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Review

Venous malformations of the head and neck: current concepts in management

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Abstract

Low-flow venous malformations are congenital lesions and they are the third most common vascular anomaly in the head and neck. In this paper, the third in a series of three educational reviews, we discuss current trends in their management, and include a summary of common sclerosant agents used in their control.

Keywords: Vascular anomalies; Venous malformations; Sclerotherapy

Introduction

The management of vascular anomalies, a diverse collection of vascular malformations and tumours, has developed rapidly as knowledge has increased. The work of Mulliken and Glowacki[1] has enabled better understanding of the distinction between different clinical entities, and the development of a classification based on histopathological findings, which divides anomalies into vascular tumours and malformations (Fig. 1).[1–3] Concurrent advances in imaging and interventional radiological approaches have also improved treatment.

In this paper we discuss the clinical features and management of venous malformations. Although these are the most common vascular malformation in the body[4] (with a prevalence of 1%[5] and an incidence of 1 in 10 000[6]), they are only the third most common to present in the head and neck after infantile haemangiomas and lymphatic malformations.[7]

Diagnosis

History and examination

More than 40% of lesions are found in the head and neck,[1] most commonly in the muscles of mastication, and in the upper aerodigestive tract, submandibular triangle, and buccal space.[8] Anatomical planes do not restrict them, and they have the potential to arise in any location, tissue, or organ.

A thorough history and clinical examination will usually enable diagnosis and exclude potentially sinister conditions. The most common symptoms at presentation relate to the focal lesion and include intermittent pain and swelling caused

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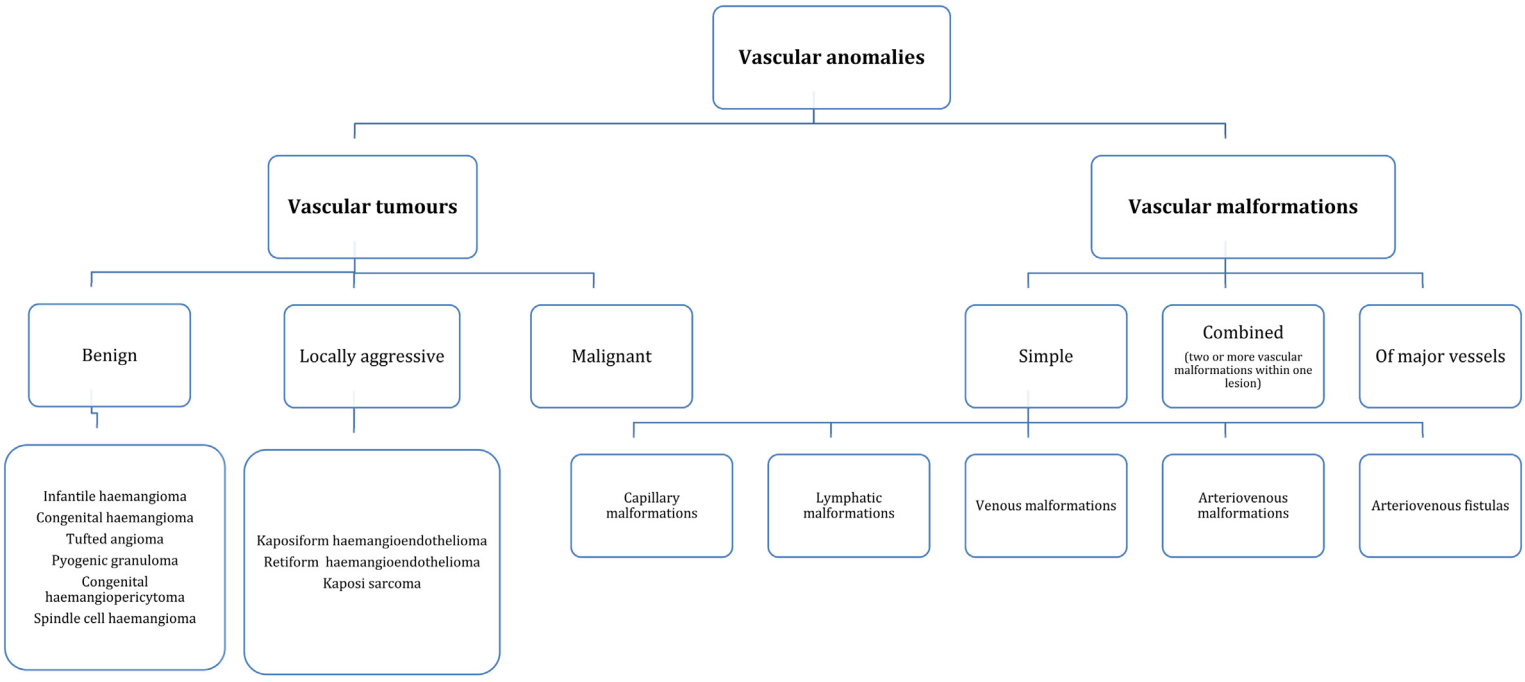

Fig. 1. International Society for the Study of Vascular Anomalies (ISSVA) classification of vascular anomalies.[2]

by intermittent microthromboses. Unlike haemangiomas, vascular malformations are present at birth. They grow with the patient, do not regress,[9] and develop steadily, although episodes of rapid progression are well known secondary to traumatic injury, infection, attempts at intervention,[1,10] and hormonal fluctuation.[11] Depending on their size and location, they may first be noticed in childhood, adolescence, or early adulthood. They often remain asymptomatic, although they can cause discomfort in the form of a dull ache which, depending on their position, can be worsened by physical activity and extremes in temperature. Long-standing lesions can lead to chronic pain through persistent venous stasis, formation of a thrombus, and local intravascular coagulation. This is a unique feature, as other vascular anomalies rarely present with pain.[7] Acute formation of a thrombus, which leads to pain and rapid enlargement is also recognised, and is sometimes the first presentation.[7] Involvement of the mucosal surfaces of the head and neck also leads to the other classic symptom, intermittent bleeding.[8]

Lesions typically present as soft, compressible, non-pulsatile masses with blue discolouration (Figs. 2 and 3). They typically expand with dependency when sited below the heart, and those within the skeletal muscles of mastication increase on clenching.[10] Initiation of a Valsalva manoeuvre will also increase their size, and this is a unique feature.[9] They range in size from small and asymptomatic varicosities to massive cervicofacial lesions that cause functional and aesthetic deformity.

The main differential diagnosis includes other low-flow vascular anomalies such as capillary and lymphatic malformations. Capillary malformations, which affect the capillaries of the papillary dermis, are the most important.[12]

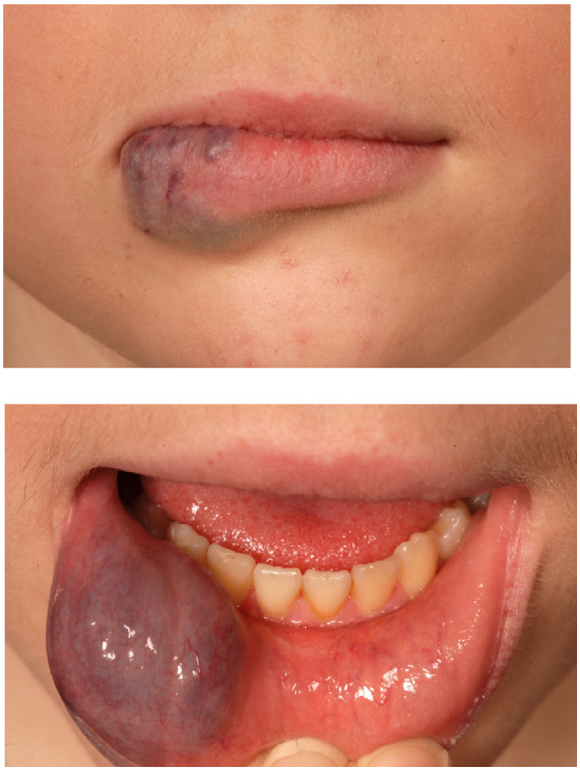

Fig. 2. Isolated venous malformation of the right lower lip (published with the patient's consent).

They are commonly called "port wine stains" and they present in a range of sizes and colours, most commonly pink or purple. They often arise on the face, and unlike venous malformations, tend to follow the branches of the trigeminal

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