

DENTAL ARTICLE

Multiple Venous Malformations with Phleboliths: Radiological-Pathological Correlation

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ABSTRACT

Vascular malformations are congenital lesions that are present at birth and do not regress. However, they often present later in life. They are subdivided into two categories: (1) slow- or low-flow and (2) fast- or high-flow malformations. Low-flow malformations contain combinations of capillary, venous, and lymphatic components. Venous malformations can occur anywhere in the body, but are most frequently seen in the head and neck (40%). These lesions present in a variety of ways, from a vague blue patch to a soft blue mass, which may be single isolated or may occur in multiple areas. Treatment depends on the type of lesion, the location, degree of involvement, and the clinical symptoms. Here we are report the imaging and histopathologic findings in a patient with multiple venous malformations affecting the left side of the face and trunk.

Key words: Hemangiomas, magnetic resonance imaging, sclerotherapy, vascular malformations, venous malformations

INTRODUCTION

Venous malformations are part of a spectrum of vascular malformations. Venous malformation is defined as malformations comprised of slow-flowing, abnormal dilated veins, and venous network.^[1] Clinically venous

malformations (VM) are present at birth and tend to grow steadily in proportion to the somatic growth of the child. Venous malformations are congenital lesions that affect boys and girls equally with a reported risk of developing other conditions within a specified period of time. The occurrence rate is 1-2 per 10,000 births and 0.1-1% of a population are found to have this condition.^[2] Histologic and histochemical studies show that abnormalities are formed by small and large dysplastic post-capillary, thin-walled vascular channels with patchy deficiency of mural smooth muscle.^[3]

A 28-year-old male was referred to the oral and maxillofacial clinic for evaluation of multiple swellings on the left side

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of the face and inside his mouth. The patient first noticed the swelling over the temple when he was 11-years-old. As the boy grew, the swelling also proportionately increased. There were no associated symptoms.

On clinical examination, multiple poorly defined swellings were noted on the left side of the head and neck region around the temple, cheek, and submandibular region [Figure 1a and b]. Similar lesion was noted in the supraclavicular and scapular region [Figure 2a and b]. These lesions were approximately 3 × 2 cm in size. They had a slight purplish discoloration. The swelling was pulsatile and increased in size when the patient was standing (dependent position). On palpation, these lesions were found to be soft and easily compressible. Intraorally lobulated swelling was noted on the floor of the mouth in the left sublingual region. The lesion measured 3 × 4 cm, filling the entire floor of mouth, and purplish discoloration was also noted over the swelling [Figure 3a and b]. Venous channels became engorged in the dependent position. On palpation, these lesions were soft in consistency and readily flattened on compression.

RADIOLOGIC FEATURES

X-ray of paranasal sinus showed no involvement of the bone. Multiple small phleboliths were noted in the left submandibular region. Ultrasonography of the



Figure 1: 28-year-old male with multiple swellings on the left side of the face. (a) Photograph (front view) shows swelling over the temple region and (b) Photograph (side view) reveals well defined swelling over temple (large arrow) diffused swelling in the cheek, and submandibular region (small arrow).

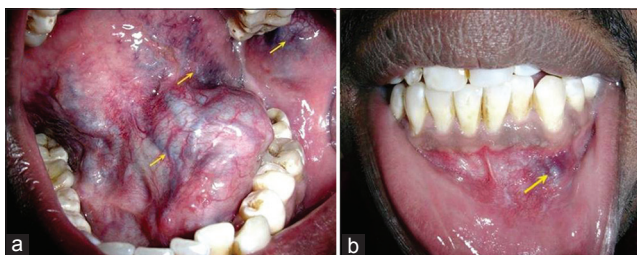


Figure 3: 28-year-old male with multiple swellings on the left side of the face. (a) Intraoral photograph shows swelling in the floor of mouth filling the entire floor of mouth (arrow) and a small swelling in the buccal mucosa that has a purplish hue (small arrow). (b) Intraoral photograph of the labial sulcus reveals a purplish swelling (arrow).

submandibular region showed a well-defined hypoechoic lesion having anechoic areas with septations, showing flow inside the lesion. Small hyperechoic foci of calcifications were noted [Figure 4].

Magnetic resonance imaging (MRI)-T2-weighted fat suppression, post-contrast axial images showed hyperintense lesions, which are well-defined with no flow voids. These lesions had homogeneous contrast enhancement. All the lesions in the submandibular, cheek, and temple region had similar findings [Figure 5]. Direct puncture phlebogram of the left frontal scalp region revealed pooling of contrast into cavernous spaces draining into external jugular vein [Figure 6].

PATHOLOGIC FEATURES

Incisional biopsy was performed from the lower lip lesion. The specimen obtained was stained using hematoxylin and eosin stain. Histopathological examination revealed several thin-walled venous channels lined by flattened endothelium, supported by a dense uninfamed fibrous connective tissue stroma. These features were suggestive of venous malformation [Figure 7].

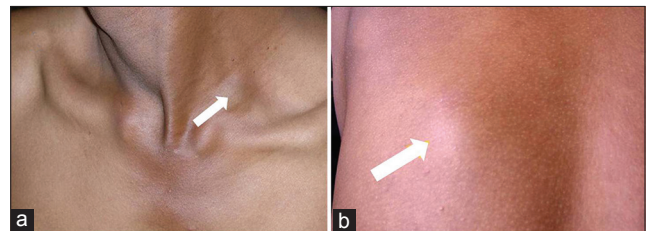


Figure 2: 28-year-old male with multiple swellings on the left side of the face. (a) Photograph shows a well-defined swelling measuring 1 × 1 cm in the left supraclavicular region and (b) Photograph of the scapular region shows a diffused swelling.

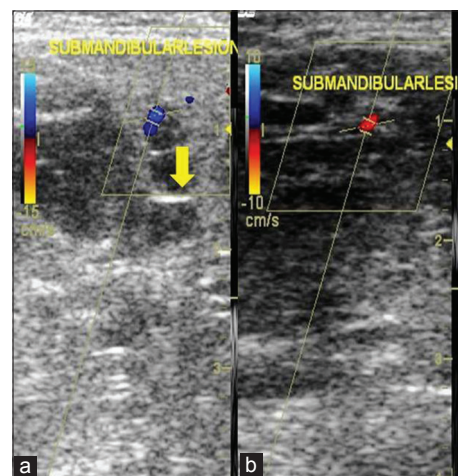


Figure 4: 28-year-old male with multiple swellings on the left side of the face. Ultrasonography of the submandibular region with 7 MHz small parts transducer shows heterogeneous hypoechoic lesion with slow flow and hyperechoic foci suggestive of calcifications (arrow).

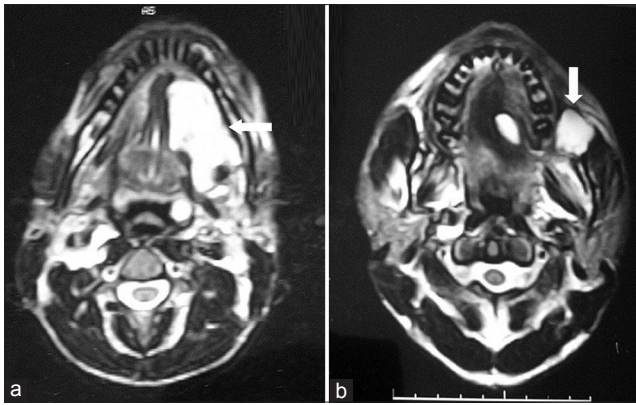


Figure 5: 28-year-old male with multiple swellings on the left side of the face. T2-weighted magnetic resonance imaging. (a) Axial section at the submandibular region shows well-defined hyperintense lesion (arrow) in the floor of mouth. (b) Axial section at the level of cheek shows well-defined hyperintense lesion in the cheek region (arrow).

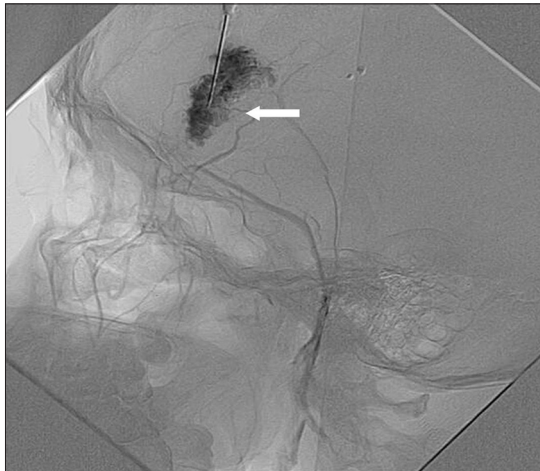


Figure 6: 28-year-old male with multiple swellings on the left side of the face. Phlebography of the lesion in the temple region shows filling of the entire lesion and draining to the regional area (arrow).

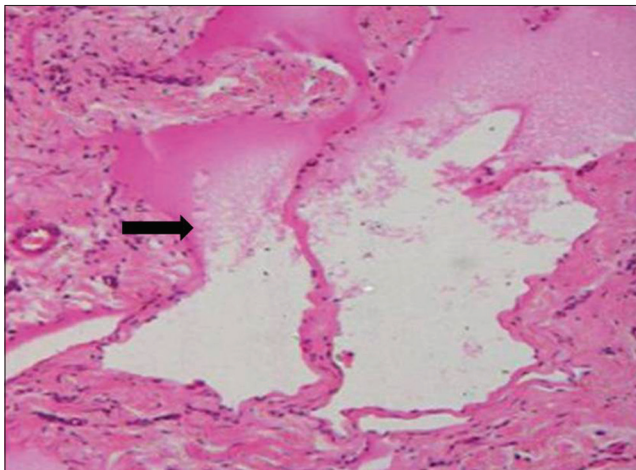


Figure 7: 28-year-old male with multiple swellings on the left side of the face. Hematoxylin and eosin stained specimen ($\times 40$) shows irregular venous channels (large arrow) lined by thin endothelium.

DISCUSSION

Venous malformation (VM) is the second most common vascular anomaly of the head and neck after hemangioma.^[2] Venous malformations can occur anywhere in the body but are most frequently seen in the head and neck (40%), extremities (40%), and trunk (20%). VMs are slow-flow vascular anomalies composed of ectatic venous channels that will continue to grow throughout the patient's lifetime. They grow slowly in size with age, but their growth may be exacerbated following trauma, sepsis, or hormonal changes and they do not regress spontaneously. Both men and women are equally affected.^[4]

Vascular malformations are believed to be the result of a congenital error of vascular morphogenesis that occurs between the 4th and 10th weeks of intrauterine life.^[5] Vascular malformations have a quiescent endothelium and are considered to be localized defects of vascular morphogenesis, likely to be caused by dysfunction in pathways regulating embryogenesis and vasculogenesis.^[3]

These lesions vary in color depending on depth of involvement and range from mild detectable color change to deep purple color.^[5] These lesions fill when the patient is standing and are compressible, which helps to distinguish them from lymphatic malformations on physical examination.^[6] Areas frequently involved in the head and neck region are masseter, temporalis, tongue musculature, as well as oral and airway mucosa.^[3] Soft tissue lesions are most frequently facial in location, with the buccal region being the most common site followed by the mandibular space, sublingual space, tongue, and orbit.^[4] Intraosseous calvarial involvement is most frequent in the frontoparietal region and the mandible is the most frequent location within the facial skeleton. There may, however, be no visible manifestations with deeper lesions.^[2] In our case, multiple VMs were present, which was not associated with any other syndrome.

The first anatomopathologic classification of vascular lesions based on the microscopic appearance was developed by Virchow and Wegner. They classified vascular lesions into simple, cavernous, and racemic types.^[4] Mulliken and Glowacki (1982) developed a biological classification of vascular anomalies that included physical findings, clinical behavior, and cellular kinetics and classified them as hemangiomas and VMs.^[6] A more recently updated classification of vascular anomalies by International Society for the Study of Vascular Anomalies is now widely used [Table 1].^[7]

Table 1: Classification of vascular anomalies (ISSVA 2007)

| Vascular tumors | Vascular malformations |
|--|---|
| Infantile hemangiomas | Slow-flow vascular malformations |
| Congenital hemangiomas | Capillary malformation (CM) |
| Tufted angioma | Venous malformation (VM) |
| Kaposiform | Lymphatic malformation (LM) |
| Spindle cell hemangioendothelioma | Fast-flow vascular malformations |
| Other, rare hemangioendotheliomas (epithelioid, composite, retiform, polymorphous, etc.) | Arterial malformation (AM) |
| Dermatologic acquired vascular tumors | Arteriovenous fistula (AVF) |
| | Arteriovenous malformation (AVM) |
| | Complex-combined vascular malformations |
| | CVM, CLM, LVM, CLVM, AVM-LM |

ISSVA: International society for the study of vascular anomalies, CVM: Capillary venous malformation, CLM: Capillary lymphatic malformation, LVM: Lymphatic venous malformation, CLVM: Capillary lymphatic venous malformation, AVM-LM: Arteriovenous malformation-lymphatic malformation

Plain radiographs have limited role in the investigations as they can identify only phleboliths.^[8] Ultrasound (US) is often the initial investigation to evaluate vascular malformations and it may characterize and define the extent of more superficial lesions. On gray-scale imaging, venous malformations can appear as hypoechoic or heterogenous lesions with anechoic structures visible in 50% of cases. In addition, the Doppler flow is generally monophasic low velocity flow, and in some cases flow is only discernible with compression and release of the lesion.^[9] Phleboliths that confirm the diagnosis of VMs may be detected.^[8]

Venous malformations on MRI are very well-discerned. They typically appear as isointense or hypointense lesions on T1-weighted images, but could be hyperintense depending on the presence of intralesional fat. Lesions are typically lobulated, which gives them the characteristic “bunch of grapes” configuration. Septations and rounded signal voids corresponding to phleboliths are additional distinguishing features.^[10]

In T2-weighted or inversion recovery sequences, VMs demonstrate high-signal intensity. This imaging modality is used to determine the full extent of the lesion and its relationship to adjacent vital structures. Gradient echo sequences reveal areas of low signal corresponding to calcification or hemosiderin or thrombosis. T1-weighted post contrast imaging demonstrates homogenous or heterogeneous enhancement, and dynamic contrast-enhanced MRI has increased the specificity of venous malformation diagnosis.^[9]

VMs are best demonstrated by direct phlebography, which fills the sinusoidal spaces and any anomalous veins, allowing assessment of the size and extent of the lesion.^[3]

In the present study, phleboliths were seen on plain radiography and US features were suggestive of vascular anomaly with slow flow rate. Whereas, the MRI features were suggestive of venous malformation, which was confirmed by direct phlebography where the lesion was seen draining the regional vein. These investigation features were suggestive of venous malformation. Venous malformations usually are associated with syndromes like Proteus syndrome and Blue rubber bleb nevus (Bean) syndrome.

Multiple treatment options exist for venous malformations, including conservative measures such as head of bed elevation and compression, laser therapy, sclerotherapy, and surgery.^[2] Conservative management of venous malformations is usually reserved for smaller isolated asymptomatic lesions and is also important in controlling the growth and symptoms.

Elevation of the head of the bed is important as it decrease hydrostatic pressure in the malformation, which can lead to expansion and can also decrease symptoms of airway obstruction, swelling, and pain that are experienced.^[1]

Laser therapy is a mainstay of management of mucosal and skin malformations.^[5]

Sclerotherapy remains a good option for the treatment of venous malformations in the head and neck. Sclerotherapy involves percutaneous injection of a substance to induce inflammation and thrombosis of the lesion, which then will lead to more long-term fibrosis and hopefully decrease or eliminate the expansion of the lesion. Sotradecol foam or ethibloc (glue), or the sclerosant is mixed with fibrin glue or ethyl cellulose, bleomycin (pingyangmycin) and picibanil (OK-432) have recently been used as sclerosants in Asia with promising results.^[10]

Large cervicofacial venous malformations present a much greater challenge, and one must be prepared to use multimodal therapy to keep the lesion under control. These lesions generally cannot be cured as doing so would leave devastating functional and cosmetic results. Therefore, therapy is used to control growth, maintain cosmesis, and decrease symptoms.^[5]

CONCLUSION

Venous malformations are either superficial or deep veins that are abnormally formed and dilated. A thorough

examination and investigation of the condition is needed to establish the exact extension of the condition and plan proper treatment.

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