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Short communication

Glomangiopericytoma-type glomus tumour/myopericytoma of the lip

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Abstract

We describe the case of a 46-year-old man who presented with a four-month history of a reddish, otherwise asymptomatic, nodular lesion that involved the mucosal side of the upper lip. The lesion consisted of myoid glomus-like cells arranged around branching and thin-walled haemangiopericytoma-like vascular structures, and was classified as glomangiopericytoma. No recurrences occurred during an eight-month follow-up. To the best of our knowledge, this tumour has never previously been described in the mucosa of the lip.

Keywords: Glomus tumour; Myopericytoma; Glomangiopericytoma; Oral cavity; Lip

Introduction

The World Health Organization[1] includes glomus tumours and myopericytoma in the category of pericytic (perivascular) tumours. While glomus tumours are defined as "mesenchymal neoplasms composed of cells that closely resemble the modified smooth muscle cells of the normal glomus body", myopericytoma "forms a morphological spectrum with myofibroma as well as with so-called infantile haemangiopericytoma, angioleiomyoma, and glomus tumour".[1] Pericytic (perivascular) tumours can arise at any age, usually in the superficial soft tissues, are rarely multiple, and exceptionally malignant.[1–4] Local trauma has been associated with their development, also in the oral cavity, where, however, these tumours are not common.[5–8]

Haemangiopericytoma-like vasculature can be present in glomus tumours, and glomus-like cells can be found in myopericytoma; the term glomangiopericytoma is currently used to identify these hybrid tumours.[1–5]

Case report

A 46-year-old man presented with a four-month history of a reddish, asymptomatic, soft tissue swelling of the mucosa of the upper lip. The overlying skin and rest of the oral cavity looked normal, and there was no relevant medical history. The patient denied any local trauma. Preoperative diagnosis included haemangioma and angiofibroma. The nodule was completely excised under local anaesthesia. Histological examination of the lesion showed a highly vascularised and cellular neoplasm with an overall pattern reminiscent of a haemangiopericytoma. Intralesional vascularisation consisted of variable-sized vessels with a predominance of branching and thin-walled vascular structures. Neoplastic cells were oval to round-shaped, showed centrally-placed nuclei, and were arranged around (and separated) the vascular spaces (Fig. 1). Stromal hyalinosis was detected focally. There was no mitosis, atypia, or necrosis.

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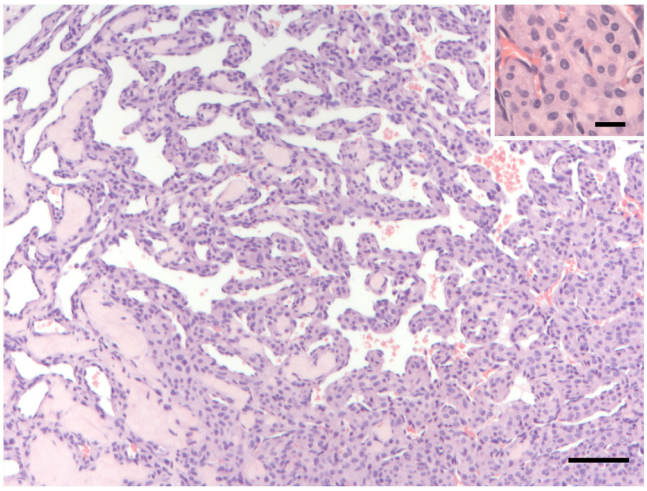

Fig. 1. Low-power magnification histological image of the tumour (bar = 200 μm). The insert highlights the perivascular glomus-like cells (haematoxylin and eosin stain, bar = 50 μm).

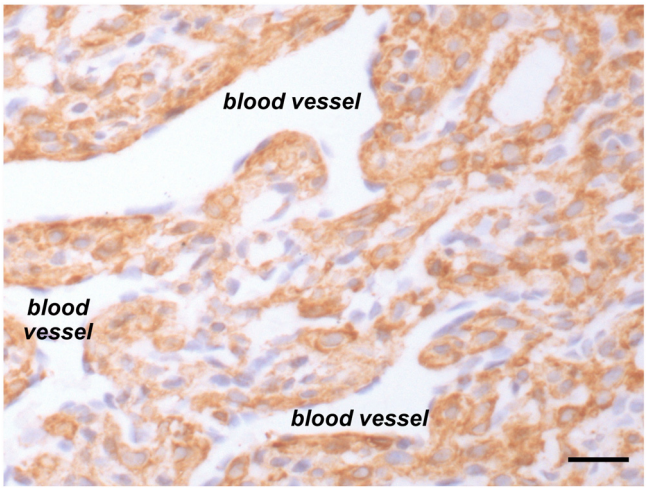

Fig. 2. Immunoreactivity of the perivascular glomus-like cells for smooth muscle actin (haematoxylin and eosin stain, bar = 100 μm).

Neoplastic cells stained for vimentin, smooth muscle actin (Fig. 2) and CD34, were surrounded by a collagen type IV-enriched "net-like" meshwork (Fig. 3), and were negative for S100, desmin, CD117, pan-cytokeratin, p63 and β-catenin. The proliferative activity (Ki67) was less than 5%. The pathological findings overall were consistent with a glomangiopericytoma-type glomus tumour/myopericytoma. At eight-month follow-up the patient had a well-healed surgical site and no recurrence.

Discussion

Glomangiopericytoma was first described in 1998 as a soft tissue tumour that was characterised by branching haemangiopericytoma-like vessels surrounded by glomus-like cells.[2] To date, the term glomangiopericytoma is also

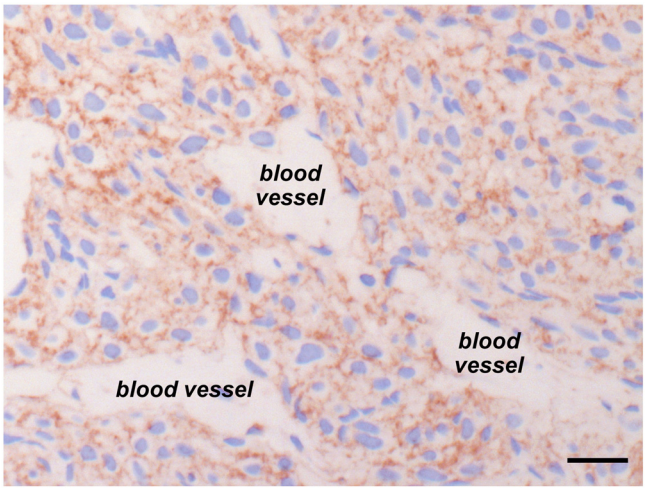

Fig. 3. Pericellular "net-like" immunoreactivity for collagen type IV (haematoxylin and eosin statin, bar = 100 μm).

used as a synonym for a sinonasal haemangiopericytoma-like tumour, a perithelial myoid tumour that is virtually specific to the sinonasal area [9,10] and differs from a glomangiopericytoma-type glomus tumour/myopericytoma in the nuclear expression of β-catenin as a result of gain-of-function *CTNNB1* mutations. [10]

Despite this, glomangiopericytoma-type glomus/myopericytoma is recognised as a specific histological entity,[1] and glomangiopericytoma-like areas can be detected in other pericytic (perivascular) tumours, including glomus tumours, myopericytomas, myofibromas, and angioleiomyomas.[1–8] As these tumours form a morphological continuum, this is not unexpected. Indeed, they share perithelial myoid differentiation and their overall morphological variations are thought to reflect the predominant differentiation of pluripotent perivascular cells along smooth muscle cells, pericytes, or glomus cells.[5]

In general, once perithelial myoid differentiation has been immunohistochemically established, the recognition of pericytic (perivascular) tumours does not pose much difficulty for pathologists. In our case, the absence of nuclear immunoreactivity for β-catenin in the perithelial myoid cells supported the diagnosis of glomangiopericytoma-type glomus tumours/myopericytomas[1,10] and their immunoreactivity for CD34 did not contrast with this conclusion.[1,4,8] In contrast, maxillofacial surgeons, dentists, and otolaryngologists might not be familiar with this family of neoplasms because of their non-specific gross appearance. In our case, for example, the lesion was misinterpreted as a vascular tumour.

A correct knowledge of glomangiopericytomas, and in general of the whole range of pericytic (perivascular) tumours, is important for appropriate treatment. These tumours are typically benign, and complete excision is generally curative. Local recurrences may occur when tumours have not been completely excised.[1–8] Malignant glomus tumours and myopericytomas have also been described as

glomus tumours of "uncertain malignant potential", the recognition of which is based on specific clinicopathological features including being >2 cm in size, deep location or superficial location with high mitotic activity or atypical mitosis, and the absence of nuclear atypia.[1,4,8]

Conclusions

In the range of extra-sinonasal pericytic tumours, the term glomangiopericytoma identifies glomus tumours with histological features of haemangiopericytoma/myopericytoma, and vice versa. We have described a glomangiopericytoma-type glomus tumour/myopericytoma that had arisen within the mucosa of the lip. To the best of our knowledge, this variant has never previously been reported at this site.

Conflict of interest

We have no conflicts of interest.

Ethics statement/confirmation of patient's permission

Ethics approval not required. The patient's permission has been obtained.

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