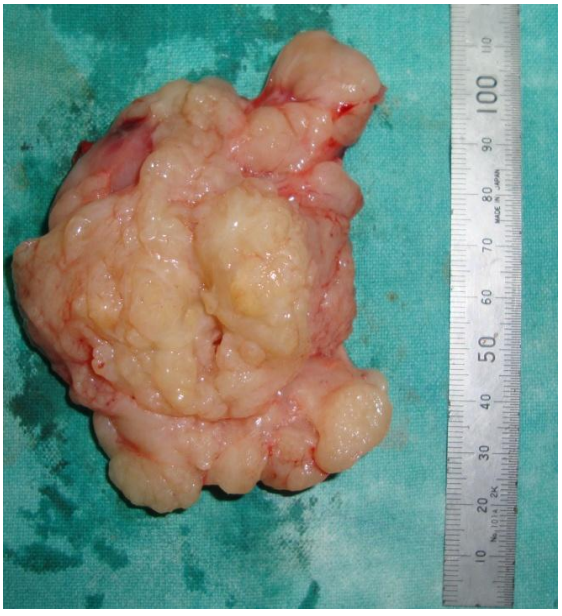

Figure 4. Cut surface of the specimen

thought of as the precursor of neurofibroma, the schwnnoma and most likely the neurogenic sarcoma[14]. Neurilemmoma or Schwannoma are relatively solid or compact arrangement of neoplastic tissue that consists of Schwann cells arranged in twisting bundles and associated with delicate reticulin fibres; the nuclei of the Schwann cells are frequently grouped in parallel rows (so-called palisades), and the nuclei and fibres sometimes form exaggerated tactile corpuscles, called *Verocay bodies*. Neurilemmomas can be found anywhere in the oral cavity, with a predilection for the tongue, but occurrence in the floor of the mouth is uncommon[6, 15]. Central neurilemmomas of the jaws have been reported, mostly located in the mandible[6]. The tumour develops in patients of all ages without an obvious preference for either sex[4, 6]. The reported duration of neurilemmomas

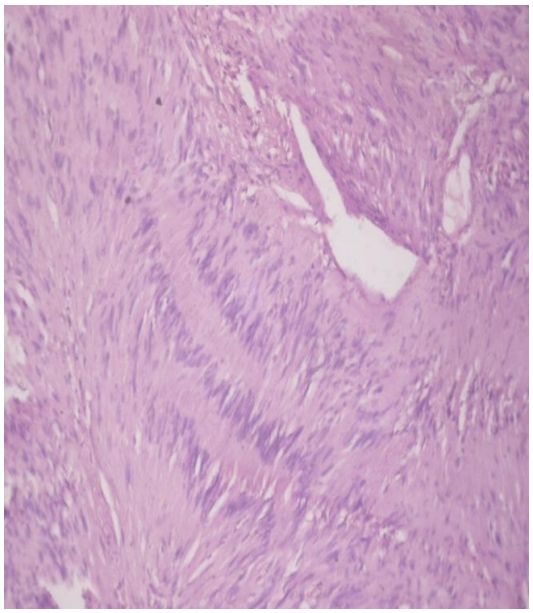

Figure 5. Histopathological view of Haematoxylin and Eosin stained slide under 100x magnification showing the presence of arrangement of cells in palisades and verocay bodies

before treatment ranges from 5 months to 49 years, with 45% of them having been present for less than 1 year[4]. The tumour in this case was large when compared with those previously reported, because neurilemmomas rarely exceed a few centimetres in diameter, and generally measure less than 20 mm[4]. Neurilemmomas are usually slow growing tumours but may have sudden increase in size in some cases possibly due to internal haemorrhage[13] . As it grows, it pushes the nerve aside which is thus not usually enmeshed in the tumour. It must be histopathologically differentiated from the neurofibromas in neurofibromatosis as neurofibromas are associated with much higher rate of the malignant transformation than neurilemmomas in which it's rather rare. The schwannomas have capsule while neurofibromas lack capsule. The myxoid changes as well as the high cellular areas as found in neurilemmoma can also be found in neurofibroma although nuclear palisading usually is not a prominent feature feature in neurofibroma[16.] Solitary neurofibroma is not usually seen in neurofibromatosis which is of considerable clinical relevance.

Neurilemmomas of the hypoglossal nerve usually originate from the skull base segment of the nerve, being entirely intracranial or located peripherally and having a dumb-bell shape. Neurilemmomas of the most peripheral segment of the nerve, especially those located in the floor of the mouth, are uncommon[7, 8, 15, 17]. Damage to the infranuclear portion of the hypoglossal nerve results in deviation of the tongue on protrusion and tongue atrophy.

Malignant transformation of neurilemmomas is very rare[4, 6]. Malignant schwannomas are highly aggressive nerve sheath tumours capable of arising de novo or from previously present neurofibroma. Neurilemmomas of the floor of the mouth should be considered in the differential diagnosis of malignant tumours (on the basis of data relating to speed of growth and clinical appearance of the neoplasm) and numerous benign epithelial and connective tissue neoformations (lipoma, traumatic fibroma, leiomyoma, granular cell tumour, neuroma and adenoma)[6]. A malignant neoplasm and a secondary hypoglossal nerve paralysis must be ruled out in the presence of hemiatrophy of the tongue. One should also keep in mind the signs and symptoms of neurological dysfunction, as the tumour may possibly originate from a cranial nerve.

When a palpable mass is found in the floor of the mouth, both CT and MRI can give elucidative information. It has been stated that MRI may be helpful in evaluating neurogenic tumours[5]. Radiographically, neurilemmomas appear as well circumscribed, encapsulated, soft tissue masses. Secondary manifestations of hypoglossal dysfunction may be seen, particularly fatty infiltration and unilateral volume loss involving the tongue musculature[7]. The treatment of choice for the tumour is surgical removal, but even in the case of incomplete excision, recurrence is low[3, 6].

CONCLUSION

Among the peripheral neurilemmomas floor of the mouth is not a frequent site. Neurilemmomas should be considered in the differential diagnosis of well-circumscribed sublingual space mass lesions, although they are rare and a biopsy would be required for definitive diagnosis. Surgical excision is the treatment of choice and recurrence rate is very low.

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