Rare case of spindle cell haemangioma of oral cavity

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SUMMARY

Spindle cell haemangioma (SCH) is a slow growing, benign vascular lesion with a preference for the distal extremities. Its occurrence in the oral cavity is rare. Clinically, it presents as solitary or multiple subcutaneous nodules, therefore, it could be considered in the differential diagnosis of benign soft tissue tumours. Microscopically it mimics some malignant vascular tumours and it is necessary to differentiate it from other malignant vascular lesions. We report a case of SCH in anterior mandibular region of a young male in his 20s. Although it is a benign lesion, the reported case displayed extensive areas of muscle infiltration and necrosis. After studying the radiographic findings and considering the absence of cellular atypia, a final diagnosis of SCH was made. Literature survey suggests that this is the eleventh case of SCH reported in oral cavity.

BACKGROUND

Spindle cell haemangioma (SCH) is a benign vascular tumour that usually occurs in the subcutaneous tissue of distal extremities.[1] Previously, it was considered as haemangioendothelioma by Weiss and Enzinger and shared the features of benign haemangioma and malignant angiosarcoma in its behaviour.[2] In 1996, WHO renamed it as spindle cell haemangioma. Now, according to the classification International Society for the Study of Vascular Anomalies (2018), it has been included under the category of benign vascular tumours.[3]

SCH can occur either sporadically or in association with syndromic disorders (10% of cases) such as Klippel-Trenaunay-Weber, Maffucci, epithelioid haemangioendothelioma, early onset varicose veins, lymphoedema and superficial cutaneous lymphatic malformations.[1 4] It mainly affects the distal extremities, but may occur at other sites such as the chest wall, genital area, head and neck region. It has been rarely reported in oral cavity. Microscopically, it is characterised by presence of cavernous vascular channels and spindle cell proliferation.[4]

Here, we describe a case of SCH in mandibular anterior region of a young male. To the best of our knowledge, this is the eleventh case of SCH reported in the oral cavity.

CASE PRESENTATION

A male in his 20s reported to our clinic with a primary symptom of swelling in the chin region noticed over 2 years. On intraoral examination, firm, well-defined, compressible, irreducible, non-fluctuant, painless and non-pulsatile swelling of size 3×3 cm was palpated in mandibular labial vestibule.

INVESTIGATIONS

On radiographic examination, MR angiography revealed lobulated hyperintense lesion showing postcontrast enhancement in the subcutaneous plane of the submental region in midline. Lesion was extending to the right side with multiple tortuous vascular channels, suggestive of a vascular malformation. MRI (T2-weighted sequences) revealed mildly hyperintense lesion, which led to scalloping and thinning of outer cortex of mandible. The appearance was suggestive of a soft tissue tumour, possibly of vascular origin (figure 1A,B). Ultrasonography showed well-defined echogenic lesion measuring approximately 3.6×1.9 cm nestled in the subcutaneous plane of the right side of chin. It displayed a bunch of vessels with predominantly low resistance arterial flow evident on doppler imaging (figure 1C,D). Based on clinical and radiological information, a series of differential diagnosis were considered including haemangioma/vascular tumour, peripheral giant cell lesion, pyogenic granuloma, peripheral ossifying fibroma and other mesenchymal tumours such as neurofibroma and schwannoma.

The macroscopic examination of the excised lesion showed dark brown tissue and was firm in consistency (figure 2). On light microscopy study, the lesional tissue was composed of variably sized blood vessels, budding capillaries and highly cellular areas. The tumour cells were seen infiltrating the muscle and adipose tissue. Cellular areas consisted of spindle shaped tumour cells with vesicular to hyperchromatic nuclei and a few regions showed epithelioid tumour cells with enlarged vesicular nuclei and eosinophilic cytoplasm. The multiple, thin walled blood filled spaces lined by endothelial cells were seen, engorged with red blood cells and eosinophilic material. Stroma showed mild chronic inflammatory cell infiltrate interspersed with areas of necrosis and haemorrhage. Tumour cells were immunopositive for CD31 and CD34 (figure 3). Based on the radiological and histopathological features, the final diagnosis of spindle cell haemangioma was tendered.

TREATMENT

After the discussion with multidisciplinary team and considering the high vascularity of the lesion and young age, the lesion was removed surgically followed by electrocauterisation to avoid inadvertent bleeding under general anaesthesia. The overlying mucosa was excised. The borders of the lesion were demarcated in a meticulous manner by dissecting around the lesion followed by its complete removal. As the posterior margin was not easily discernible, the periosteum attached to the lesion was removed in this region.

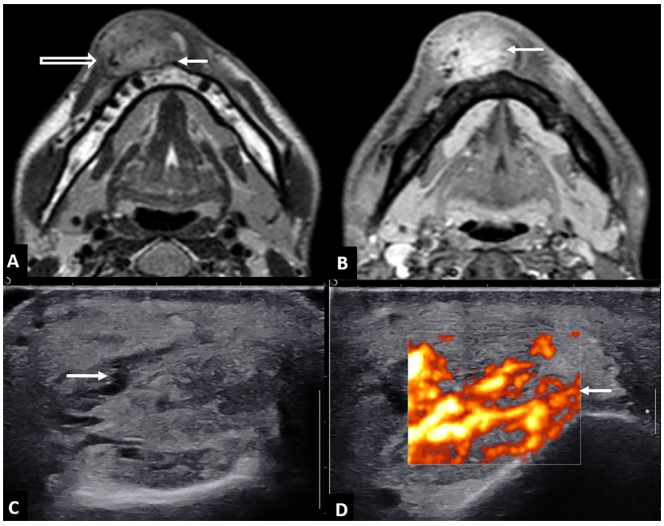

Figure 1 (A) MRI face-axial T2-weighted image shows a lobulated lesion in the subcutaneous plane of the chin, slightly to the right of the midline. Lesion is of intermediate signal intensity with few internal flow voids (arrow). The posterior margin is not well defined and is causing scalloping and mild thinning of the cortex of the underlying bone (open arrow). (B) Postcontrast T1-weighted image shows the moderate postcontrast enhancement (arrow). (C) Sagittal ultrasonography image reveals a heterogenous, solid lesion in the subcutaneous plane. The mass is predominantly echogenic with linear anechoic areas (arrow) which show colour flow on Doppler evaluation. (D) Power Doppler evaluation shows the markedly increased vascularity (arrow) within the lesion.

Since the recurrence rate of head and neck SCH is low as compared with cutaneous SCH,[5 6] surgical excision was performed, which is considered as the standard treatment. Follow-up at 8 months revealed no recurrence of the lesion.

DISCUSSION

Oral pathologists may be unfamiliar with the histopathological features of SCH in view of the rarity of its occurrence in the oral cavity. SCH is a slow growing tumour; presenting as dermal or subcutaneous nodules and mainly affects the distal extremities. Superficial lesions appear bluish while the deeper lesions appear skin coloured due to the difference in the thickness of skin cover over the lesion. SCH mostly occur in middle-aged patients but may appear at any age[1 4 7] (table 1)

Clinically, it presents as solitary or multiple subcutaneous nodules, so it could be considered in the differential diagnosis of benign soft tissue tumours. Published literature suggests that majority of cases occur in adult population. The mean age of occurrence is in the fourth decade with two cases reported from a younger population.[5 8] Male to female ratio was 1.2:1. The maximum reported size of the lesion was 3 cm and the longest duration of lesion existence prior to its diagnosis was reported as 5 years.[6] Approximately 10% of cases were associated with some inherited syndromes. Cai *et al* reported a case of SCH of lower lip in a 34 years female associated with Maffucci syndrome.[9]

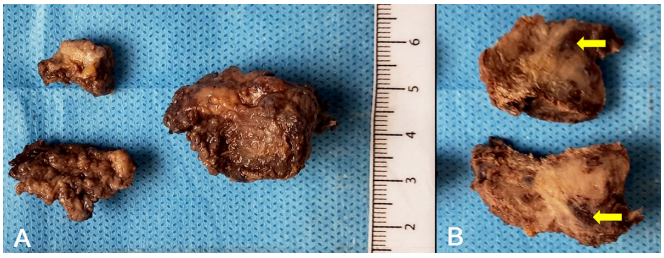

Figure 2 Gross specimen. (A) Well circumscribed brownish firm tissues. (B) Cut surface was smooth and light brown in colour. Focal dark brown necrotic areas (yellow arrow) were also appreciated.

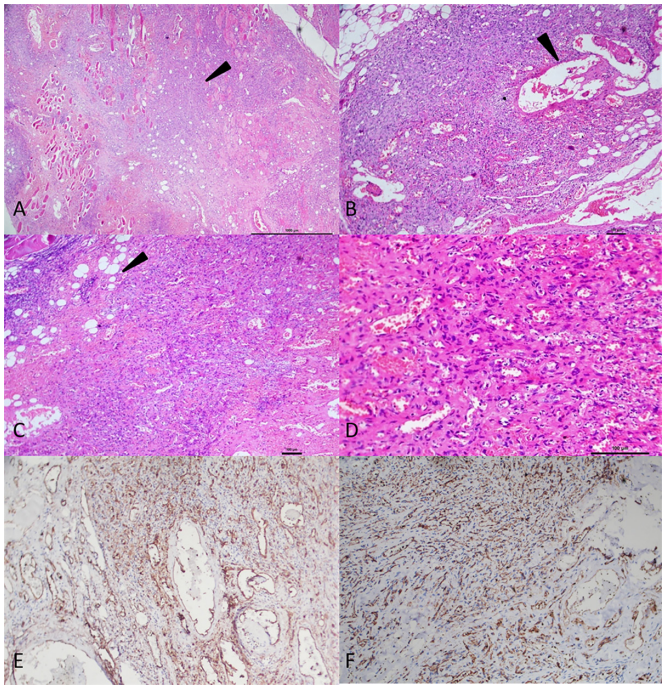

Figure 3 (A) Photomicrograph showed variably sized blood vessels, budding capillaries and highly cellular areas (black arrow) with tumour cells infiltrating the muscle. (B) Dilated blood vessels (black arrow) present adjacent to cellular areas of spindle cells. (C) Tumour cells showed infiltration into adipose tissue (black arrow). (D) Cellular areas were consisting of spindle shaped tumour cells and variably sized blood vessels. (E and F) Tumour cells were immunopositive for CD34 and CD31, respectively.

Microscopically, it presents as well-circumscribed mass surrounded by fibrous connective tissue showing a range of cellularity, imparting it a lobular architecture.[10] Variably sized blood vessels along with solid cellular areas are recognised as characteristic features. Solid cellular areas consist of spindle shaped cells with plump vesicular to hyperchromatic nuclei with some regions also showing epithelioid shaped endothelial cells. Vascular cavernous spaces are lined by endothelial cells and also contain erythrocytes.[1 11] Similar features were found in our case. Additionally, there was evidence of muscle and adipose tissue infiltration with a few areas of necrosis and the lesion lacked a definite capsule. However, prominent cytological atypia and mitotic figures were absent. It is necessary to differentiate SCH from other vascular tumours such as Kaposi sarcoma and angiosarcoma because the latter are malignant lesions.[7] In SCH, the presence of phleboliths in cavernous vessels and plump endothelial cells can differentiate it histologically from Kaposi's sarcoma. Slit-spaces can be seen in Kaposi's sarcoma, but it is not a prominent feature of SCH. Ancillary methods like immunohistochemistry are very useful to differentiate SCH from Kaposi's sarcoma. Human herpesvirus 8 positivity is found in Kaposi's sarcoma but is absent in SCH. Histologically, a lack of infiltrative growth pattern, significant nuclear atypia, high mitotic rate and

Table 1 Review of literature showing spindle cell haemangioma of oral cavity since 1995–2021

S. No.	Author(s)	Age (years)/sex	Site	Duration	Size of tumour (cm)	Immunohistochemistry	Associated syndrome
1	Tosios *et al*[8]	12/F	Mandibular buccal fold	N/A	1	Vimentin, factor VIII-associated antigen	N/A
2	Ide *et al*[11]	55/M	Palate	3 months	1.2	Factor VIII-related antigen, CD34, CD31, vimentin, SMA	N/A
3	Sheehan *et al*[7]	44/M	Buccal mucosa	N/A	1	CD31 and CD34	N/A
4	Tosios *et al*[4]	29/F	Upper lip	1 year	1×0.7	Factor VIII, CD34, SMA and Ki-67, CD68-focal positive. Oestrogen receptor-negative	N/A
5	Cai *et al*[9]	34/F	Lower lip	2 years	2×2×1	Vimentin, CD34, CD31, lymphatic endothelial cell marker D2-40 and α-SMA. S-100 protein, keratin (AE1/AE3) and CK19-negative	Maffucci syndrome
6	Chavva *et al*[13]	33/M	Below tongue	8 months	1×1.5	CD34 and CD31	N/A
7	French *et al*[1]	52/F	Dorsum of tongue	6 months	2	CD31	N/A
8	Murakami *et al*[6] 2018	41/F	Upper lip	5 years	3×2	CD34, CD31, factor VIII, SMA and WT-1 S100 protein, AE1/AE3, D2-40 and EMA-negative	N/A
9	Saikrishna *et al*[5]	10/M	Maxillary buccal vestibular fold	2 weeks	2.5×1.5	CD31	N/A
10	Panda *et al*[10]	32/M	Lower lip	N/A	2.5×1.5×1	CD31	N/A
11	Our case	25/M	Mandibular labial vestibule	2 years	3×3	CD 31, CD34	N/A

EMA, epithelial membrane antigen; F, female; M, male; N/A, not available; SMA, smooth muscle actin.

atypical mitotic figures differentiate SCH from angiosarcoma.[12] On immunohistochemical analysis, CD31 and CD34 markers are helpful to deduce the origin of tumour cells in SCH. Along with these, IHC positivity for factor VIII and SMA is also present in SCH and can help in its diagnosis.[4 7 13]

Clinical and radiological characterisation of vascular anomalies of the head and neck is necessary for guiding the appropriate treatment. A multidisciplinary approach is essential for the management of head and neck vascular anomalies. Spectral and colour Doppler ultrasonography and dynamic time resolved MR angiography are helpful to differentiate high and low flow vascular anomalies. Embolisation and sclerotherapy are the primary treatment options for these vascular lesions.[14] Nair *et al* described a simplified algorithm for effective management of vascular lesions requiring surgery.[15]

SCH is a rare benign vascular tumour of oral cavity and histologically it mimics some malignant vascular tumours. Therefore, it is essential to recognise this entity to avoid misdiagnosis and to differentiate it from other malignant vascular lesions.

Learning points

- Spindle cell haemangioma (SCH) is a rare slow growing benign vascular tumour.
- Present report is the eleventh case of SCH reported in oral cavity.
- Histopathologically, it mimics some malignant vascular tumours, which need to be carefully excluded to avoid misdiagnosis.

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