International Surgery Journal
Mona H et al. Int Surg J. 2023 Apr;10(4):795-797
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20231001

Case Report

An unexpected case of lower lip benign Schwannoma: diagnosis, management and review of literature

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Received: 18 February 2023
Accepted: 18 March 2023

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ABSTRACT

Schwannoma also known as neurinomas of verocay is a benign, slow growing, nerve sheath tumor arising from the Schwann cells of the nerve. Around 25-40% of the schwannoma arises from the soft tissues of head and neck areas. Out of which, only <1% arises from the oral cavity. The most common intra-oral site is the tongue and the least common site being the lips. Moreover, Schwannoma in the lower lip are excessively rare during childhood and adolescence. Due to its rarity in lower lip, it's diagnosis is usually overlooked during the primary examination. Complete surgical excision of the tumor is the treatment of choice. Recurrence cases and malignant degeneration are extremely rare. Here, we reported a case of a 17 year old female with peripheral Schwannoma of the lower lip, presented to the Otorhinolaryngology department, Indira Gandhi Hospital, Dwarka, Delhi with complaints of painless swelling in the lower lip causing cosmetic deformity for the patient. She underwent complete enucleation of the mass and is being follow-up in the outpatient department.

Keywords: Case reports, Benign peripheral Schwannoma, Lower lip, Verocay bodies

INTRODUCTION

Schwannoma was first described in 1910 by Verocay.[1] Benign Schwannoma is a rare, slow growing neurogenic tumor with unknown etiology arising from the nerve sheath's Schwann cells. Less then 1% cases arises from the oral cavity despite the lip and oral cavity being a highly innervated areas. The least common site of origin in the oral cavity is the lip. Lip schwannoma tends to occur in adults between the third and fifth decades of life with no sex predeliction.[2]

They usually presents as single, mobile, painless swelling which may or may not be symptomatic to the patient. The diagnosis is made histopathologically when it demonstrates cytologically bland spindle cells in hypercellular antoni A and hypocellular Antoni B patterns.Intense S-100 protein immune-reactivity for cytoplasmic and nuclear patterns indicates neural origin.[3]

Being a benign, encapsulated tumor, complete surgical resection while preserving the main nerve truck is the treatment of choice. Cases of recurrence or malignant transformation in lower lip schwannoma has been reported.[4]

Malignant peripheral nerve sheath tumors are rare tumors which can arise from a pre-existing benign nerve sheath tumor like neurofibromas or neurilemmomas. It is an aggressive tumor which are highly metastatic with high recurrence rate and poor outcomes.[5]

Due to its rarity and asymptomatic presentation, it is diagnosed later on when it causes mass effect or visual deformity. And while performing complete resection of this tumor in the oral cavity, it can lead to loss of aesthetics and functions. Though malignant transformation is rare, cases have been reported. So, benign Schwannoma should

also be included in the differential diagnosis of a slow growing mass in the oral cavity for early management.

CASE REPORT

A 17 year old female presented with complaint of painless swelling in her lower lip since 5 years which was insidious in onset, gradually progressive to the current size causing problem of cosmesis to the patient. The swelling was not associated with history of bursting or discharge. There was no history of trauma.

On examination, a single 1.5×1 cm resilient mass, slightly pale in colour, non-tender and non-mobile, present in the lower lip of the patient (Figure 1).

The patient underwent excision of mass under local anaesthesia. During the procedure, an encapsulated mass was enucleated and was sent for histopathological examination.

Histopathological reporting showed a well encapsulated neoplasm composed of hypercellular areas which showed organised spindle cells with verocay body formation (antoni A area) (Figure 2) and focal hypocellular areas in which oval to spindled cells were arranged in haphazard pattern in loose myxoid stroma (Antoni B area). Occasional interspersed thickened hyalinised blood vessels were noted.

No evidence of mitosis/necrosis/atypia was seen. The findings were suggestive of peripheral nerve sheet tumour consistent with schwannoma (Figure 3). The patient is under regular follow up (Figure 4).

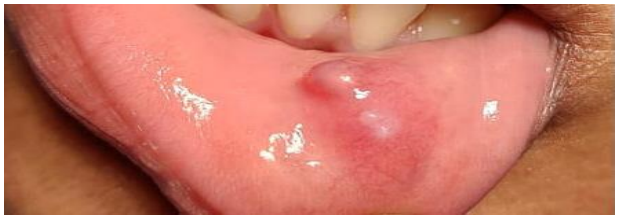

Figure 1: Pre-operative picture showing a single, slightly pale in colour, resilient mass in the lower lip.

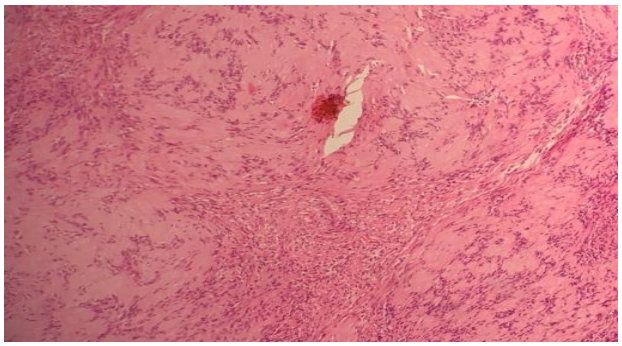

Figure 2: Histopathology slide showing verocay body formation in the hypercellular area.

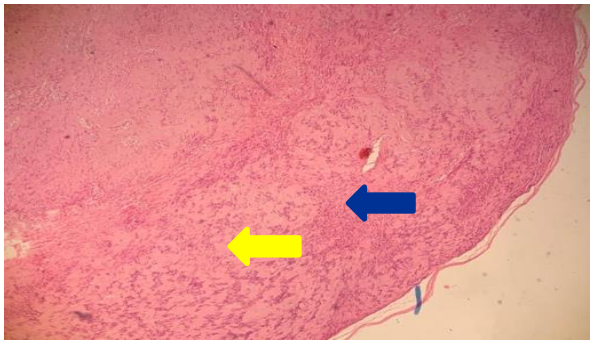

Figure 3: Histopathology slide showing a well encapsulated mass showing hypercellular areas (Antoni A) (blue arrow) and hypocellular areas (Antoni B) (yellow arrow) with haphazard arrangement of cells in myxoid stroma.

Figure 4: Day 14 post-operative picture showing the lower lip after removal of tumor.

DISCUSSION

In 2013, WHO classified the nerve sheath tumors into benign and malignant. The exact etiology is not known and it may be associated with syndromes like neurofibromatosis type 1.[4]

Benign Schwannoma of oral cavity is an uncommon presentation. Less then 1% arises from the oral cavity even when these areas are highly innervated. Of all the subsites of oral cavity, lip is the least common site.Lip schwannoma tends to occur in adults between the third and fifth decades of life, with a mean age of 26.4 years (range: 7-82) and do not show sex predilection.[2] Being a solitary, benign, slow growing tumor, they remain asymptomatic for a long period of time. Long standing cases in oral cavity can presents with dysphagia, dysarthria, restricted mouth breathing, snoring, speech and swallowing interference, and in rare cases paresthesia and pain. Furthermore, schwannomas of the lip produce apparent visual deformity for the patients and induce mental discomfort like in our case.[6]

Malignant peripheral nerve sheath tumor (MPNST) are rare and only 10-12% are reported in oral and maxillofacial area.[7] It may arise sporadically in the setting of previous radiation, from pre-existing neoplasms, including neurofibromas and neurilemmomas, or as part of neurofibromatosis type 1 (NF1). MPNST is consider if one

of the following criteria can be established: (I) origin of the tumour from a peripheral nerve; (II) origin from a pre-existing benign lesion of the nerve sheath; and (III) microscopically, the tumour reveals differentiation of Schwann cells. They are extremely aggressive with high local recurrence rate, high metastatic potential and poor survival rate. In the soft tissue of oral and maxillofacial areas, gingiva is the most affected part followed by lip. They commonly presents as rapidly growing tumor with pain, tenderness, numbness, paraesthesia and burning sensation.[4]

The diagnosis is based on the histopathological examination after biopsy or excision. Histologically, they are of seven subtypes: classical (verocay), plexiform, cellular, cranial nerve, melanotic, degenerated (ancient), and granular cell schwannomas.[8]

The classic schwannoma presents as an encapsulated tumor with two distinct histological regions. Antoni A tissue shows hypercellular spindle cells, sometimes palisade around eosinophilic areas (verocay bodies). Immunohistochemistry shows positivity for S100 protein staining. Antoni B tissue shows a hypocellular myxomatous pattern with a background of loose connective tissue. Cyst, hemorrhage, and fatty degeneration may be present. Calcifications and mitotic figures are rare.[9]

Benign Schwanommas are encapsulated and are always eccentric to the axis of the nerve. So the complete surgical excision while preserving the nerve trunk is the treatment of choice.

Recurrence of tumor in lip are reported in three cases, which maybe due to incomplete excision or potential malignant transformation.[5] It was seen that recurrence rates for lip schwannomas (5.3%) were significantly less than the recurrence rates of peripheral and head and neck schwannomas (8-24%) as described in the literature. Additionally, malignant transformation is considerably greater in lip schwannomas when compared with schwannomas in other locations (9.47% verses 0.001%).[10] Lip Schwanommas being an uncommon presentation are usually diagnosed late and can presents with cosmetic disfigurement.

Removal in such cases can lead to aesthetic and functional loss. Furthermore, since only a few cases of malignant transformation of a benign Schwanommas have been reported, wide local excision is not preferred as primary surgery.[11] However, presence of malignancy warrants removal of surrounding neovascular, subcutaneous, and bony tissue along with the tumor mass. Adjuvant radiotherapy or chemotherapy are recommended for higher stages malignant schwanommas.[12]

CONCLUSION

This rare entity must be kept under consideration while diagnosing patients presenting with mass in lip and oral cavity in order to treat it promptly to prevent aesthetic and functional loss cause by large size benign schwannomas and also to prevent chances of malignant transformation.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Mona H, Anand R, Sharma A. An unexpected case of lower lip benign Schwannoma: diagnosis, management and review of literature. Int Surg J 2023;10:795-7.