

# Spindle cell haemangioma – Report of a rare entity

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## Abstract

Spindle cell haemangioma is a rare benign vascular neoplasm. Its occurrence in the oral cavity is uncommon and may be mistaken for other common oral lesions such as mucoceles or fibromas. Microscopically, it has features which are similar to those of other vascular neoplasms and may be a cause for potential mis-diagnosis. In this article, we report a case in the lower lip of a young male patient.

**Keywords:** Endothelial cells, epithelioid haemangioendothelioma, haemangioma, lip, spindle cell haemangioma, vascular neoplasms

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## INTRODUCTION

Spindle cell haemangioma (SCH) is a benign vascular tumour that typically occurs in acral locations.<sup>[1]</sup> Its occurrence in the oral cavity is very rare. Only 13 cases have been reported previously in the head and neck region.<sup>[2]</sup> We present a case of SCH in the lower lip of a young male patient.

## CASE REPORT

A 27-year-old male patient visited our institution with a complaint of a swelling in the lower lip since the past 6 months. He had noticed a swelling in the lower lip which seemed to have episodes of growth and remission in the past 6 months. He had no other symptoms. On examination, a sub-mucosal nodular swelling was seen in the inner aspect of the lower lip in relation to 33. It had the same color as that of the surrounding mucosa and measured about 0.7 × 0.7 cm. On palpation, it was firm, sessile, and non-tender. A differential diagnosis of mucoceles and

fibrosed minor salivary gland was given, and the lesion was excised. The gross specimen was greyish-white and firm in consistency measuring 1 × 0.6 × 0.5 cm [Figure 1a]. On sectioning, a circumscribed lesion was seen in the connective tissue below the epithelium [Figure 1b]. Microscopically, a well-circumscribed lesion composed of thin-walled cavernous vessels lined by flat endothelial cells was seen in the deeper connective tissue stroma [Figure 2]. These cavernous spaces were filled with red blood cells and thrombi [Figures 3 and 4]. In between the cavernous spaces, cellular areas composed of proliferating bland spindle cells were seen [Figure 5]. A few vacuolated endothelial cells were also seen in between. Numerous plump endothelial cells showing lumina formation were noted. Mitosis and atypia were not observed. Immunohistochemical stains for vascular markers CD31 and CD34 were positive for the endothelial cells, whereas the spindle cells were negative [Figure 6a and b]. A diagnosis of spindle cell haemangioma was given.

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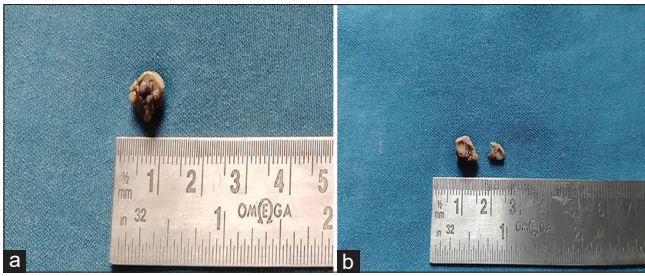


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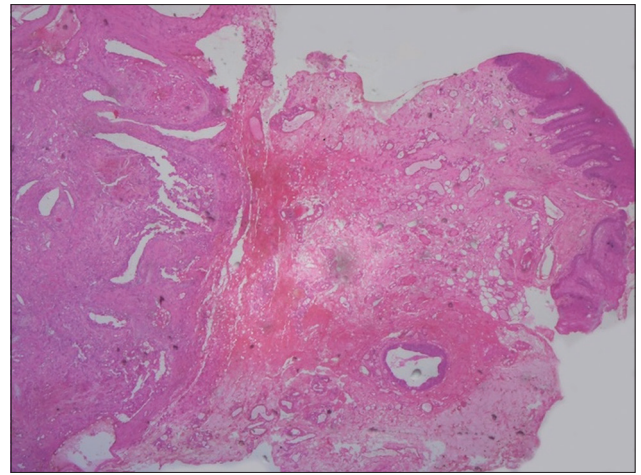
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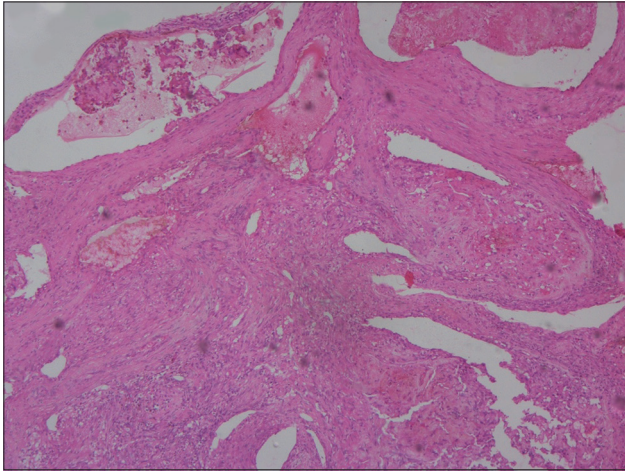
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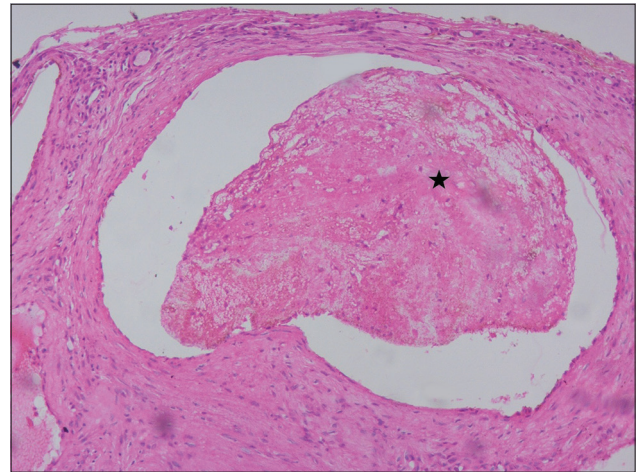
**Figure 1:** (a) Gross appearance of the excised lesion; (b) Cross-section showing the circumscribed lesion in the connective tissue



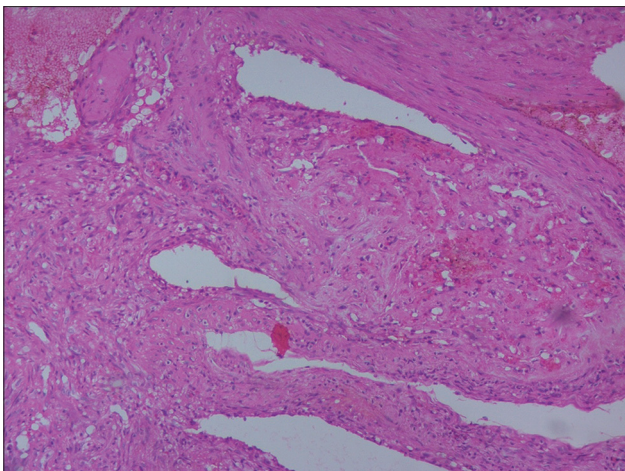
**Figure 2:** Photomicrograph showing the circumscribed tumour in the deeper connective tissue (H&E,40x)



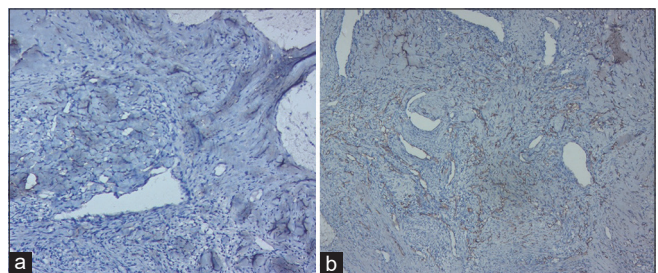
**Figure 3:** Photomicrograph showing cavernous vascular spaces (H&E,100x)



**Figure 4:** Photomicrograph showing a large thrombus (asterisk) inside a vessel (H&E,200x)



**Figure 5:** Photomicrograph showing a high-power view of proliferating spindle cells in between the cavernous spaces (H&E,200x)



**Figure 6:** Photomicrograph showing immunohistochemical staining with (a) CD31 and (b) CD34

## DISCUSSION

SCH was first described in 1986 as a vascular tumour with areas resembling both cavernous haemangioma and Kaposi's sarcoma by Weiss and Enzinger.<sup>[3]</sup> They named it as spindle cell haemangioendothelioma with the belief that it was a neoplasm of intermediate grade malignancy. It was later renamed in 1996 by the World

Health Organisation as SCH, suggesting its benign course. It can occur singly or as multiple lesions.<sup>[4]</sup> Perkins and Weiss<sup>[5]</sup> proposed the term SCH for single lesions and spindle cell haemangiomatosis for multiple lesions.<sup>[2]</sup> SCH can be associated with other developmental anomalies or syndromes such as early onset varicose veins, lymphedema, Klippel–Trenaunay–Weber syndrome, Maffucci's syndrome, epithelioid haemangioendothelioma, and superficial



cutaneous lymphatic malformations.<sup>[4]</sup> Studies have shown the strong association between Maffucci's syndrome and Ollier's disease and the occurrence of SCH and specifically the presence of mutations of IDH1 or IDH2.<sup>[6]</sup>

Imayama *et al.*<sup>[7]</sup> suggested that haemangioendothelioma is a reactive process associated with vascular damage. They also hypothesised that the behaviour of the endothelium may facilitate thrombosis, and thrombus organisation with new vascular proliferation may explain the pathogenesis of the lesion. Fletcher *et al.*<sup>[8]</sup> thought that the presence of clusters of abnormal vessels close to the tumour and a smooth muscle component within the tumour as well as the association of some SCH with early varicosities was indicative of a non-neoplastic and possibly reactive lesion. They proposed that SCH was caused by abnormalities of blood flow because of an arteriovenous shunt at the affected area. Perkins and Weiss<sup>[5]</sup> noticed that SCH resembled angiomatosis in the presence of abnormally engorged vessels, herniations, and intra-luminal webs, and hence, they suggested that SCH was a benign vascular neoplasm. They felt that alteration in blood flow could explain the biphasic nature of the lesion.

SCH mostly affects the dermis and epidermis of distal extremities and is very rare in the head and neck region. Both the solitary and multiple lesions may vary in size from a few millimetres to a few centimetres, and most lesions reported are less than 2.0 cm.<sup>[9]</sup> Only one case reported in the head and neck was over 3.0 cm in size.<sup>[2]</sup> It may be normal in colour or may be bluish and is usually firm in consistency. They have been reported in a wide age range. There was no significant gender predilection, although solitary lesions are found to be more common in males and multiple lesions in females. A PubMed search of English literature for cases of SCH in the oral cavity was performed and is given in Table 1. Most of the cases were given a clinical diagnosis of common oral lesions such as mucoceles and pyogenic granuloma.

Microscopically, SCH shows a variable architecture and has a lobular architecture on low power.<sup>[2]</sup> It has a biphasic

pattern composed of cavernous vascular spaces and solid cellular stroma with varying proportions.<sup>[9]</sup> The cavernous spaces are thin-walled and lined by flat endothelial cells. They may be filled with blood, thrombi, or phlebolith. The cellular areas are composed of spindle cells forming short fascicles and small slit-like spaces. Atypia and mitotic figures are not seen. Many immunohistochemical studies have shown that the cells lining the cavernous vessels and vacuolated epithelioid cells stain positive for vimentin, CD31, CD34, and factor VIII-related antigens, thus proving the vascular origin.<sup>[2,9]</sup> However, in most reports, the spindle cells do not stain with endothelial markers, but in a few studies, they have shown focal positivity.<sup>[1,4]</sup> The lesional cells do not react for S-100 protein, type IV collagen, cathepsin B, cytokeratins, epithelial membrane antigens, or herpes virus 8 latent nuclear antigen 1.<sup>[9]</sup> Wang L *et al.*<sup>[14]</sup> performed an immunohistochemical study in 12 cases of SCH. They found that the tumour cells were positive for Prox1 (expressed in lymphatic endothelial cells), focally positive for D2-40, and negative for WT1 (Wilms tumour 1), which suggested that this disease could be a lymphatic malformation rather than a neoplasia.

The differential diagnosis of SCH includes other vascular tumours such as pyogenic granuloma, Kaposi sarcoma, cavernous haemangioma, epithelioid haemangioma, intra-vascular papillary endothelial hyperplasia, and Kaposiform haemangioendothelioma.<sup>[2,3,9]</sup> The presence of spindle cells and occasional multiple lesions makes this tumour similar to Kaposi sarcoma. However, Kaposi sarcoma has an infiltrative growth pattern with some amount of nuclear atypia and a higher mitotic rate, which is not seen in SCH. Moreover, cavernous spaces with thrombi and phlebolith are usually not seen in Kaposi sarcoma. SCH shows negative for immunostaining with herpes virus 8 latent nuclear antigens, and the spindle cells are also negative for CD34 unlike Kaposi sarcoma.<sup>[1,2,9]</sup> Spindle cell proliferation is not seen in cavernous haemangioma, although the cavernous vascular spaces may look similar. Epithelioid

**Table 1: Previously reported cases of SCH in the oral cavity**

Author	Age	Gender	Site	Clinical diagnosis
Tosios K <i>et al.</i> <sup>[10]</sup>	12	F	Mandibular vestibule	Haemangioma/pyogenic granuloma
Ide F <i>et al.</i> <sup>[11]</sup>	55	M	Palate	Pyogenic granuloma
Lade H <i>et al.</i> <sup>[12]</sup>	25	M	Posterior pharyngeal wall	Synovial sarcoma
Sheehan M <i>et al.</i> <sup>[11]</sup>	44	M	Buccal mucosa	Vascular tumour
Tosios K <i>et al.</i> <sup>[9]</sup>	29	F	Upper lip	Mucocele
Cai Y <i>et al.</i> <sup>[13]</sup>	34	F	Lower lip	Chondroma
Chavva S <i>et al.</i> <sup>[4]</sup>	33	M	Floor of the mouth	Minor salivary gland tumour
French KEM <i>et al.</i> <sup>[3]</sup>	52	F	Tongue	Not mentioned
Murakami K <i>et al.</i> <sup>[2]</sup>	41	F	Upper lip	Haemangioma

haemangioendothelioma has a more solid appearance and lacks cavernous spaces. Pyogenic granuloma is a polypoid, circumscribed, exophytic lobular proliferation of capillaries in a fibromyxoid stroma with surface ulceration and cavernous spaces with spindle cell proliferation absent, thus helping to differentiate it from SCH.<sup>[3]</sup> SCH may show areas similar to intra-vascular papillary endothelial hyperplasia, but the latter is more acellular, has many papillae, and does not present spindle and epithelioid endothelial cells.<sup>[9]</sup> Kaposiform haemangioendothelioma exhibits distinct glomeruloid nests, which are not seen in SCH.<sup>[9]</sup>

Surgical excision is the best treatment for SCH with good prognosis. Although more than 50% of the patients may develop new lesions in the same anatomic region several years after initial excision, these are not considered a recurrence but new primaries or continuous multi-focal intra-vascular growth.<sup>[2]</sup> Post-operative radiotherapy, low-dose interferon  $\alpha$ -2b, and intra-lesional and intra-arterial administration of recombinant interleukin 2 have been successful in treating and/or preventing recurrence of inaccessible or multiple SCH.<sup>[9]</sup> Local excision was performed in our case, and the patient is on regular follow-up for the past 2 years and has not shown any recurrence.

## CONCLUSION

We are reporting the 14<sup>th</sup> case of SCH in the head and neck region and the 10<sup>th</sup> case in the oral cavity. SCH is a rare vascular neoplasm which may be mistaken for more common oral lesions such as mucoceles when occurring in the lip. Our patient reported with a solitary lesion and did not show features of any other associated syndromes. Histopathological features were suggestive of SCH, and immunohistochemical studies were performed to confirm the diagnosis.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will

not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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