CASE REPORT

Vascular Malformation of Lower Limb- A Diagnostic Dilemma

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Abstract

Intramuscular venous malformations are rare presentations and when occur they are most commonly found in the head and neck and extremities but are relatively rare in the trunk, they are well localized to a single muscle or adjacent muscle groups. We present here a case report of patient who presented with venous malformation confined to the lower extremity which was successfully treated with surgical excision.

Key Words

Vascular malformation, Venous, Lower limb, Surgical excision

Introduction

Venous malformation is the most common congenital vascular malformation. It is noted in 1-4% of total patients presenting with vascular malformations. There is bluish discoloration of the skin, which is compressible and soft on examination. They are also dependent lesions, as a result of which it expands or contracts depending on the position of the patient. The malformation is noted in up to 40% of cases in the skeletal muscles. Vascular malformations of the skin manifest much earlier than the vascular malformations of the muscles. Patients present with pain and swelling of the muscle. This progressively increases, because there is proportional increase in the size of the malformation with the growth of the patient. This worsens when the patient attains puberty, has hormonal changes or has an infection. This worsens the symptoms (1-2).

These venous malformations are of different morphologies with abnormal venous channels, adipose tissue and degenerative muscle matrix. Tissue planes are crossed and enter adjacent tissue (3). Most venous malformations are misdiagnosed as haemangiomas. Doppler ultrasonography and Magnetic resonance imaging have been used for the diagnosis of the condition and its characterization.

These malformations could be treated by multiple methods. Excision of the malformation by surgery, using ablation, or sclerotherapy has been mentioned in the treatment of venous malformations (4). The purpose of this case report is to understand the method of diagnosis and its surgical management.

Case Report

A 19-year-old presented to the OPD with complaints of swelling in the anterior aspect of left thigh for 4-5 years. There was no history of trauma or any other medical history of any disease related to swelling. On examination it was diffuse swelling over the anterior aspect of left thigh above the knee, swelling was irregular, non-tender, diffuse, soft in consistency, non-pulsatile in nature (Fig. 1a). There was no evidence of neurovascular deficit.

Laboratory investigations were normal other than anemia (Hb, 7gm %). Specific investigations were done in the form of ultrasound Doppler left lower limb which showed dilated venous channels along the left anterior and medial aspect of left thigh and left suprapatellar region suggestive of venous malformation. MRI of left leg revealed large, ill-defined, infiltrative type of lesion in the antero-medial compartment of left thigh, suggestive of vascular malformation, probably hemangioma (Fig. 1b,c).

Surgical excision of the venous malformation was done (Fig. 2) and specimen was sent for the histopathological examination which reported it as hemangioma. Postoperative period of the patient was uneventful. Patient was discharged in satisfactory condition and is on regular follow up with us in the Out Patient Department.

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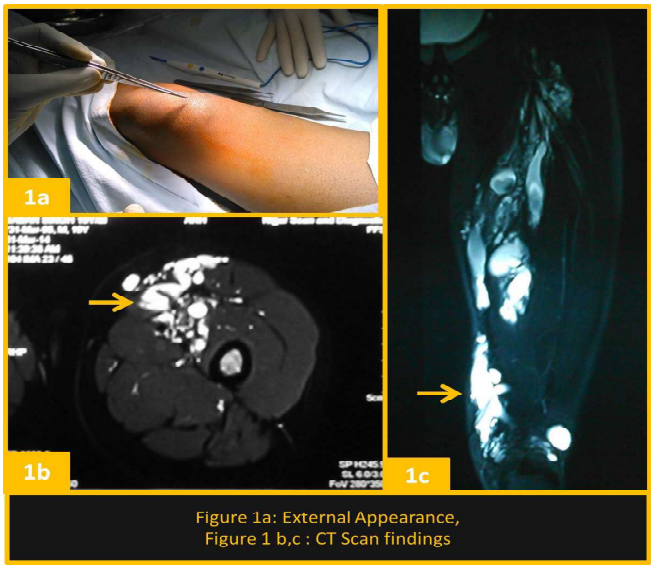

Figure 1: a) External Appearance; b&c) CT Scan Findings

Figure 2: a) Before Excision; b) After Excision; c) Feeder Artery

Discussion

Vascular malformations are congenital morphogenic anomalies of various vessels that can present at any age (5). Descriptions are often confusing with terms such as venous angioma, cavernous angioma or haemangioma being used. According to the International society for the study of vascular anomalies, vascular anomalies are classified into either vascular tumours (mostly haemangiomas) or vascular malformations (6-7). Vascular malformations have a different origin. They are rare congenital lesions caused by abnormal embryonic development of vascular structure. Although these lesions are always present at birth, they may not be visible until week or even years after birth (8). These lesions will typically grow in proportion to the growth of the child, meaning they tend to grow steadily throughout life. They are subdivided into 2 categories: slow-or-low flow and fast-or high flow malformations (5,6). Low flow malformations are further subdivided into arteriovenous, capillary, venous, lymphatic, and combined malformation. Among them, venous malformation is the most common form and they are mostly in skin and subcutaneous tissues.

In our case report, it was an intra-muscular venous malformation of the lower limb which sometimes can cause a diagnostic dilemma. The venous portions of malformations contain abnormally formed and dilated superficial or deep veins, the wall of which are thin because they lack smooth muscle. The clinical presentation of low-flow vascular malformations varies because of the many combinations of venous and lymphatic components, as well as variation in size and location. Because venous malformations are lesions due to abnormal embryonic development, it is assumed that localized venous malformations result from insults of the specific neurovascular bundles during development, which is the origin of localized vessels and muscles.

Although venous malformations most often involve the face, limbs and trunk, they may be found within the internal viscera, bones and skeletal muscle. In our patient, a venous malformation was noted intramuscularly. Venous malformation is commonly confused with hemangiomas in adult, especially in the liver (9). Typical venous malformations are grossly detectable and easily diagnosed by skin color changes, focal edema, asymmetry of muscles or pain. venous malformations in the skeletal muscles, however, has the potential to be missed because their involved sites are invisible and the disease is rare. These lesions are diagnosed by both physical examination and by using a number of imaging techniques, including magnetic resonance imaging, computed tomography, and ultrasonography, in some cases, an angiogram is needed to assist in detailed treatment planning.

Due to the rarity of vascular malformations, experience in diagnosis and treatment is limited. This may lead to misdiagnosis and a poor outcome. Vascular malformations are generally treated in specialized centers by a multidisciplinary team and are determined by the extent

and location of the lesions. Although many are managed expectantly, some require treatment because they are painful or may be, they are present in a location where they can cause significant morbidity. Therapeutic options ranges from observation, sclerotherapy to finally, surgical excision (10). Our patient underwent surgical excision. Recurrence, focal fibrosis or contractures following surgery is more common with diffuse venous malformations. Due to frequent infiltrative, widespread and unpredictable nature, treatement for vascular malformation is often challenging and interesting.

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