Case Report

Vestibular Schwannoma in a paediatric case—A rare entity

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Abstract

Neurilemmoma, also known as Schwannoma, is a benign, slow-growing, encapsulated neoplasm that arises from Schwann cells of the peripheral nerve sheath. Although it is extremely rare, it affects the head and neck region in roughly 25–45% of all cases. Intraorally, the tongue is the most common site and is rarely involves vestibular mucosa. Here, we report a rare case of Schwannoma of vestibular mucosa in a 13-year-old girl and contribute a review to the current literature. Clinically, based on age, site, and appearance, a diagnosis of a benign tumour such as lipoma, fibroma, neurofibroma, and benign lesions of salivary glands was made. An excisional biopsy showed solely Antoni type A tissue with central acellular eosinophilic Verocay bodies surrounded by spindle-shaped neurilemma cells arranged compactly with wavy, twisted nuclei arranged in a palisaded manner. The patient was recurrence-free after 6 months. Hence, this case is of interest due to its rarity in terms of age, site, and histopathology (Antoni type A neurilemmoma) for a presumed initial diagnosis. Here, we also hypothesize regarding the type of growth pattern in the earlier diagnosed cases.

Keywords: Antoni type A, child, intraoral, S-100 protein, Schwannoma, vestibular mucosa

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Submitted: 18-Nov-2022, **Revised:** 25-Nov-2022, **Accepted:** 16-Feb-2023, **Published:** 11-Jul-2024

INTRODUCTION

Neurilemmoma, also known as Schwannoma, is a benign, slow-growing, encapsulated neoplasm that arises from Schwann cells of the peripheral nerve sheath.[1] Although it is extremely rare, it affects the head and neck region in roughly 25–45% of all cases.[2,3] Of these, 0.1–9.3% are found within the oral cavity.[3,4] It affects females more than males,[1-4] and occurs most often during the second and third decades.[1,5,6] Intraorally, the tongue (55.7%) is the most common site involved followed by the palate (10.4%), the floor of mouth (9.4%), buccal mucosa (8.4%), gingiva and lip (5.7%), and the vestibular mucosa (4.7%).[6] It usually appears as a solitary submucosal mass in the oral cavity that is asymptomatic. Intraosseous presentation is unusual (less than 1%), with the mandible being the most common site of involvement.[7]

Histologically Schwannoma is usually an encapsulated tumour with two characteristic tissue patterns Antoni type A and Antoni type B.[8] An immunohistochemical examination of the tumour may reveal S-100 antigen positivity.[9] The treatment of choice for both peripheral and intraosseous Schwannoma is surgical excision, as it has a low rate of recurrence or malignant transformation.[1]

To the best of our knowledge, the first case of intraoral Antoni type A neurilemmoma over vestibular mucosa in a paediatric patient is being reported here. Hence, this

Access this article online

Quick Response Code:

Website: https://journals.lww.com/JPAT/

DOI: 10.4103/jomfp.jomfp_489_22

How to cite this article: Augustine J, Urs AB, Singh K, Gupta S. Vestibular Schwannoma in a paediatric case—A rare entity. J Oral Maxillofac Pathol 2024;28:297-300.

case is of interest due to its rarity in terms of age, size, and histopathology (Antoni type A neurilemmoma) for a presumed initial diagnosis.

CASE

A 13-year-old child reported a complaint of painless swelling over the left lower back tooth region that appeared abruptly 2 weeks before she presented at our institute. There had been no previous trauma or infection in the area, and there was no change in sensation. Intraoral examination revealed a well-circumscribed round to oval, sessile mass of about 15 × 10 mm dimension mass with a slightly shiny surface with a yellowish hue over the vestibular mucosa with respect to 36, 37. The mass was firm, smooth, non-tender, and non-mobile on palpation. Other findings were unremarkable.

Clinically, based on age, site, and appearance, a diagnosis of a benign tumour such as lipoma, fibroma, neurofibroma, and benign lesions of salivary glands was made. Mass was easily visible and palpable in the soft tissue; hence, no radiological investigation was required [Figure 1].

An excisional biopsy was done under local anaesthesia. Grossly, the specimen revealed an entirely encapsulated tumour mass measuring 13 × 6 × 4 mm, with a soft and greyish-white surface [Figure 2].

A histopathological examination was performed, and the lesion was diagnosed as a neurilemmoma.

Sections showed an encapsulated hypocellular tumour of loosely collagenous tissue with spindle-shaped cells. The tumour consisted entirely of Antoni type A tissue with central acellular eosinophilic Verocay bodies surrounded by spindle-shaped neurilemma cells arranged compactly having wavy, twisted nuclei arranged in palisaded manner. Variably sized blood vessels and lymphocytic infiltrations are also seen. Immunohistochemical staining was strongly positive for S-100 protein [Figure 3]. The post-operative course was uneventful, with the patient remaining free of disease during 6 months of follow-up.

DISCUSSION

Neurilemmomas are benign, slow-growing encapsulated neoplasms that arise from Schwann cells in the peripheral nerve sheath. Due to their rarity, Schwannomas are not commonly included in the differential diagnosis of oral cavity lesions. While the tongue (55.7%) is one of the most common tumour locations in the oral cavity, the location in the vestibular mucosa, as in the case presented, is quite rare. It was reported in 1957 by Mckay for the first time at this site.[6] Eleven cases of vestibular mucosal Schwannoma have been reported to date in the English literature, which is summarized in Table 1.

Vestibular Schwannoma affects all age groups, being most commonly found in the second and third decades, half of the cases are of mandibular vestibular mucosa with a slight female predisposition. On this site, they usually present as a submucosal mass of an average size of 3 cm, appearing as slow growing, without any symptoms that justify the long duration in most of the cases, and are usually noticed because of discomfort.

Clinically, Schwannomas may be indistinguishable from other benign tumours like lipoma, fibroma, neurofibroma, and benign lesions of salivary glands like mucocele. These differential diagnoses must be considered especially in the case of paediatric patients. Due to the small size, an excisional biopsy and histological examination are essential to give a definitive diagnosis. Treatment is exclusively

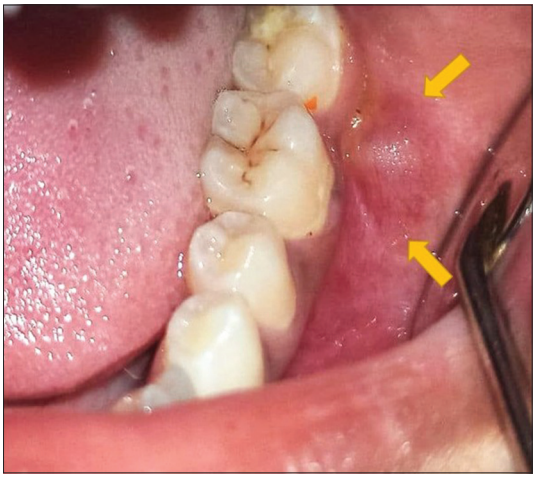

Figure 1: Solitary mass in the left mandibular vestibular region

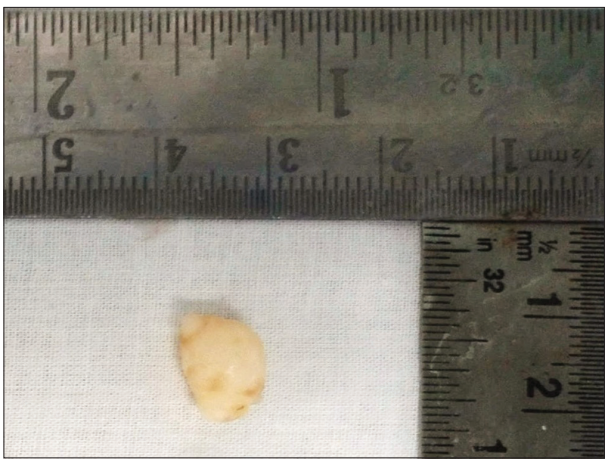

Figure 2: Photograph of the excised specimen

Table 1: Reports of Schwannomas of Vestibular mucosa published in English literature

Case no.	Year	Author	Age/ sex	Site	Duration	Size (cm)	Symptoms	Growth pattern
1	1957	Mckay *et al*.[10]	19/M	Lower labial sulcus	15 days	6	None	Antoni type A
2	1959	Winters and Simard *et al*.[11]	25/F	Upper anterior region	3 years	3×1.5	None	-
3	1960	Hénault and Laufer *et al*.[12]	13/M	Lower left region	-	2×1	None	-
4	1963	Thoma *et al*.[13]	20/F	Mental foramen	-	-	Pain	Antoni type A and B
5	1964	Putney *et al*.[2]	38/F	Lower posterior sulcus	-	-	None	Antoni type A and B
6	1983	McCoy *et al*.[14]	36/F	Maxillary left posterior mucobuccal fold	14 months	2.0	-	Antoni type A
7	1989	Dayan *et al*.[15]	52/F	Upper left vestibulum	-	0.9	-	Antoni type A
8	2006	Di Giovanni A *et al*.[16]	16/M	Vestibular mucosa		3	None	Antoni type A
9	2009	Subhashraj K *et al*.[17]	19/M	Lower left buccal vestibule	8 months	3×2.5	None	Antoni type A
10	2014	Arwade RV *et al*.[18]	40/F	Mandibular left labial vestibule	1 year	3×2.5	None	Antoni type A and B
11	2019	Sholapurkar A *et al*.[19]	26/M	Maxillary vestibule	2 years	4×3	None	Antoni type A and B
12	-	PRESENT CASE*	13/F	Mandibular left posterior buccal vestibule	2 weeks	1.5×1.0	None	Antoni type A

M=Male, F=Female

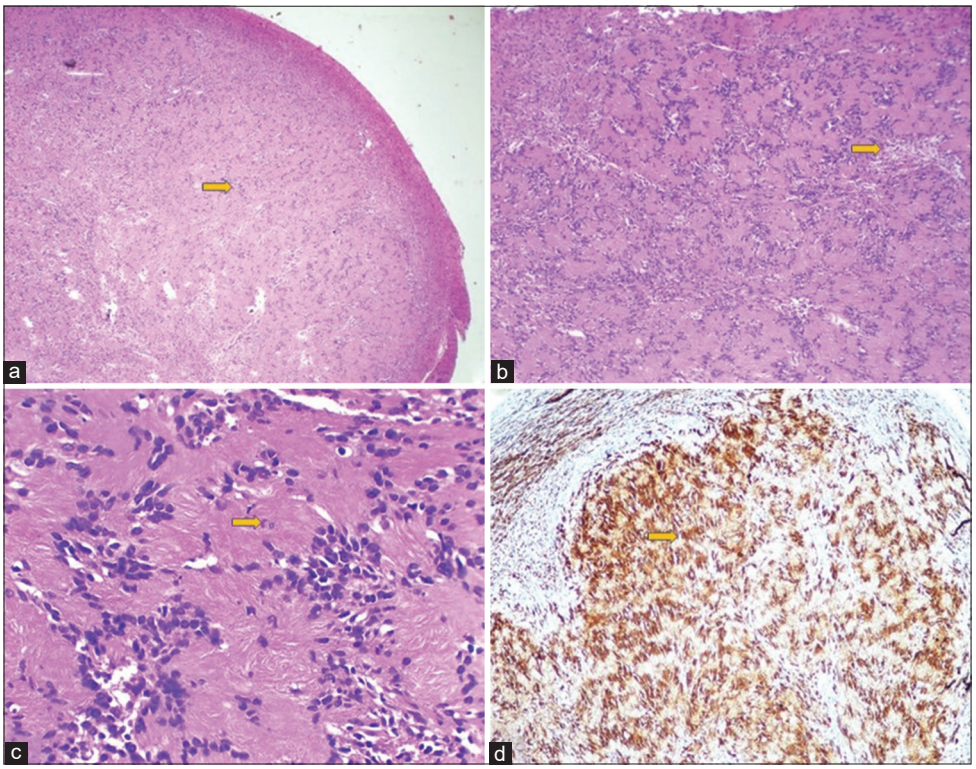

Figure 3: (a) Photomicrograph shows Antoni type A areas composed of spindle-shaped tumour cells with palisaded nuclei surrounding amorphous eosinophilic central Verocay bodies. (b) Focal areas show interstitial collagen fibres with blood vessels and lymphocytic infiltration. (c) Antoni A areas show central acellular eosinophilic Verocay bodies surrounded by spindle-shaped Schwann cells arranged compactly having wavy, twisted nuclei arranged in a palisaded manner (d) S-100 positive tumour cells

surgical. The option of complete excision was chosen due to the small size of the lesion.

Histologically, Schwannoma is usually an encapsulated tumour with two characteristic tissue patterns Antoni type A and Antoni type B.[8] Schwann cells that are closely packed, forming bundles, or arranged in rows with elongated, palisading nuclei characterize Antoni type A. Antoni type B is a mixture of Schwann cells, fibroblasts, and nerve fibres that are widely separated, dispersed loosely and randomly, and have a network of delicate reticulin fibres and numerous microcystic spaces.[2,4,8] One-third of vestibular mucosal Schwannoma cases are diagnosed with Antoni type A, within two years of duration, consistent with the present cases, while the duration of others is not known. It can be hypothesized that earlier diagnosed cases typically show one type of growth pattern, Antoni type A. However, a lager case series should be examined to prove this hypothesis significantly.

Treatment of benign Schwannoma is complete excision. Recurrence, as well as malignant transformation, is seen rarely after complete excision.[4]

CLINICAL SIGNIFICANCE

Vestibular mucosal Schwannoma is extremely rare, especially in children, and there are very few similar case reports in the English literature. As an example of a lesion frequently not considered during clinical practice or even considered a possible diagnosis, we would like to present a new case of Schwannoma of the vestibular mucosa in a paediatric patient. Given the rarity of this lesion, caution is advised because it may be clinically indistinguishable from a lipoma, fibroma or neurofibroma, and mucocele. For asymptomatic, slow-growing intraoral growths, clinicians should consider neurilemmoma as a differential diagnosis and ensure complete excision to avoid tumour recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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