## Clinicopathological Case Report

# Management strategy for facial venous malformations

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### ····· ABSTRACT

Venous malformations (VMs) are slow-flow vascular malformations, caused by abnormalities in the development of the veins. Venous malformations vary in size and location within the body. When the skin or tissues just under the skin are affected, they appear as slightly blue-colored skin stains or swellings. These can vary in size from time to time because of swelling within the malformation. As these are vascular malformations, they are present at birth and grow proportionately with the child. Venous malformations can be very small to large in size, and sometimes, can involve a significant area within the body, When the venous malformation is well localized, this may cause localized swelling, however, when the venous malformation is more extensive, there may be more widespread swelling of the affected body part. Some patients with venous malformations have abnormal blood clotting within the malformation. Most venous malformations cause no life-threatening problems for patients. Some venous malformations cause repeated pain due to intermittent swelling and congestion of the malformation or due to the formation of blood clots within the malformation. Rarely, venous malformations may be part of a syndrome (an association of several clinically recognizable features) or be linked to an underlying genetic abnormality. We present 12 cases of venous malformations of the head and neck area, which have been managed at our hospital.

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#### Introduction

Vascular malformations (VMs) comprise of a spectrum of lesions involving all parts of the body. Mulliken and Glowachi, in 1982, proposed classification of the vascular abnormalities on the basis of cellular kinetics and clinical behavior. There are vascular tumors (lesions that arise from the endothelial hyperplasia) and vascular malformations (lesions that arise by dysmorphogenesis and exhibit normal endothelial turnover). A hemangioma is the most common vascular tumor, occurring on the skin of 4 to 10% of the infants. Vascular malformations are localized or diffuse errors of embryonic development at some stage of either vasculogenesis or angiogenesis.

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Most of these lesions are obvious at birth, while some are obvious during adolescences or adulthood. It is believed that enlargement is the result of changes in pressure and flow, ectasia, shunting, and collateral proliferation, rather than cellular proliferation. VMs have a tendency to grow with the child and after the individual has attained full growth, the VMs remain stable throughout life. Unlike hemangiomas, they do not regress, but some enlargement may occur in response to trauma or hormonal disturbances during puberty or pregnancy.[2] Vascular malformation classification was revised in 1992, during the meeting of the International Society of Vascular Anomalies, on the basis of histological appearance of the abnormal channel, flow characteristics, and clinical behavior.[3] They are classified as low-flow and high-flow vascular malformations. Venous malformations are characterized by a soft compressible, non-pulsatile tissue mass. The overlying skin usually has a bluish tint, but may be normal. The main locations are the head and neck (40%), trunk (20%), and extremities (40%). Although most venous malformations are in the skin and subcutaneous tissue, they often also involve the underlying muscle, bone, and abdominal viscera.

Treatment of a Venous Malformation depends upon the location of the lesion and its symptoms and complications. Asymptomatic venous malformations must not be treated by any interventional method. The patient must be aware of its worsening during puberty and pregnancy. Trauma must be avoided and extremity lesions must be supported by elastic stockings. Preoperative control of intravascular coagulopathy with low molecular weight heparin must be considered before resection of large venous malformations. [4] We report here our experience of venous malformations of the head and neck region, which were encountered and treated in the Department of General Surgery and Plastic Surgery.

#### MATERIALS AND METHODS

Twelve patients with venous malformations of the facial region were treated at the King George Medical University (KGMU) in the period between February 2013 and June 2014. There were five males and seven females). The patients' age at presentation, which ranged from four to seventy years. Eight patients (67%) had a vascular malformation at birth, during infancy. In six patients (50%) the majority of malformation



Figure 1: Venous malformation of the nose

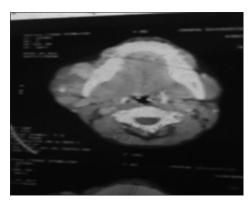


Figure 3: CT scan showing venous malformation involving the parotid muscle and parotid gland of the girl depicted in figure 2

occurred in the middle area of the face (cheek, nose, and upper lip). Two malformations (17%) occurred in the gingiva. One patient had a lesion involving the entire tongue. The interval between the onset and presentation varied from a few days to 17 years. It tended to present later in men than women. Some patient photographs are given below [Figures 1-9]. The most common presenting feature was facial disfigurement, with swelling, skin discoloration, and gingival bleeding. All the patients were evaluated preoperatively and a detailed history was taken with regard to the onset and progression of the lesions. The size and the site of the lesion were documented and associated symptoms and findings in the form of pain, ulceration or bleeding were also noted. All venous malformations were confirmed by color Doppler and CT angiography; one of the parotid region lesions has been depicted in Figure 3. Computed tomography was also done to exclude skeletal involvement in some of the patients. Sclerotherapy with sodium tetradecyl sulfate was done in all of the cases, except two, where direct excision was done. Direct excision was done in small and chronic lesions, where chronic phleboliths and inflammations led to fibrosis, with less compressibility and defined borders. Surgical



Figure 2: Venous malformation of the right parotid region



Figure 4: Venous malformation of the left upper eyelid



Figure 5: Venous malformation of the ear lobule



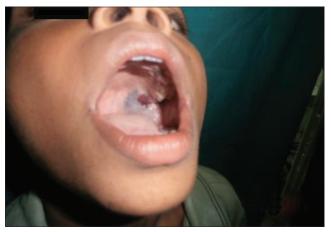


Figure 7: Venous malformation of the upper jaw



Figure 8: Venous malformation of the right mandibular area involving the neck



Figure 9: Venous malformation of the tongue

resection after sclerotherapy was performed in lip and nose lesions. Surgical resection was facilitated by subcutaneous infiltration of adrenaline in normal saline (1: 2, 00.000). Four patients required resection of a portion of the involved skin along with resection of the malformation. Tissue deficits were covered with a split skin graft (on the forehead) and the rotation of a skin flap (on the cheek). The lesions were successfully resected, with no significant blood loss. In two patients surgical resection was performed with sclerosant injections into the inaccessible portion of the malformation.

#### DISCUSSION

Venous malformations are soft, compressible, nonpulsatile masses of the skin and subcutaneous tissue. There are few deep soft and intramuscular lesions that often cause discomfort after exertion or at the end of the day. Intraoral lesions can bleed, distort dentition, cause speech problems or can obstruct the upper respiratory pathway.<sup>[2]</sup> The main problems with venous malformation is esthetic disfigurement, bleeding sometimes, and rarely localized intravascular coagulopathy (LIC).[3,4] The treatment of vascular malformation is really difficult. Only symptomatic patients must be treated. An esthetic problem is the prime indication for intervention. Other indications like pain, functional problems, bleeding, LIC, speech disturbances, and respiratory obstructive symptoms are less commonly encountered. Sclerotherapy is the mainstay of treatment and absolute alcohol and sodium tetradecyl sulfate are most commonly used as sclerosing agents. [5] We have used sodium tetradecyl sulfate in cases where sclerotherapy has been employed. This agent is less toxic than because of lower reported rates of skin necrosis, neurological complications, and systemic complications. [6] Anaphylactic reaction with sodium tetradecyl sulfate has been reported,[7] but was not seen

in our series. Sclerotherapy induces an inflammatory reaction that worsens the symptoms initially, but subsides gradually. Anti-inflammatories and analgesics were given during the course of sclerotherapy. Two to four sessions were needed for subsidence of the lesion to an optimal size. Venous anomalies have a propensity for recanalization and recurrence. Spongy patterns, especially when intramuscular, are more difficult to treat. We have found a better result in cavitary and dysmorphic vein patterns. Success rates of 30 to 95% have been described. In our series, all patients are doing well as there is a short follow-up and the failure rate and other interventional complications will be observed in long-term follow-ups.

Surgery is indicated only in those cases where residual lesions remain after sclerotherapy or when there is esthetic prejudice. [9] We have performed surgical resection in only two cases where small circumscribed lesions were present and were comparatively less compressible and easily accessible and disfiguring the face. Other therapeutic modes like laser therapy can be useful in superficial venous malformations and in oromucosal lesions. [10-12] Satisfactory minimal scarring, especially for discoloration has been reported, but recurrence and repeated treatments are common. The Nd YAG laser has deeper penetration and can be used for deeper lesions. For deeper lesions, laser probes can be inserted subcutaneously.

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