

CASE REPORTS

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Congenital smooth muscle hamartoma of the tongue in a neonate — a case report

Amar A. Shah^{1*} , Abdelilah Lahmar² and Anirudh V. Shah¹

Abstract

Background: Hamartoma is a benign, tumor-like proliferation of mature tissue that is natural to the organ from which it developed. Oral hamartomas are rare and can occur as an isolated finding or as a part of a syndrome.

Case presentation: We report a 4-day-old male neonate who was postnatally noted to have a large 3.5 × 3.5 cm lobulated mass over the tip of the tongue. The mass was causing protrusion of the tongue and feeding difficulties following which surgical excision was planned. The postoperative recovery was uneventful.

Conclusion: Although rare, lingual hamartomas should be considered in the differential diagnosis algorithm for congenital tongue lesions. The definitive diagnosis is histopathological with complete resection of the tumor resulting in a favorable prognosis.

Keywords: Smooth muscle hamartoma, Tongue, Neonate

Introduction

Hamartoma is a benign, tumor-like proliferation of mature tissue that is natural to the organ from which it developed [1]. Most hamartomas are located in the liver, spleen, pancreas, and kidney [2]. Oral hamartomas are rare [2]. Lingual hamartomas can occur as an isolated finding or as part of a syndrome such as oral-facial-digital syndrome (OFDS), a set of genetic disorders characterized by oral abnormalities, facial dysmorphism and hand/foot abnormalities [3]. Only 63 cases not associated with identifiable syndromes are reported in the English literature [3]. We present a rare case of a large hamartoma over the anterior tongue in non-syndromic newborn.

Case presentation

A 4-day-old male was brought to our hospital with an intraoral mass since birth (Fig. 1). This was associated with difficulty in breast feeding. The child was born at term by vaginal delivery with a birth weight of 2.2 kg.

The antenatal history was normal and there was no mention of the mass on antenatal scan. On examination, the child had a large lobulated 3.5 cm × 3 cm mass over the tip of the tongue. The size of the mass caused protrusion of the tongue. The rest of the tongue and oral cavity was normal. The systemic examination was unremarkable. Serological evaluation revealed hemoglobin of 9.1 g/dl (normal range 11.5–16.5 g/dl), lymphocyte count 70.5% (normal range 30–70%), platelet count 187,000/cmm (normal range 200,000–500,000/cmm), and alpha-feto-protein 24,392.3 ng/ml (normal range < 0.8 ng/ml).

In view of the size of the mass and the symptoms, surgical removal was planned. The procedure was performed under general anesthesia with oral intubation. Traction was applied over the tongue and the mass delivered (Fig. 2). A “V”-shaped incision over the anterior aspect of the tongue was made, and the mass arising from the tongue musculature was completely dissected and excised (Fig. 3). Hemostasis was achieved using diathermy. The tongue reconstructed in layers with 5-0 polyglactin sutures (Fig. 4). The postoperative course was uneventful. The child was commenced on feeds with cup and spoon on the first postoperative day. Gradually, breastfeeding was started, and the child was established

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Fig. 1 Clinical photo showing the tongue mass



Fig. 2 Preoperative photo showing the tongue mass



Fig. 3 Preoperative photo following excision of the mass

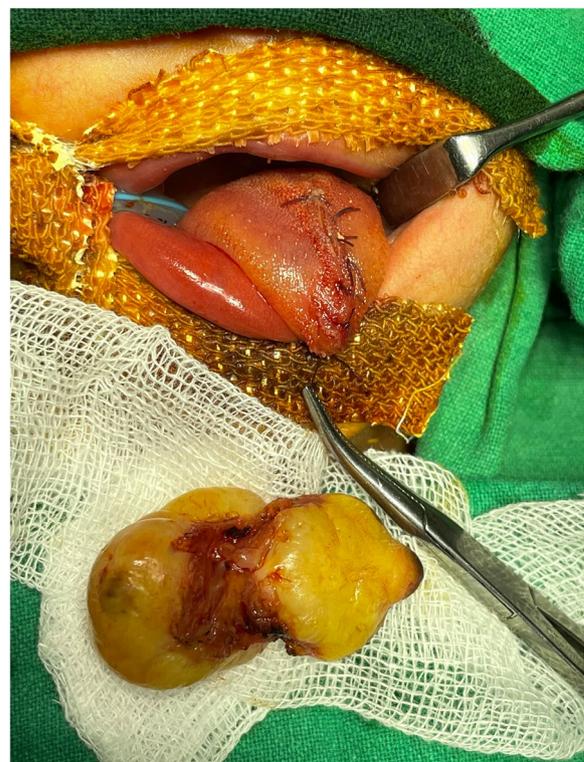


Fig. 4 Preoperative photo following reconstruction of the tongue

on full breast feeds prior to discharge on the 7th postoperative day. Histologically, the lesion showed scattered vessels with thick fibrous collar, hypocellular fibrous tissue with spindle cell proliferation consistent with benign smooth muscle hamartomatous lesion (Figs. 5, 6, 7).

The child has been in follow-up for 6 months. There has been no recurrence of the lesion. The elevated alpha

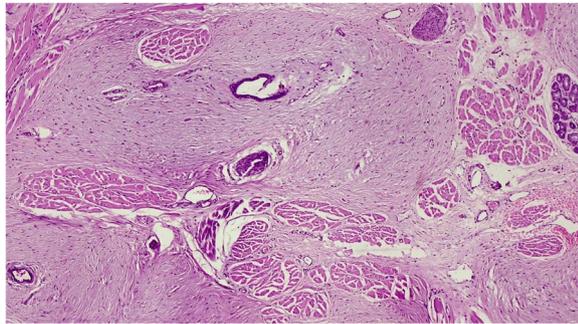


Fig. 5 Section shows fascicles of smooth muscle and salivary gland acini and ductule (hematoxylin and eosin $\times 4$)

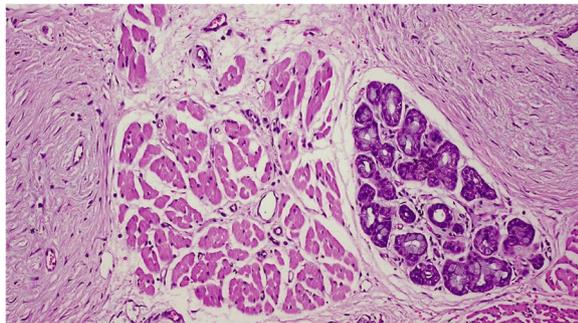


Fig. 6 Section shows smooth muscle, fibrocollagenous stroma, and salivary gland acini (hematoxylin and eosin $\times 10$)

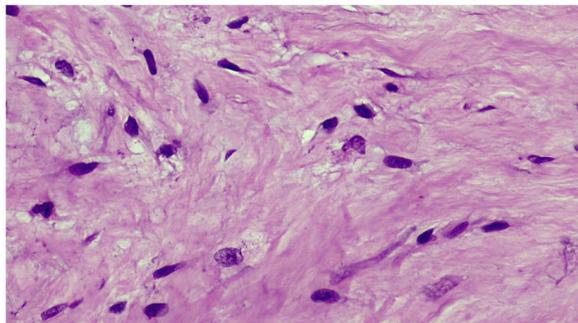


Fig. 7 Section shows plump spindle cells in a collagenous stroma (hematoxylin and eosin $\times 40$)

fetoprotein levels gradually reduced and have come back to normal over the past 6 months.

Discussion

Lingual hamartomatous lesions can be divided into 4 types depending on the predominant tissue. They are (1) neurovascular, (2) predominantly fatty, (3) smooth muscle, and (4) fatty and predominantly smooth muscle hamartomas which are also called leiomyomatous

hamartomas [1]. As per the histopathological description, the present case can be classified as a smooth muscle hamartoma. The majority of lingual hamartomatous lesions have been observed in children and adolescents [4]. In a review by Kreiger et al. [1], the dorsal anterior tongue was the most commonly affected site. In a literature review by Liu et al. [3], most of these types of lesions were located at the base of the tongue in the mid-line, while the second most affected area after that was the back of the tongue. Hamartomas of the tongue have been described in the medical literature in patients with tuberous sclerosis and orofacial-digital syndrome [3, 4]. Lingual hamartomas can be pedunculated, polypoid, or sessile.

Patients with lingual hamartomas are usually asymptomatic. Symptomatic children may have feeding difficulties and even failure to thrive [3]. A lingual hamartoma located posteriorly should be thoroughly investigated particularly when the lesion is adjacent to the foramen cecum. A lingual thyroid can be located in the same area, and surgical excision of the mass without adequate work up may render the child hypothyroid as it may be the patient's only functioning thyroid tissue [3]. Clinical examination and ultrasound of the neck can confirm whether the cervical thyroid is present. Lingual thyroid tissue can however coexist with a normal carotid thyroid. In the event of any doubt, a thyroid uptake scans can be done for confirmation [3]. In our case, the anterior tongue lesion ruled out the possibility of a lingual thyroid, and hence, no further imaging study was carried out.

The differential diagnosis of congenital lesions of the tongue include lingual thyroid, vascular and lymphatic lesions (capillary hemangioma, lymphangioma, pyogenic granuloma), traumatic lesions and benign and malignant neoplasms [1, 3]. Because lingual hamartomas can clinically resemble other lesions, histopathologic evaluation is required for a definitive diagnosis [3]. In some cases, CT and MRI are used, although their usefulness is limited for two reasons. The first is that since the oropharynx is often apparent on inspection, CT and MRI findings are less useful in characterizing lingual hamartomas. The second reason is that radiation exposure, especially in children, carries a high risk [3, 5].

Complete excision of the tumor is the treatment of choice with the inclusion of margin when there is suspicion of malignancy in the differential diagnoses [3]. Surgical approach is usually transoral using either conventional methods or a laser if the size of the tumor does not exceed 3 cm [6, 7]. In the case of a large tumor involving the tongue base and the vallecula, a suprahyoid pharyngotomy approach is recommended by some authors because it allows for better vascular control in

richly vascular lesions [4]. The prognosis is favorable with no recurrence of cases after complete excision surgery [8–10].

Conclusion

Although rare, lingual hamartomas should be considered in the differential diagnosis algorithm for congenital tongue lesions. The number of cases in the literature demonstrates that these lesions may be more common than previously suspected, particularly in asymptomatic patients. Additionally, high-risk patients such as those with tuberous sclerosis and orofacial-digital syndrome should be well assessed. The definitive diagnosis is histopathological with complete resection of the tumor resulting in a favorable prognosis.

Acknowledgements

None

Authors' contributions

AAS, operating surgeon, literature review, interpretation of the data, and drafted the manuscript. He is the corresponding author and will be accountable for all aspects of the work. AL, literature review, interpretation of the data, and drafted the manuscript. AVS, conceptualized the report and critically revised the manuscript. The authors read and approved the final manuscript.

Funding

None

Availability of data and materials

The article is about a case report of a neonate who has been operated on at the hospital owned by the authors.

Declarations

Ethics approval and consent to participate

Ethics approval was waived as the presentation is from a private children's hospital owned by the authors. Also, the study is not an experimental study. This is an uncommon case of a large hamartoma of the tongue in a neonate.

Consent for publication

The authors have obtained the necessary approval for the publication of the data from the hospital. The parents of the child have given written consent to operate the child and to share the photograph for academic purposes in the journal.

Competing interests

The authors declare that they have no competing interests.

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Received: 4 April 2022 Accepted: 2 August 2022

Published online: 17 August 2022

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