Venous Malformations of the Head and Neck

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KEYWORDS

• Venous malformation • Head and neck • Vascular malformation

KEY POINTS

- Venous malformations (VMs) arise from deficits in the development of venous network, leading to dilated and dysfunctional venous channels that are deficient in smooth muscle cells.
- Clinical features of head and neck VMs are highly variable, ranging from small and asymptomatic varicosities to massive cervicofacial lesions.
- Several therapeutic approaches exist, including surgery; laser photocoagulation; sclerotherapy; and, more recently, systemic targeted drugs.

With an incidence of approximately 1 in 2,000 to 5,000, venous malformations (VMs) represent a vascular malformation frequently observed in specialized multidisciplinary centers.[1] They arise from deficits in the development of the venous network, leading to dilated and dysfunctional venous channels that are deficient in smooth muscle cells. These slow-flow venous sacs progressively expand with stagnation of venous blood. This results in growing lesions that do not spontaneously regress, and that ultimately infiltrate and compress normal adjacent tissues.[2,3]

More than 40% of VMs occur in the head and neck (H&N) region, representing, with infantile hemangiomas (IHs) and lymphatic malformations (LMs), the third most common vascular anomaly affecting this area.[4,5] Clinical features of H&N VMs are highly variable, ranging from small and asymptomatic varicosities to massive cervicofacial lesions. These VMs are not only disfiguring but also induce functional comorbidities with potential life-threatening complications. Several therapeutic approaches exist, including surgery; laser photocoagulation; sclerotherapy; and, more recently,

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systemic targeted drugs. Even though superficial and small VMs are successfully treated by single-treatment modality, management of deep and infiltrative VMs represents a medical challenge and requires a multidisciplinary approach with individualized treatment modalities.

NATURAL HISTORY AND SYMPTOMS

H&N VMs, as other VMs, are always present at birth but not always apparent. They grow with the child, slowly expand, and do not regress. The slow growth and deep location of some VMs can result in a late presentation during childhood, adolescence, or early adulthood. Environmental factors, such as traumatic injury, intervention, infection, or hormonal fluctuation (pregnancy or puberty) can exacerbate VM progression.[6–8]

More than 90% of VMs occur sporadically and consist of a unifocal lesion. Because VMs are able to arise in any location, tissue, or organ, H&N VMs can be well localized or extensive, superficial or deep (**Fig. 1**). They commonly involve buccal space, cheek, neck, eyelids, lips, parapharyngeal space, and submandibular triangle. Muscles of mastication, such as the temporal muscle, the masseter muscle, and the tongue muscles are also commonly affected. Other locations, such as pterygopalatine fossa and infratemporal fossa, may be affected, rendering the initial diagnosis difficult. Involvement of craniofacial skeleton can also occur, more commonly in the mandible and less commonly in maxilla, nasal, and cranial bones. Extensive H&N VMs diffusely spread along different tissue planes and ultimately involve adjacent structures, including skin, parotid gland, cervicofacial musculature, oral cavity, and respiratory and digestive tracts.[6,7,9]

Protrusion may be the only presenting symptom. Lesions typically present as a soft, compressible, and nonpulsatile mass with rapid refilling after compression. The overlying skin may appear normal or exhibit a bluish discoloration. Cutaneous involvement leads to a darker blue or purple discoloration. With time, calcified thrombi or phleboliths appear and are palpated within the venous mass.[10]

Symptoms are usually absent when the lesion is small or superficial; with expansion of the VM, symptoms appear, depending on the location and the mass effect on surrounding tissues and organs. Pain and swelling are observed in most patients. Sluggish flow progressively leads to increased blood stasis and alternated cycles of thrombosis and thrombolysis; that is, localized intravascular coagulopathy (LIC), which is responsible for chronic and recurrent pain. Physical activity, hormonal fluctuations (menstrual periods and puberty), extreme temperature, and Valsalva maneuver increase vein dilatation and exacerbate pain. Pain may also be more pronounced in the morning at awakening due to stasis and swelling. Acute pain should make suspect an acute venous thrombosis that leads to rapid enlargement of the malformation; this can sometimes be the first indication of a deep-seated H&N VM.[11,12]

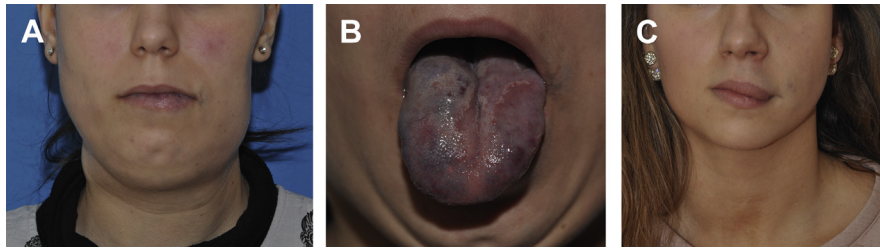

Fig. 1. Several aspects of facial VM. (*A*) Lower face involving the skin, mucosa, muscle and the tongue (*B*), causing facial asymmetry (*C*).