Case Report

Giant arteriovenous malformation of the face and upper lip

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Abstract

A 35-year-old male patient reported with the chief complaint of a large swelling in the right side of the upper lip. The lesion measured 6.5 cm measuring from the infraorbital region to the lower border of mandible on the right side of the face and 7.5 cm from the right ear lobule to the contralateral commissure of the left side of the face. On palpation, the swelling was soft, fluctuant and compressible. Pulsations were felt, and on auscultation, bruit was also heard. Computed tomography angiogram of the neck and circle of Willis showed serpiginous hyperdense vascular channels causing significant soft-tissue thickening of the upper lip, right cheek region and philtrum. Hence, the diagnosis of diffuse subcutaneous facial arteriovenous malformation involving the right cheek and philtrum was given. The entire lesion was excised. In the postoperative 2nd month, secondary cosmetic correction or lip reconstruction was done. The patient was reviewed after 3 years; there was no recurrence of the lesion.

Keywords: Arteriovenous malformation, computed tomography angiography, face, upper lip, vascular

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INTRODUCTION

Anomalies of vascular tissue are considered to be a group of lesions originated from blood vessels and lymphatics which show extensively varying histopathology and clinical behavior. These vascular anomalies represent the most frequent inherited abnormalities in infants and children. Long ago, the difference between the true hemangioma and the less common vascular malformation was first recognized by James Wardrop, a London surgeon, in 1818.[1]

Vascular tumors and vascular malformations show huge differences in clinical, radiological and histological appearance. Thus, vascular tumors are endothelial neoplasms characterized by increased cellular proliferation. On the other hand, vascular malformations are the result of abnormal development of vascular elements during embryogenesis and fetal life. In disparity to vascular tumors, vascular malformations do not have a growth phase or an involution phase. Vascular malformations tend to grow proportionately with the child, never regresses, but persist throughout life.[2]

The classification of malformation is based on the clinical, radiological and histological appearance of the abnormal channels, which may be either hematic or lymphatic in nature. A vascular malformation can be slow flow (that is, capillary, lymphatic or venous) or fast flow (that is, arterial). If there are combinations of these elements, the malformation is called an arteriovenous malformation (AVM), lymphaticovenous malformation (LVM) or capillary LVM (CLVM).[3]

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The diagnosis of this group of lesions primarily depends on the history of the lesion and the clinical presentation. Radiographic evaluation may be helpful in determining the exact extent, location and flow dynamics of some lesions.[1] The present case report describes AVM of the face and upper lip in a 35-year-old male.

CASE REPORT

A 35-year-old male patient reported with the chief complaint of a large swelling in the right side of the upper lip. The swelling was noticed by his parents at the age of 10 years, which was the size of a peanut. The patient underwent multiple treatments including intralesional injections and surgical excision for the same in the past at other centers with failures.

When the patient reported to the department, the lesion measured 6.5 cm measuring from the infraorbital region to the lower border of mandible on the right side of the face and 7.5 cm from the right ear lobule to the contralateral commissure of the left side of the face. The thickness of the lesion was variable with maximum thickness of 5 cm in the region of right side of the upper lip. The overlying skin appeared nodular over the swelling, and multiple intraoral mucosal telangiectasias were visible. On palpation, the swelling was soft, fluctuant and compressible. Pulsations were felt, and on auscultation, bruit was also heard. Stone-hard nodules were also palpated on the superficial aspect of the lesion, suggestive of phleboliths [Figure 1].

Computed tomography (CT) with angiography was done. CT angiogram of the neck and circle of Willis showed serpiginous hyperdense vascular channels causing significant soft-tissue thickening of the upper lip, right cheek region and philtrum [Figure 2]. Postcontrast, there is an avid enhancement. Feeding arteries appear to arise from maxillary and facial branches of the right external carotid artery. Draining vein is seen draining into the right internal jugular vein. There is early opacification of the right internal jugular vein. There is also a feeding artery from maxillary branch of the left external carotid artery. Hence, the diagnosis of diffuse subcutaneous facial AVM involving the right cheek and philtrum was given.

The patient underwent surgery as the principle modality of treatment. The surgery was planned in two phases, in the first phase ligation of right facial artery, external jugular vein; and the left facial artery was performed. This resulted in decrease in the blood volume and shrinkage of the lesion by 2 cm in overall dimensions. One unit of blood was transfused before the surgery and one unit intraoperatively. In the second phase, upper labial incision was placed along the vermillion border, bleeders were identified and circumferential ligation was done using 4'0 vicryl. Around 100 circumferential ligating sutures placed. Orbicularis oris muscle reduction was done minimally; debulking of excess tissue was done. The entire lesion was excised. In the postoperative 2nd month, secondary cosmetic correction or lip reconstruction was done.

Histopathological findings include stratified squamous epithelium and fibrous connective tissue infiltrated by neutrophils, lymphocytes and mast cells. Deeper down connective tissue showed large vascular channels of varying sizes, thick and thin walled, some empty and interspaced with areas of hemorrhage [Figure 3]. Based on clinical, radiographic and histopathological findings, the lesion was diagnosed as AV malformation. The patient was reviewed after 3 years; there was no recurrence of the lesion [Figure 4].

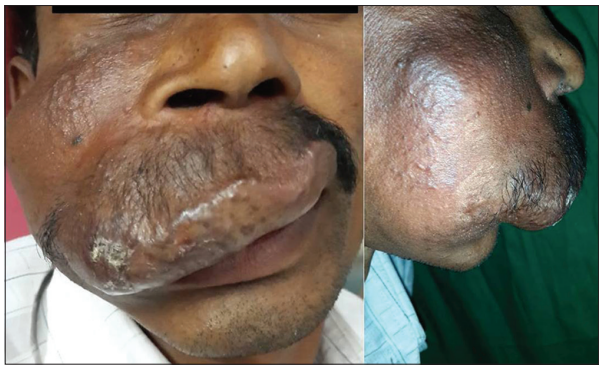

Figure 1: Giant arteriovenous malformation involving the upper lip and right side of the face

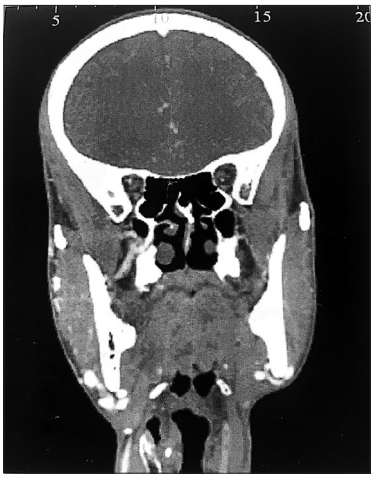

Figure 2: Computed tomography angiography of the neck and circle of Willis

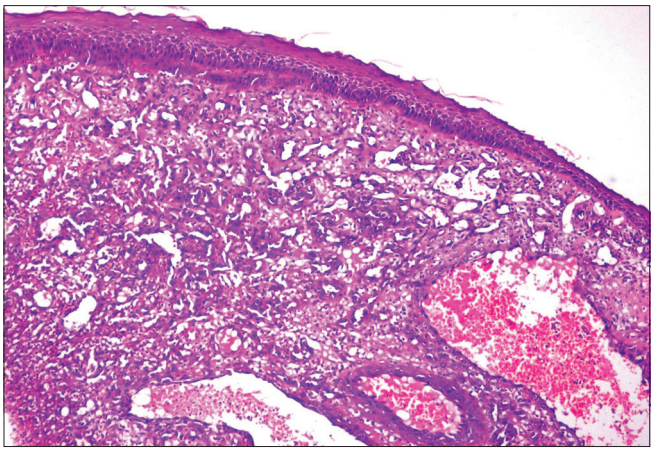

Figure 3: Connective tissue with large vascular channels of varying sizes, thick and thin walled, some empty and interspaced among areas of hemorrhage (H and E)

DISCUSSION

Vascular malformations are less frequently seen in about 0.5% of all newborns. These lesions result from abnormal blood/lymphatic vessel morphogenesis. Histopathologically vascular malformations are characterized by normal endothelial cells and normal number of mast cells throughout their history. They are present at birth but may not become clinically apparent, until late infancy/childhood. Under the influence of hormones, trauma and sepsis, changes may occur. Their growth is commensurate with that of the patient, and they do not involute.[1]

An AVM of the face/scalp is an abnormal fistulous connection between the feeding artery and the draining veins without an intervening capillary bed within the subcutaneous layer. The draining veins are grossly dilated and tortuous and may show variceal dilatation.[4]

AVM signs and symptoms vary between asymptomatic lesions to pulsatile swelling, throbbing pain and uncontrollable hemorrhage following trauma or tooth extraction and cardiac failure. When the lesions are small and asymptomatic, close observation is sufficient.

However, with continuing growth of AVMs, signs and symptoms outbreak and result in cosmetic and functional problems, which necessitate active treatment.[5]

Magnetic resonance imaging is the investigation of choice as it provides accurate information about the extent of the lesion and better contrast between the different types of vascular anomalies. Angiography is usually reserved for therapeutic endovascular interventions. Direct intralesional injection of contrast medium may have a role in the analysis of venous malformations.[6] Thus, in the present case, CT with angiography was done.

Figure 4: A 3-year follow-up visit after surgery showing no recurrence

Surgical excision of small vascular growths is possible without undue risk of hemorrhage or of causing any cosmetic or functional deficiency, whereas total resection of large, localized deforming vascular lesions involves the possibility of massive hemorrhage during surgery and the need for major reconstructive procedures after resection.[4] With our case, surgical debulking for lip swelling was done, and 3-year follow-up showed no recurrence.

CONCLUSION

Correct classification and diagnosis of vascular anomalies is imperative to accurately ascertain prognosis and direct treatment. Multimodal therapy is frequently indicated, and in complex patients, a referral to a multidisciplinary vascular anomaly team should be considered.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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