CASE REPORT

Gingival leiomyomatous hamartoma of the maxilla: a rare entity

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SUMMARY

Hamartoma is a tumour-like malformation appearing as a focal overgrowth of normal cells. Leiomyomatous hamartomas (LHs) are rare in the oral cavity and commonly seen in the Japanese and less than 40 cases have been reported in the Japanese and English literature. The clinical differential diagnoses are irritational (traumatic) fibroma and congenital epulis. It has to be differentiated histopathologically from its neoplastic counterparts and mesenchymomas. Hence, we report such a case of LHs, which presented as a sessile gingival growth occurring in the midline in a 15-year-old girl. The final diagnosis was based on the histopathological appearance which was confirmed by immunohistochemical staining of various markers. A review of the literature of previous cases was also carried out.

BACKGROUND

Oral hamartomas are defined as tumour-like malformations of oral tissues, developmental in origin, with the tissue being native to the site.¹ The common ones are usually composed of blood and lymphatic vessels (haemangiomas and lymphangiomas).² However, some rare ones show the presence of fibrous, muscle, neural and epithelial tissues³ and even much rarer are those composed of smooth muscles.⁴ Most of the published cases have presented in the posterior tongue, hard palate and gingiva. Hitherto, fewer than 40 cases have been reported in the Japanese and English literature.^{5–9} Thus, we report a rare case of leiomyomatous hamartoma (LH) of the median maxillary gingiva in a 15-year-old girl with immunohistochemical findings to confirm the diagnosis.

CASE PRESENTATION

An apparently healthy 15-year-old girl reported to the outpatient department with symptoms of an asymptomatic, slowly enlarging and soft tissue mass with displaced teeth in the maxillary front gingival region of 6 months duration. No medical/social/family history of any importance pertaining to the lesion was reported. No extra oral swelling or intraoral examination revealed a smooth surfaced, sessile, ovoid/tubular, soft tissue mass measuring about 1.5×2 cms in size between the permanent maxillary anterior incisors (figure 1). No signs of bleeding or pus discharge could be elicited and the mass was firm in consistency. It had led to forward and lateral displacement of the left central incisor. The rest of the oral tissues were normal and the

Figure 1 Preoperative view of a reddish, smooth, non-tender mass in the midline between maxillary central incisors.

oral hygiene was reasonably good. The remainder of the physical examination was normal.

INVESTIGATIONS

No investigations were deemed necessary at this point and hence were not carried out before the treatment. The patient presented clinically with a benign fibrous growth, and a routine complete haemogram was advised, the results of which would be suggestive of a definitive diagnosis. A complete haemogram was performed and all the values were within the normal range. An excisional biopsy was carried out and sent for histopathological examination.

DIFFERENTIAL DIAGNOSIS

Lesions commonly manifesting as gingival epulides are fibrous epulis, pyogenic granuloma and peripheral giant cell granuloma. Pyogenic granuloma/lobular capillary haemangioma and giant cell granuloma are more vascular lesions and soft to firm in consistency. Keeping in mind the benign nature and presenting features of the lesion and the reasonable good oral hygiene, it was provisionally diagnosed to be a fibrous epulis.

TREATMENT

The lesion was surgically excised under local anaesthesia.

OUTCOME AND FOLLOW-UP

The excised tissue was fixed in 10% neutral-buffered formalin, embedded in paraffin and sectioned for histopathological and immunohistochemical observations. Microscopic examination of the H&E stained tissue sections revealed spindle-shaped cells forming discrete, whorled or short/long fascicles of varying thickness resembling smooth muscle tissue (figure 2A, B). These were haphazardly arranged and were surrounded by a loose collagenous connective tissue (figure 3A, B). The spindle cells had single elongated blunt-shaped

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Figure 2 (A and B) Photomicrograph showing a fibrous connective tissue lesion covered by a stratified squamous epithelium (H&E stain $\times 50$).

nuclei and there was no evidence of mitoses or cellular atypia. Towards the basal (attached) of the lesional tissue, the bundles became prominent, thick and convoluted and resembled that of nerve fascicles (figure 3C). Capillaries and small muscle-walled vessels were noted amid these fascicles, but a direct relationship between the two could not be established. Added to that, a focal, basophilic, loose fibrillary area (myxoid) with ovoid to spindle-shaped cells was noted. A clearly identifiable lamina propria separated the lesional tissue from the overlying hyperplastic, keratinising and stratified squamous epithelium (figure 3D).

In a few sections of the tissue, muscle bundles, blood vessels and fibrous connective tissue were noted in the tumour covered by a stratified squamous epithelium (figure 4A). The tumour was largely composed of smooth muscle bundles, blood vessels within fibrous stroma (figure 4B, C). The smooth muscle bundles were admixed with small nerve bundles and blood vessels (figure 4D).

Further special staining with Van Gieson stain revealed these fascicles to be smooth muscle bundles which stained yellow against the red coloured collagen bundles (figure 5). Immunohistochemical staining using an indirect immunoperoxidase method with labelled streptavidin-biotin with antibodies against α -smooth muscle actin (SMA; figure 6A) and desmin (DAKO, Denmark) further confirmed the diagnosis (figure 6B). Vimentin (DAKO, Denmark) expression highlighted the connective tissue component, blood vessels and fibroblasts (figure 6C) and anti-S-100 protein polyclonal antibody (DAKO, Denmark) immunostain showed a negative expression (figure 6D). A final diagnosis of LH of gingiva was based on clinical, histological, immunohistochemical findings. No follow-up could be carried out as the patient failed to return.

DISCUSSION

LHs present very rarely in the oral cavity. An English and Japanese literature search for oral LH until now yielded only 18 cases reported in the maxillary alveolar and gingival region. The remainder of the cases are reported in other areas like the tongue and palate.⁵⁻⁹ All the 18 cases including our case occurred in the median maxillary region (table 1).

The majority of cases occurred in the age group 2 months to 19 years and exhibited a strong female predilection with M:F=1:2.6. Most of the reported cases were noted in the incisive papilla region and measured <1 cm.⁵⁻⁹ They presented as a sessile/pedunculated, non-ulcerated, single/multilobular mass.⁶ The prognosis of these lesions is normally reasonably good. Recurrences have not been reported following a simple surgical excision of these masses.^{5,6}

The present case is the first to be reported in the Indian scenario. The lesion measured up to 1.5×2 cms and presented clinically as a fibrous epulis. Histopathologically, LHs are

Figure 3 (A and B) Photomicrograph showing fibrous connective tissue with admixture of smooth muscle bundles and blood vessels with a covering hyperplastic, para-keratinised oral epithelium (H&E stain $\times 100$). (C) Photomicrograph showing the focal area of myxoid connective tissue surrounded by collagenous tissue and blood vessels (H&E stain $\times 200$). (D) Photomicrograph showing fibrous connective tissue with an admixture of smooth muscle bundles and blood vessels with a covering hyperplastic, para-keratinised oral epithelium (H&E stain $\times 400$).

Figure 4 Microscopic images in few sections showing (A) muscle bundles, blood vessels covered by the by stratified squamous epithelium, (B) mainly composed of smooth muscle bundles, blood vessels within the fibrous stroma (C and D) smooth muscle tissue admixed with nerve bundles and blood vessels.

non-encapsulated lesions composed of thick interlacing fascicles of elongated deeply eosinophilic spindle cells surrounded by fibrous stroma, interspersed with nerve fibres and vascular channels. In our case, in addition to the classic features, a focal area of myxoid degeneration not in association with the nerve fibres was observed.

Its origin is unknown according to some researchers; the cells accompanying arteries passing through the nasopalatine foramen undergo differentiation into smooth muscle cells forming a hamartomous proliferation. During the embryonic development, fusion of several tissues has been considered due to the predilection of such occurrence at the base of the tongue and upper median gingiva.^{6 9 10} These remain inconspicuous at birth and are apparent when they enlarge rapidly following a traumatic episode. However, any event of trauma could be elicited in our case.

LHs should be differentiated histopathologically from leiomyoma, benign mesenchymoma and congenital epulis.^{7 11} Leiomyoma is a benign tumour of the smooth muscle tissue with a wide range of occurrence, affecting more commonly the lip and tongue regions. It displays large interlacing fascicles of smooth muscle cells and is encapsulated.³ In contrast, LHs are well circumscribed and, in addition to smooth muscles, contain supporting blood vessels, nerve fibres and glands.⁶ Benign mesenchymoma is a lesion characterised by two or more distinct, histologically benign mesenchymal lines of differentiation exclusive of a fibroblastic line of differentiation. They are believed to represent neoplasm and are very similar to LHs, but tend to be larger, occur in a wide age range and show infiltration into the adjacent tissues.¹¹ Fibromas are reactive lesions; histologically, they contain dense collagenous tissue with occasional spindle cells which may mimic smooth muscle tissue.

Figure 5 Photomicrograph highlighting the yellow coloured smooth muscle cells with surrounding red coloured collagenous tissue (Van Gieson stain).

Figure 6 Photograph showing immunohistochemical positivity by smooth muscle actin (A) and desmin (B), negativity by vimentin (C) and S-100 of smooth muscle fibres (D), respectively.

The use of special stains and immunohistochemistry are rather unnecessary for the histological diagnosis of intraoral LH. Still, a simple panel of immunohistochemical markers is tried in most published cases to confirm the diagnosis. Immunohistochemical staining of smooth muscle cells helps in establishing a definitive diagnosis.^{12 13} Immunohistochemical markers such as SMA and S-100 protein are most commonly used to confirm the diagnosis of LH histologically.^{2 6 7}

However, few authors have used vimentin, desmin and HHF35 markers.^{1 14}

This case thus describes such a rare case of gingival LH showing focal myxoid areas in addition to smooth muscle bundles and also is the first case of its kind to be reported in the Indian context.

To conclude, most cases go unnoticed or are excised as simple fibroma or epulis without any histological investigations.

Table 1 Summary of clinical details of reported cases of LH of the maxillary gingiva in the literature*

Author	Year	Age	Gender	Site	Size (cms)	Ethnicity	Clinical diagnosis
Takahashi <i>et al</i>	1962	2–3 months	F	Median maxilla	0.4×0.5×0.7	Japanese	Congenital epulis
Mushimoto <i>et al</i>	1982	11 months	F	Median maxilla	0.5×0.5×0.7	Japanese	Congenital epulis
Kajiyama <i>et al</i>	1983	4 years 5 months	F	Median maxilla	1.5×0.6×0.7	Japanese	Congenital epulis
Kanekwa <i>et al</i>	1990	3 years	M	Median maxilla	0.5×0.5×0.5	Japanese	Congenital epulis
Seki <i>et al</i>	1991	2 years 3 months	F	Median maxilla	0.8×0.5×0.4	Japanese	Congenital epulis
Ng <i>et al</i>	1992	3 months	F	Incisive papilla	0.9×0.4	Malay	NS
Semba <i>et al</i>	1993	2 years 2 months	M	Anterior maxilla	0.5×0.5×0.4	Japanese	Congenital epulis
Misawa <i>et al</i>	1994	1 year 7 months	F	Median maxilla	0.3×0.2	Japanese	Congenital epulis
Takeda <i>et al</i>	2000	10 months	M	Anterior maxilla	0.6×0.6×0.6	Japanese	Congenital epulis
Correa <i>et al</i>	2001	6 years	F	Incisive papilla	0.5	Latin American (Brazil)	Congenital epulis or fibroma
Kujan <i>et al</i>	2005	11 months	F	Anterior maxilla	1×0.5×0.5	Caucasian (UK)	Congenital epulis
Iida <i>et al</i>	2007	2 years 7 months	M	Incisive papilla	0.5×0.3×0.4	Japanese	Benign tumour
Zaitoun and Triantfyllou	2007	8 years	F	Incisive papilla	0.6	Caucasian	NS
Nava-Villalba <i>et al</i>	2008	19 years	F	Incisive papilla	0.5×0.8×0.4	Latin American	Fibroma
Scarpelli <i>et al</i>	2008	6 months	F	Midline of the maxilla	0.3×0.5×0.2	Latin American (Brazil)	Congenital epulis
Zhang <i>et al</i>	2011	2 years	F	Incisive maxilla	0.5×0.5×0.7	Japanese	Congenital epulis
ALQahtani D and Qannam A	2012	18 months	M	Anterior maxilla	3×2×3	Middle East (Arab)	Gingival epulis
Our case	2015	15 years	F	Median maxilla	1.5×2.0	Indian	Fibrous epulis

*Modified from refs. 5–9
F, females; M, male; NS, not specified.

Learning points

- Oral hamartomas are commonly of vascular origin, for example, haemangiomas and lymphangiomas. However, sometimes they can be composed of other tissues like smooth muscle, etc.
- Leiomyomatous hamartomas present as asymptomatic, slow growing, soft tissue swellings in the maxillary anterior region (gingival epulides), palate and tongue, with no recurrences reported so far.
- Hence, differentiation from other common benign or reactive lesions and even mesenchymomas is of academic and prognostic importance.

Competing interests None declared.

Patient consent Obtained.

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