CASE REPORT

Leiomyomatous Hamartoma of Incisive Papilla with High Frenal Attachment: A Case Report

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Abstract

Aim: This case report aims to describe a rare congenital lesion of the incisive papilla with a high labial frenulum attachment, clinically mimicking congenital epulis but histopathologically diagnosed as an oral leiomyomatous hamartoma.

Background: Oral leiomyomatous hamartoma is a very rare congenital lesion, mainly appearing on the median anterior maxilla/incisive papilla and tongue.

Case description: This clinical paper is about a rare lesion in a 6-year-old female child whose parents reported to the department with the complaint of slow-growing soft tissue overgrowth between the front teeth of the upper jaw, present since birth. The soft tissue growth is now causing difficulty in biting food and is visible during smiling and speaking, causing an esthetically unpleasing appearance. The clinical examination also revealed a high labial frenulum attached to the lesion. The lesion was provisionally diagnosed as congenital epulis based on the clinical picture. However, after excisional biopsy and histologic evaluation with special stains, the lesion was finally diagnosed as leiomyomatous hamartoma.

Conclusion: Surgical excision of the lesion followed by frenectomy was performed with no postoperative complications.

Clinical significance: Owing to the rare occurrence and nature of mimicking congenital epulis, it is important for a dental practitioner to have knowledge about these types of lesions. The final diagnosis of such lesions can only be made after histopathological evaluation using special stains. This case report describes the clinical and histopathological features of a rare leiomyomatous hamartoma of the incisive papilla, along with high frenulum attachment and its management.

Keywords: Case report, Congenital epulis, Hamartoma, Incisive papilla, Smooth muscle.

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Introduction

A hamartoma is defined as a benign growth composed of an abnormal mixture of cells and tissues normally found in the area of the body where the growth occurs. They are haphazardly arranged and are native to their origin.[1] Hamartomas are not very common in the oral cavity and are usually found in the liver, spleen, pancreas, lungs, and kidneys. Most oral cavity hamartomas are of vascular origin, and those originating from other tissues are rare.[2] Oral hamartomas composed of smooth muscle fibers are known as leiomyomatous hamartomas and are rarer than those composed of fibrous, neural, and epithelial tissues. Among published cases of leiomyomatous hamartoma, most are found on the posterior tongue, hard palate, and gingiva.[3] To the best of our knowledge and literature search to date, only 19 cases of leiomyomatous hamartoma of the median maxillary gingiva have been reported in Japanese and English literature worldwide. All these cases mimicked clinical features similar to congenital epulis, but their histological features differed from those of congenital epulis. The literature also lacks reports of leiomyomatous hamartoma with high labial frenulum. Thus, this paper intends to report a rare case of congenital oral midline leiomyomatous hamartoma of the incisive papilla with high labial frenulum attachment in a 6-year-old girl, clinically mimicking congenital epulis.

Case Description

An otherwise healthy 6-year-old female child accompanied by her father reported to the Department of Pediatric Dentistry with a chief complaint of slow-growing soft tissue overgrowth over the front teeth of the upper jaw since birth. The soft tissue growth is now causing difficulty in biting food and is visible during smiling and speaking, causing an esthetically displeasing appearance. Over time, this soft tissue mass has also caused spacing between the maxillary central incisors. Medical history was normal, and there was no relevant family history pertaining to the lesion. Intraoral examination revealed a firm, pedunculated, teardrop-shaped, tubular growth on the incisive papilla between the right and left primary central incisors, extending 3–4 mm below the incisal edge of the primary central incisors. The mass was

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firm in consistency, with no signs of bleeding or pus discharge. The color of the lesion was similar to the gingiva, and its size measured approximately 8.0 × 4.0 × 4.0 mm without any signs of inflammation. A high labial frenum attachment was also present, seemingly attached to the lesion (papilla penetrating type) (Figs 1A and B). Soft tissue examination revealed normal oral mucosa, teeth, tongue, gingiva, and floor of the mouth. There were no palpable lymph nodes in the region. Based on the clinical presentation, a provisional diagnosis of congenital epulis was made.

Investigation

An intraoral periapical radiograph was advised, which revealed no bony or hard tissue changes associated with the lesion. The roots of the deciduous central incisors and the developing tooth germs of the permanent central incisors appeared normal (Fig. 2). A complete hemogram was performed before treatment, and all readings in the reports fell within normal ranges. Lastly, an excisional biopsy was performed.

Treatment

Under local anesthesia (2% lidocaine with 1:80,000 adrenaline), the lesion was excised using a #15 BP blade from the base (Fig. 3) and was sent for histopathological examination in 10% formalin solution. Following the excision of the lesion, the frenum was engaged to the depth of the vestibule using two mosquito forceps perpendicular to each other. The first mosquito forceps were engaged to the frenum as close to the alveolus as possible, and the second mosquito forceps were engaged to the frenum close to the labial mucosa of the upper lip. Incisions were made under and above the first and second mosquito forceps, respectively, using a #15 BP blade. Thus, triangular tissue was resected, resulting in a rhomboid-shaped wound, and the remaining muscle fibers were diligently dissected from the bone to prevent reattachment of the frenum. Finally, the wound was undermined before suturing, and the edges of the wound were sutured using 3-0 black silk with simple interrupted sutures, followed by COE-PAK surgical dressing. Surgical dressing and sutures were removed after 1 week, and the postoperative wound healing was uneventful. No recurrence was found after 6 months of follow-up (Fig. 4).

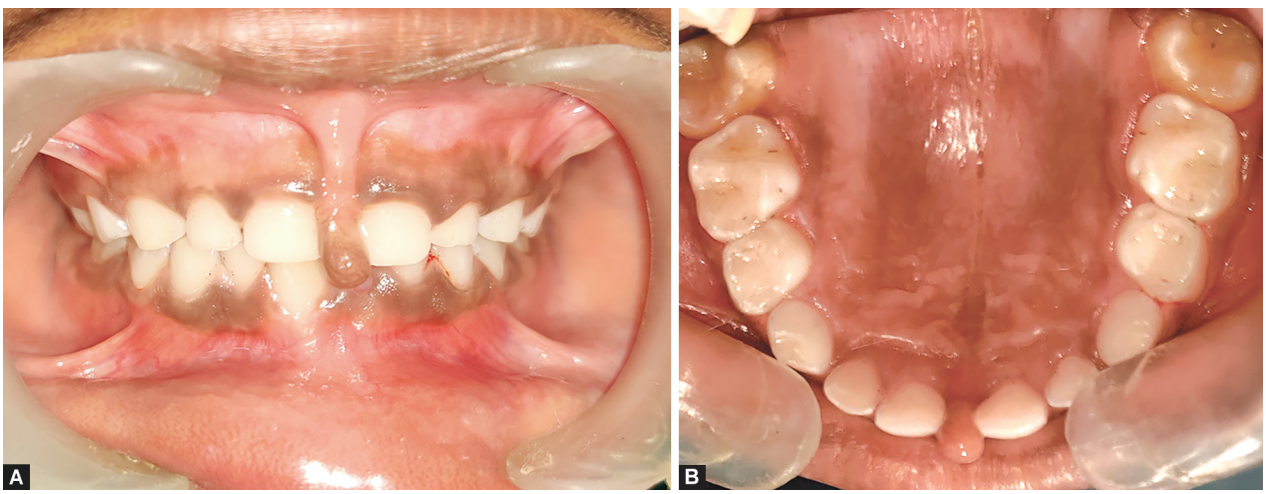
Histopathologic Examination

The excised tissue was fixed in 10% neutral buffered formalin for 24 hours, processed, and embedded in paraffin wax. 4-µm sections were subjected to conventional staining [hematoxylin and eosin (H&E)]. Microscopic examination of H&E-stained tissue sections revealed hyperplastic parakeratinized stratified squamous epithelium overlying the moderately dense connective tissue stroma with spindle to plum-shaped fibroblasts and endothelial-lined blood vessels. The central area showed the presence of smooth muscle bundles cut longitudinally and cross-sectionally. These consisted of deeply eosinophilic spindle cells with basophilic and elongated "cigar-shaped" nuclei (Fig. 5A). To confirm the presence of smooth muscle, special staining with Van Gieson's and Masson's trichrome was performed.

Van Gieson's stained sections showed yellow-colored smooth muscle fibers with surrounding red-colored collagenous tissue (Fig. 5B), and Masson's trichrome stained section showed red-colored smooth muscle fibers with surrounding blue-colored collagenous tissue (Fig. 5C). Based on the clinical and histopathological findings, a final diagnosis of leiomyomatous hamartoma was made.

Discussion

Leiomyomatous hamartoma is defined as a hamartoma composed predominantly of smooth muscle. Leiomyomatous hamartomas are usually found in the lungs, liver, spleen, or kidneys but are rarely found in the oral cavity.[4] The literature reports only 20 cases of oral leiomyomatous hamartoma in the midline of the maxillary gingiva, including the present case. All 20 cases, including the present case, are summarized in Table 1. These cases frequently occurred in females, with 15 out of the 20 reported cases occurring in females, which is about 75%. The age of first presentation of the lesion was reported to be at birth in most cases, with the maximum age of first presentation being about 2 years and 5 months, as reported by Iida et al.[4]



Figs 1A and B: Firm, pedunculated, teardrop-shaped, tubular growth on the incisive papilla between the right and left primary central incisors. (A) Frontal view; (B) Occlusal view

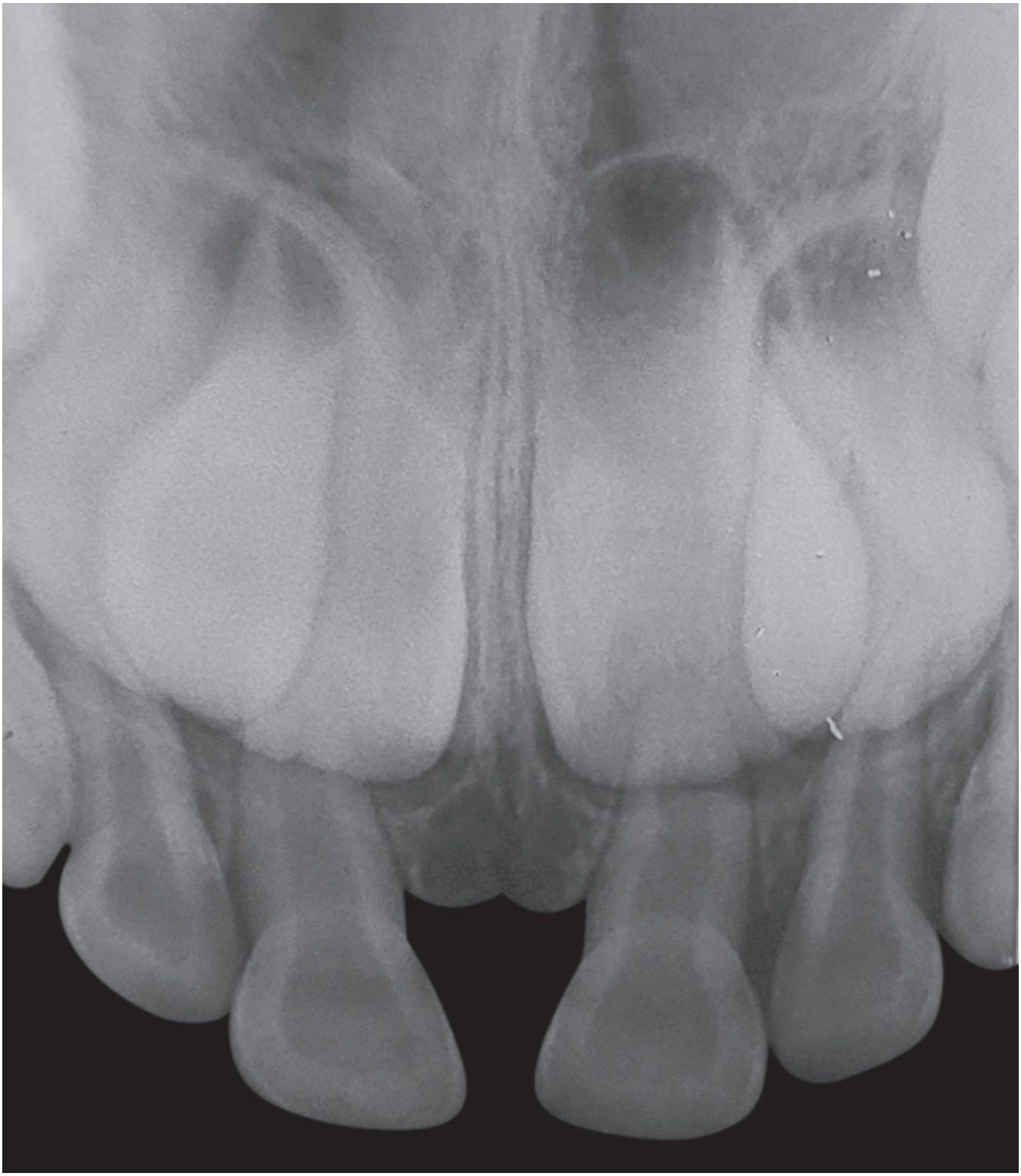


Fig. 2: Intraoral periapical radiograph showing normal radiographic appearance of the age without any bony or hard tissue changes with respect to the lesion

Oral leiomyomatous hamartomas are frequently located on the midline of the maxillary gingiva/incisive papilla and on the midline of the dorsal tongue.[2,3,5–9] This could be due to embryogenic errors during the fusion of processes and prominences in the developing embryo. Specifically, in the dental papilla, where the primary palate and palatine crests meet to form the nasopalatine foramen, from which nerves and arteries emerge, these processes are complex and prone to errors. The arteries are accompanied by cells that will

differentiate into smooth muscle cells. Dysgenic events could occur in this population of cells, giving rise to leiomyomatous hamartoma fascicles.[6] The majority of oral leiomyomatous hamartomas manifest as single lesions; however, a few cases reported by Iida et al.[4] and Kanekawa[10] revealed multiple presentations. These presentations, however, did not correspond to any syndrome and occurred in otherwise healthy individuals. The clinical examination of the present case also revealed an unusual finding of a high labial frenum attached to the lesion. Leiomyomatous hamartoma with high frenum attachment has not been reported in the literature, making this case report unique.

The clinical characteristics of the present case, including its appearance, location, sex, lack of pain, and pattern of growth, mimicked congenital epulis. However, histological findings did not show the presence of closely packed granular cells as seen in congenital epulis. Instead, histological findings revealed the presence of smooth muscle bundles, which was confirmed using special stains such as Van Gieson's and Masson's trichrome stains. Cases reported by Semba et al.,[11] Takeda et al.,[12] Correa et al.,[13] and Zhang et al.[7] revealed similar clinical and histological presentations, which confirmed the histological diagnosis as leiomyomatous hamartoma.

Few authors have used immunohistochemical markers (SMA, S-100 protein, Vimentin, Desmin, and HHF35) to confirm the diagnosis of leiomyomatous hamartoma.[4,6,7] Although

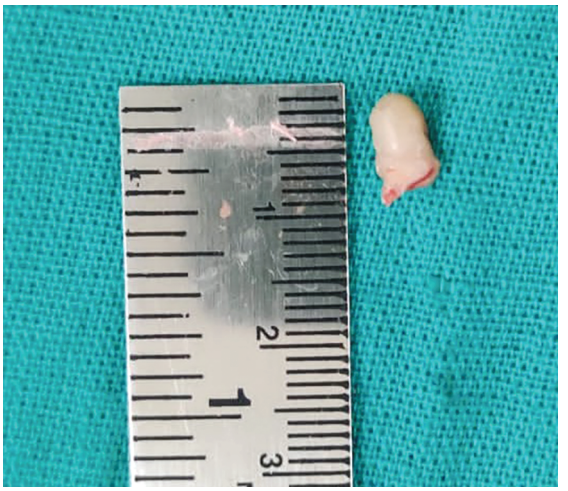


Fig. 3: Excised lesion

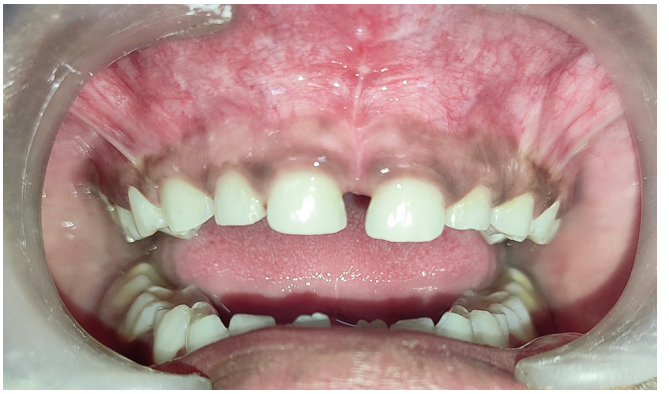
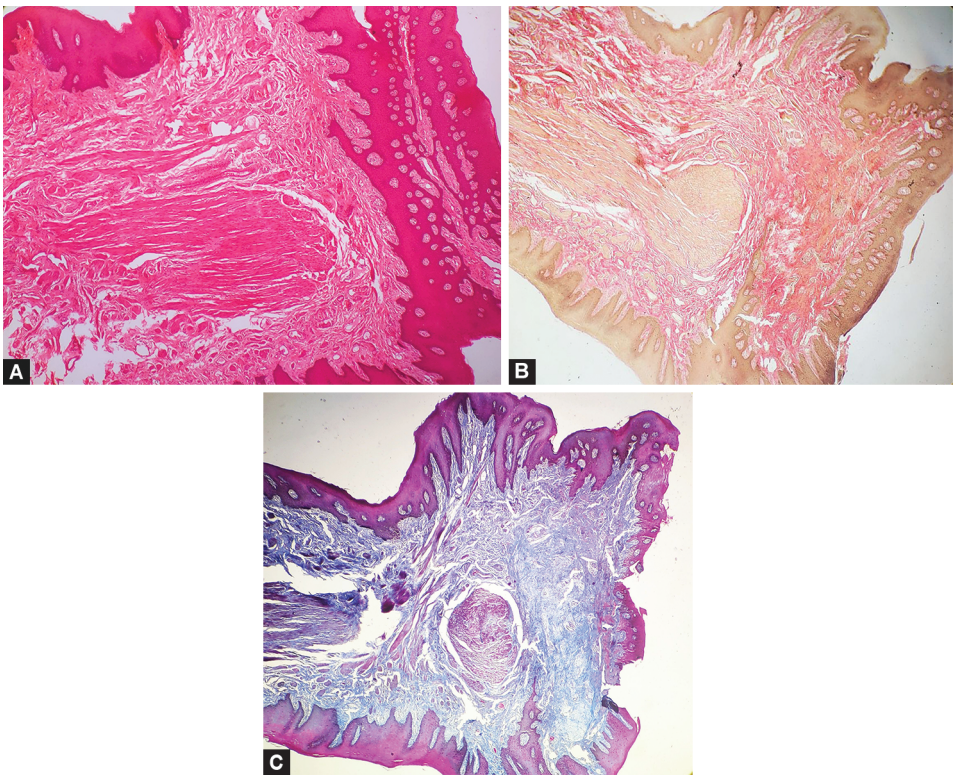


Fig. 4: After 6-month follow-up



Figs 5A to C: (A) Photomicrograph shows hyperplastic parakeratinized stratified squamous epithelium overlying the moderately dense connective tissue stroma and presence of smooth muscle fibers (H&E, 10×); (B) Photomicrograph shows yellow-colored smooth muscle fibers with surrounding red-colored collagenous tissue (Van Gieson's stain, 10×); (C) Photomicrograph shows red-colored smooth muscle fibers with surrounding blue-colored collagenous tissue (Masson's trichrome stain, 10×)

Table 1: Cases of leiomyomatous hamartoma of anterior maxilla/incisive papilla/median maxilla till date

Sr. No.	*Author*	*Year*	*Age*	*Gender*	*Site*	*Size (cm)*	*Age of first presentation*	*Ethnicity*	*Clinical diagnosis*
1.	Takahashi et al.	1962	2–3 months	F	Median maxilla	0.4 × 0.5 × 0.7	20 days	Japanese	Congenital epulis
2.	Mushimoto et al.	1982	11 months	F	Median maxilla	0.5 × 0.5 × 0.7	At birth	Japanese	Congenital epulis
3.	Kajiyama et al.	1983	4 years 5 months	F	Median maxilla	1.5 × 0.6 × 0.7	At 5 months	Japanese	Congenital epulis
4.	Kanekwa et al.	1990	3 years	M	Median maxilla	0.5 × 0.5 × 0.5	At birth	Japanese	Congenital epulis
5.	Seki et al.	1991	2 years 3 months	F	Median maxilla	0.8 × 0.5 × 0.4	At birth	Japanese	Congenital epulis
6.	Ng et al.	1992	3 months	F	Incisive papilla	0.9 × 0.4	At birth	Malay	NS
7.	Semba et al.	1993	2 years 2 months	M	Anterior maxilla	0.5 × 0.5 × 0.4	At birth	Japanese	Congenital epulis
8.	Misawa et al.	1994	1 year 7 months	F	Median maxilla	0.3 × 0.2	At birth	Japanese	Congenital epulis
9.	Takeda et al.	2000	10 months	M	Anterior maxilla	0.6 × 0.6 × 0.6	At birth	Japanese	Congenital epulis
10.	Correa et al.	2001	6 years	F	Incisive papilla	0.5	At birth	Latin American (Brazil)	Congenital epulis or fibroma
11.	Kujan et al.	2005	11 months	F	Anterior maxilla	1 × 0.5 × 0.5	At birth	Caucasian (United Kingdom)	Congenital epulis
12.	Iida et al.	2007	2 years 7 months	M	Incisive papilla	0.5 × 0.3 × 0.4	2 years 5 months	Japanese	Benign tumor
13.	Zaitoun and Triantfyllou	2007	8 years	F	Incisive papilla	0.6	At birth	Caucasian	NS
14.	Nava-Villalba et al.	2008	19 years	F	Incisive papilla	0.5 × 0.8 × 0.4	Several years	Latin American	Fibroma
15.	Scarpelli et al.	2008	6 months	F	Midline of maxilla	0.3 × 0.5 × 0.2	–	Latin American (Brazil)	Congenital epulis
16.	Zhang et al.	2011	2 years	F	Incisive papilla	0.5 × 0.5 × 0.7	5 months	Japanese	Congenital epulis
17.	ALQahtani and Qannam	2012	18 months	M	Anterior maxilla	3 × 2 × 3	18 months	Middle East (Arab)	Gingival epulis
18.	Raghunath et al.	2015	15 years	F	Median maxilla	1.5 × 2.0	6 months	Indian	Fibrous epulis
19.	Loomba et al.	2017	2 years	F	Incisive papilla	2.5 × 0.5 × 0.5	At birth	Indian	Incisive papilla growth
20.	Present case	2022	7 years	F	Incisive papilla	0.8 × 0.5 × 0.5	At birth	Indian	Congenital epulis

*Modified from reference; F, female; M, male; NS, not specified

Zaitoun and Triantfyllou[14] and Raghunath et al.[3] suggested that immunohistochemical staining is not mandatory to confirm the diagnosis of oral leiomyomatous hamartoma, as it can be diagnosed based on clinical evaluation and conventional and special staining methods, which were also employed in the present case.

Since many such cases go unreported or are surgically removed as fibromas or epulis without any histological evaluation, leiomyomatous hamartoma should be included in the differential diagnosis. There have been no reported recurrences of this lesion to date; therefore, early and prompt surgical excision is considered the treatment of choice. Delays in the excision of the lesion may cause malocclusion, resulting in developing dentition, unesthetic appearance, difficulty biting food, and speech problems.

Conclusion

Leiomyomatous hamartoma of the incisive papilla is a rare developmental abnormality of smooth muscle that clinically mimics congenital epulis. Therefore, it is important for dental professionals to have knowledge about the appearance, differential diagnosis, histology, and treatment options for this congenital lesion.

Clinical Significance

This clinical paper aims to share knowledge about a rare lesion, oral leiomyomatous hamartoma of the incisive papilla, by describing its clinical features, histopathological appearance, and treatment plan. It is essential for dental professionals to include oral leiomyomatous hamartoma in their differential diagnosis for such lesions.

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