CASE REPORT

Oral superficial haemosiderotic lymphovascular malformation: a rare presentation

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SUMMARY

We present an extremely rare case of a 53-year-old woman with an intraoral superficial haemosiderotic lymphovascular malformation (SHLM), also known as hobnail haemangioma. SHLM is a rare, benign, vascular tumour first described as targetoid haemosiderotic haemangioma, with only a handful of cases reported to present in the oral cavity. The diagnosis was established following complete surgical excision of the lesion, and after 14 months, there are still no signs of recurrence. Although SHLM is an uncommon condition, accurate and timely diagnosis is valuable in distinguishing these lesions from their more serious competing differential diagnoses.

BACKGROUND

Superficial haemosiderotic lymphovascular malformation (SHLM) is a rare vascular neoplasm of unknown aetiology, even more so when presenting in the oral cavity; although trauma has been implicated as a cause in some cases.¹

SHLM is common in young or middle-aged men, typically presents in the trunk or extremities and very rarely in the oral cavity. It is characterised by a papular or nodular lesion, which is often pigmented and may have a halo or 'targetoid' appearance.

Histological features include the presence of dilated and thin-walled vessels in the superficial layer. These vessels are lined by endothelial cells, which protrude into the lumina, giving them their characteristic 'hobnail' appearance. Deeper vascular spaces are slit-like and narrower; these vascular channels appear to dissect between collagen bundles. Haemosiderin deposition is usually seen in the surrounding stroma. Immunohistochemistry

shows that Kaposi's sarcoma-associated herpesvirus HHV8 is consistently negative for these lesions.¹⁻³

Although the presence of a hobnail appearance is a typical feature of this neoplasm, it is not a specific phenomenon. Therefore, a more appropriate term to use would be SHLM. This better delineates the aetiology of the neoplasm and eliminates any limiting or confounding terms, such as haemangioma, as these are not truly haemangiomas.

The clinical, histopathological and immunohistochemical features seen in this case were consistent with those of SHLM.

Currently, there are less than five reported cases in English literature.⁴⁻⁶ Distinguishing SHLM from the more serious differential diagnoses of progressive lymphangioma, Kaposi sarcoma, microvenular haemangioma and haemangioendothelioma variants is important.¹

CASE PRESENTATION

A 53-year-old woman presented with a vascular lesion on the lower right canine region ([figure 1](#)). She was referred to the oral and maxillofacial department under the suspected head and neck cancer referral pathway. Examination revealed an approximately 8 mm by 5 mm, purplish-red, exophytic, vascular lesion on the interdental gingival papillae between the lower right canine and lateral incisor teeth, extending buccolingually ([figure 2](#), [figure 3](#)). The lesion appeared to be surrounded by a peripheral ecchymotic ring, a clinical characteristic commonly seen in SHLM ([figure 1](#)).⁷

This slowly enlarging mass had been present for a few months and occasionally bled on brushing. There was no history of trauma to the area. She had an unremarkable medical history and was a non-smoker. Radiographs did not show any evidence of bone loss around the teeth.

The lesion was surgically excised shortly after initial presentation, and biopsy revealed a polypoid vascular lesion in keeping with a haemangioma. Interestingly, 5 months later, the patient returned with a large recurrence in the same location ([figure 4](#)). This was surgically excised a second time, and biopsy results confirmed a definitive diagnosis of a SHLM.

INVESTIGATIONS

Immunohistochemistry results from the biopsy of the recurring lesion revealed that the cells lining the vascular spaces were positive for endothelial markers CD31, CD34, Factor VIII and D2-40 and

Figure 1 Buccal view of lesion, in between the canine and lateral incisor region.

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Figure 2 Buccolingual extension of lesion, showing the full mandibular arch.

were negative for HHV8. Histopathologically, the lesion revealed variably sized thin-walled anastomosing vessels lined by endothelial cells, which were seen to bulge into the vascular spaces in places, in keeping with the distinctive hobnail appearance.

DIFFERENTIAL DIAGNOSIS

SHLM can mimic other lesions clinically, radiographically and histopathologically; these include progressive lymphangioma, Kaposi sarcoma, microvenular haemangioma and retiform haemangioendothelioma.¹ Therefore, accurate diagnosis is essential in order to deliver the most appropriate treatment.

Although the entities mentioned have several qualities in common, there are certain histological features that can separate them from SHLM. The characteristic histopathological features of SHLM include thin-walled vessels lined with focally hobnailed endothelial cells, which are not seen in microvenular haemangioma, progressive lymphangioma and Kaposi sarcoma. Furthermore, the latter two lack the haemosiderin deposition that is common in SHLM.¹ Retiform haemangioendothelioma and SHLM, on the other hand, do share the characteristic histopathological feature of vascular spaces lined by hobnail endothelial cells. However, retiform haemangioendothelioma exhibits the distinctive morphological feature of long, arborising, thin-walled vascular spaces that infiltrate into a retiform pattern, which is not seen in SHLM.⁸ In addition, SHLM lacks

Figure 3 Buccolingual extension of lesion.

Figure 4 Recurrence 5 months after initial excision of lesion. Lingual view.

Learning points

- ▶ Superficial haemosiderotic lymphovascular malformation can resemble other benign and malignant tumours clinically and histopathologically.
- ▶ Precise diagnosis is important because it may influence treatment and prognosis.
- ▶ Accurate history, clinical examination and biopsy by an experienced clinician are essential to establish a conclusive diagnosis.
- ▶ Incomplete excision results in recurrence.

the prominent lymphoid infiltrate which is typically present in retiform haemangioendothelioma.¹

This patient initially presented with what, based on the clinical findings, was thought to be a pyogenic granuloma or a vascular lesion in keeping with a haemangioma.

As previously alluded to, the initial lesion was not correctly identified either clinically or histopathologically. Reasons accounting for this potentially false-negative result include using a suboptimal biopsy technique, inadequate size and depth of biopsy, tissue not taken from the representative site and/or incorrect handling of the sample.

TREATMENT

Complete surgical excision or laser ablation can be effective. Incomplete excision can result in recurrence of lesion.

OUTCOME AND FOLLOW-UP

Patient remains free of recurrence 14 months following second excision.

DISCUSSION

The treatment of SHLM varies depending on the clinical features and the anatomic considerations, although surgical excision is generally the treatment of choice.

Delineating SHLM from other vascular lesions is important. For this reason, an accurate biopsy is needed to facilitate a conclusive histological diagnosis to be reached.

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