Solitary Extraosseous Neurofibroma in a 5-Year-Old Child - A Case Report

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Abstract

Rationale: Neurofibroma (NF) is the most common benign peripheral nerve sheath tumour that most often occurs as a solitary tumour. A clinical and imaging diagnosis may be challenging since no distinctive features are unique for this lesion. Solitary NFs are treated by complete resection that may sometimes require a nerve sacrifice, ending with a neurological deficit. **Patient Concerns:** A 5-year-old girl with mild asymptomatic slow-growing swelling in the vestibulum of her right mandible. **Diagnosis:** A solitary variant of extraosseous NF involving the right mental branch of the inferior alveolar nerve. **Treatment:** Surgical removal of the lesion while fully preserving the mental branch. **Outcomes:** No sensory deficit was identified during a 3-month follow-up. **Take-Away Lessons:** Ultrasonography is a valuable tool for evaluating soft-tissue masses of the oral cavity. A solitary extraosseous NF involving the mental branch can be surgically removed without nerve deficit.

Keywords: Extraosseous, mental branch, neurofibroma, solitary, ultrasound

Introduction

Neurofibroma (NF) is the most common benign peripheral nerve sheath tumour. Most often, it occurs sporadically as a solitary lesion (90%) and less frequently as multiple tumours associated with neurofibromatosis type 1 (10%).[1] It is composed predominantly of Schwann cells admixed with variable numbers of perineurial-like cells, transitional cells, and fibroblasts.[2] A solitary variant is most prevalent in young adults (20–30 years), without gender preference, and presents as a slow-growing, painless lesion, varying in size from small nodule to larger mass.[3] The most common site of development is the skin, but the oral cavity may also be rarely affected, especially the tongue and the buccal mucosa.[3]

Here, we report a case involving a 5-year-old girl with an extensive diagnostic workup of extraosseous NF involving the right mental branch of the inferior alveolar nerve. A detailed description of the surgical technique for removal of the lesion is presented. The result was satisfactory without a postoperative sensory deficit.

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Quick Response Code:

Website: www.amsjournal.com

DOI: 10.4103/ams.ams_54_21

Case Report

An otherwise healthy 5-year-old girl presented to our department with her parents reporting a mild asymptomatic slow-growing swelling in the vestibulum of her right mandible. No history of local trauma was reported. The patient's medical history was unremarkable with no medications. The child had no functional difficulties during eating or speaking. Two weeks prior to her visit to our department, her local dentist had extracted her primary mandibular right second molar since the swelling was suspected to be of dental origin. Following the extraction, the swelling persisted, which encouraged her parents to seek further help.

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Received: 01-03-2021 **Last Revised:** 17-09-2021
Accepted: 20-12-2021 **Published:** 01-02-2022

How to cite this article: Emodi O, Krasovsky A, Abdalla-Aslan R, Akrish S, Ginini JG, Ohayon C, *et al*. Solitary extraosseous neurofibroma in a 5-year-old child - A case report. Ann Maxillofac Surg 2021;11:336-9.

The extraoral clinical examination showed asymmetric face, with no local redness or heat. A hard small mass could be palpated through the skin at the area of the body of the right mandible, without signs of tenderness. No limitation in mouth opening was evident. No cervical or submandibular lymphadenopathy was palpated. Sensory examination was normal. The intraoral clinical examination revealed a hard vestibular mass within the mandible [Figure 1]. Teeth were neither mobile nor tender to percussion.

A panoramic radiograph revealed a postextraction socket, normal dental development, and no obvious intraosseous pathology [Figure 2a]. A periapical x-ray revealed root remnants from the extracted right second mandibular primary molar [Figure 2b]. A cone-beam computed tomography showed a well-defined soft-tissue mass of muscle density in the area of the mental foramen, causing erosion of the buccal cortex distal to the first primary right mandibular molar [Figure 2c]. The intraoral ultrasonography (US) of the vestibular area revealed a hypoechoic well defined solid mass with no significant vascularity, measuring 2 cm × 1 cm × 1.5 cm [Figure 2d].

A decision was made to proceed with exploratory biopsy under general anaesthesia. First, the primary first right mandibular molar was extracted. Then, a buccal split-thickness mucosal flap from the primary right mandibular canine to the socket of the primary second right mandibular molar was elevated. The root remnants of the primary second right mandibular molar were extracted. An extraosseous solid mass was identified just immediately under the oral mucosa, associated with an eroded buccal alveolar cortical plate [Figure 3a]. Blunt dissection was performed around the circumference of the mass, separating the lesion from the surrounding soft and hard tissues. During the exposure of the mass, it was found to be adherent around and originating from the outer perineural layer of the mental branch of the inferior alveolar nerve [Figure 3b]. Blunt dissection was continued over and along the mental nerve, slowly detaching the inner surface of the mass from the perineural sheath. Finally, the mass was fully detached preserving the integrity of the mental branch [Figure 3c]. Both ends of the nerve should be exposed and identified across the tumour before starting to detach the mass. This is mandatory for better orientation of the course of the nerve as it passes through the tumour mass. No strength or extreme traction should be used while bluntly dissecting the mass, as it shells out quite effortlessly. If resistance is felt, a different dissection plane must be identified. The use of electrocautery is advisable. The next day, sensory examination revealed no apparent deficit over the distribution of the mental nerve. The girl reacted to light touch and pain, but no quantitive evaluation (e.g., visual analog scale/numeric rating scale, etc.) was conducted due to the young age. An uneventful course of recovery was observed during 3-month follow-ups.

Based on the morphology of the tumour, together with the immunohistochemical findings, a diagnosis of NF was rendered [Figure 4].

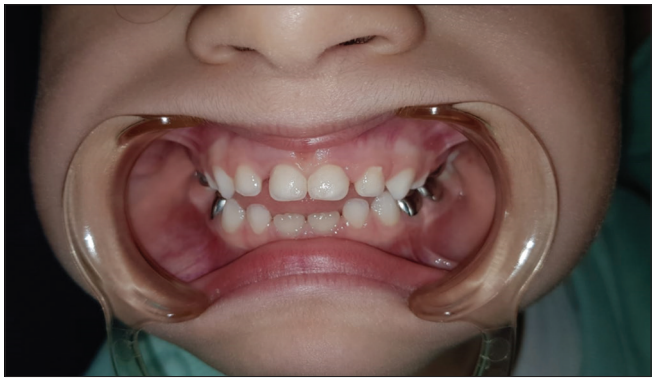

Figure 1: Clinical intraoral photograph demonstrating a vestibular mass in the mental area. Note the stretching and disappearance of the right mandibular buccal frenum

Discussion

A distinct group of extraosseous soft tissue tumours that may be relevant to the age group and the anatomical location of our patient are those of neuroectodermal origin. Common nerve sheath tumours in this category are schwannomas and NFs. Schwannomas were found to be the most common benign nerve sheath tumour of the head and neck, presenting as solitary, slow-growing, and generally asymptomatic firm masses.[4] It can range from a few millimeters to several centimeters and is most common in young and middle-aged adults.[3] Solitary NFs, although less common, tend to exhibit similar clinical features.[5] Malignant peripheral nerve sheath tumours (MPNST)

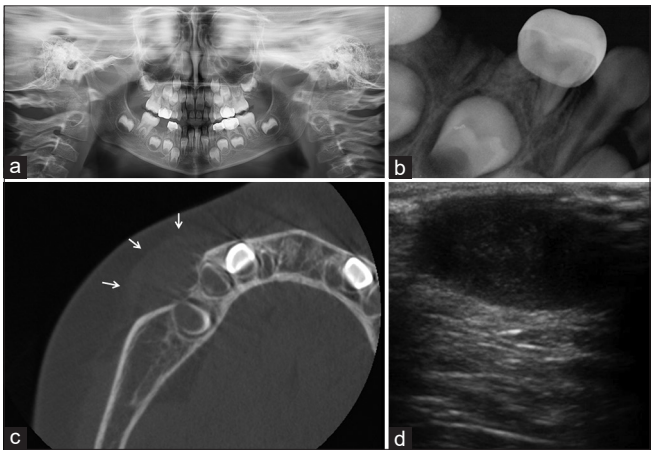

Figure 2: (a) Panoramic radiograph showing the socket of the extracted right second mandibular primary molar, with no evidence of remarkable intraosseous pathology. (b) Periapical radiograph revealing root remnants of the extracted right mandibular second primary molar, with no evidence of local intraosseous pathology. The developing teeth buds of the apically positioned premolars seem to be intact. (c) Axial view of cone-beam computed tomography of the right mandible showing a well-defined soft tissue mass (arrows) isodense to muscle in the area of the mental foramen, causing erosion of the buccal mandibular cortex with no evidence of periosteal reaction. (d) Intraoral US image in the vestibular area demonstrating a well-defined hypoechoic solid mass measuring 2 cm × 1 cm × 1.5 cm. No significant vascularity was evident

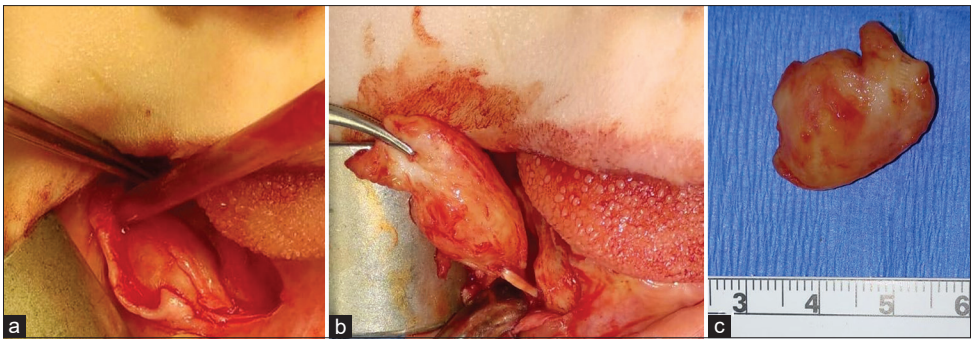

Figure 3: (a) A solid mass of approximately 2.5 cm in diameter was revealed under the oral mucosa adjacent to the buccal cortical plate. Note the bone erosion that was evident in the axial cone-beam computed tomography. (b) The mass was fully separated from the surrounding soft and hard tissues, circumferentially attached to the outer perineural layer of the extraosseous mental branch of the inferior alveolar nerve. (c) The surgical specimen of the 2.5-cm solid mass, after it was separated from the mental branch

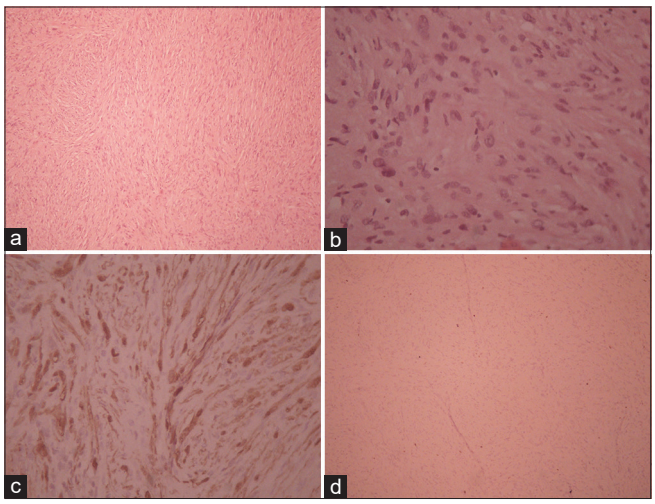

Figure 4: Histological images of neurofibroma. (a) Low power view showing a cellular soft tissue tumour. (b) Higher power view of the round and spindle cells with moderate cellularity, scant cytoplasm, and mild atypia (H and E, ×40). (c) S-100 showing strong nuclear and cytoplasmic staining of the tumour spindle cells (original magnification, ×200). (d) Ki67 demonstrating a low proliferation index (<1%) (original magnification, ×100)

constitute 10% of all peripheral nerve sheath tumours, are highly aggressive, and rapidly increase in size.[6] For this reason, MPNST were excluded from the differential diagnosis.

Fibroblastic and myofibroblastic extraosseous lesions may also be considered. Solitary myofibroma is relatively frequent in the head-and-neck region, particularly in the oral cavity, that occurs in both adults and children.[7] Clinical appearance of well localized, firm, nonmobile, and nontender swelling, together with radiographic evidence of bone erosion in the affected region can be expected.[8] A head-and-neck desmoid tumour is a benign lesion, more commonly observed and more aggressive in children, shows a predilection for the lower jaw, and appears as a firm painless mass.[3]

MRI is generally the preferred imaging modality for the evaluation of soft tissue masses. However, in this case, we performed a bedside intraoral US for several reasons, including the high availability and low cost, in contrast to MRI. In addition, an MRI examination on a 5-year-old girl would require sedation or general anaesthesia to prevent movement and claustrophobia, which we preferred to avoid. The US examination showed a well-demarcated solid mass, with no significant vascularity, which aided in arriving at the differential diagnosis and planning the surgical treatment. Solitary NFs are treated by complete resection because of the potential for extensive infiltration and malignant transformation with little chance of recurrence.[9] A complete excision may sometimes require a nerve sacrifice, ending with a neurological deficit, posing an additional treatment dilemma.[1] This case demonstrates the feasibility of the described surgical technique to preserve the full neural function of the involved branch after total excision of NF, using careful blunt dissection. Local recurrence after complete excision is unusual, <20% after near-total excision, and 40% after subtotal excision.[10] In this case, a total excision was performed; hence, local recurrence is not expected.

Conclusion

Intraoral US helped narrow the differential diagnosis and reduced time until treatment and should be considered in cases of intraoral soft tissue masses. The herein presented surgical technique can be predictably applied to avoid nerve damage and postoperative sensory deficit. A differential diagnosis of extraosseous NF should be considered in young children when intraoral soft-tissue mass is encountered.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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