Venolymphatic malformation of vastus medialis—case report

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ABSTRACT

Background: Venolymphatic malformations (VLMs) are combination of venous and lymphatic components of low flow type of category. These malformations do not connect to the main channels directly. These present in the form of swelling and other characteristics as per the type of malformation. The diagnostic evaluation becomes more baffling if the overlying skin is normal.

Case Presentation: We present a 55-year-old female who had swelling on the medial part of left knee which was causing some pain only during movements. The evaluation of the entity was done by ultrasound, color flow imaging (CFI), and magnetic resonance imaging (MRI). The diagnosis of VLM of vastus medialis was confirmed.

Conclusion: VLM swellings of extremities can be diagnosed with confidence with CFI and MRI diagnostic modalities.

Keywords: Venolymphatic malformations, low flow, ultrasound, CFI, MRI.

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Background

Venolymphatic malformations (VLMs) are of congenital in origin because of abnormal growth of the embryonic vascular tissue. The nomenclature was given as lymphangiohemangiomas or hemangiolymphangiomas [1].

Case Report

Fifty-five-year-old female reported with the complaints of pain and swelling on the medial aspect of the left upper leg. The pain was felt more while doing the movements. The intensity of pain was accentuated while performing strenuous exercises and lifting weights. There was no history of trauma. There was no past history of any systemic disease. Locally on examination, there was swelling on the infero-medial aspect of the left leg. There was no discoloration of the skin and the swelling measured 2.5 cm × 3.2 cm. There was a mild tenderness on palpation and marginally increase in size while in movements. Systemic examination was unremarkable. Plain X-ray done of the left leg did not reveal anything in both the lateral and antero-posterior projections (Figure 1).

Ultrasound (US) and color flow imaging (CFI) evaluation had revealed multicystic anechoic mass with a few low-flow vascular channels.

Patient was subjected to magnetic resonance imaging study which revealed a hypo intense region at the lower end of vastus medialis (VM) in T1W sequences (Figure 2a–c). T2W, STIR and T1W contrast enhanced MR images

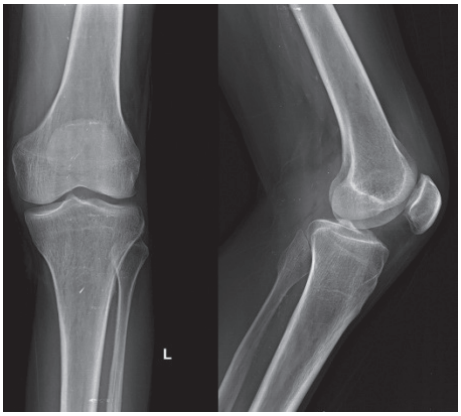

Figure 1. *Plain radiograph of left knee joint including the proximal and distal bones. There was no pathology seen in both the views.*

clinched the diagnosis as per the classical appearances (3 a and b, 4 a and b and 5 a, b and c).

The patient was diagnosed as a case of VLM of the lower part of VM. There was no need of immediate management as the patient was asymptomatic. Six-months follow up with subsequent surgical intervention had been needed.

Discussion

There are three categories of vascular malformations as slow flow, fast flow, and mixed type. Our present case

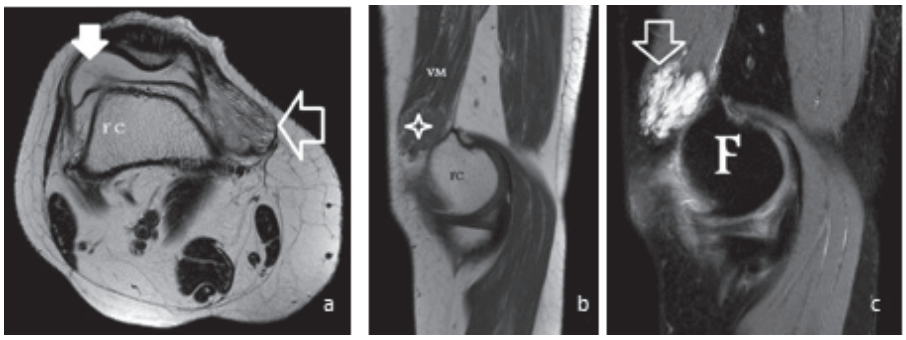

Figure 2. *Sections of the left upper leg and knee joint. (a) Axial section shows effusion in the supra patellar fossa (white arrow) and mixed intensity region (white hallow arrow). (b) T1W sagittal section shows the region having different intensity as that of muscle (star) above femoral condyle (FC). (c) STIR sagittal section shows hyperintense region at the lower end of the VM muscle (inverted arrow) just above the femoral condyle (F).*

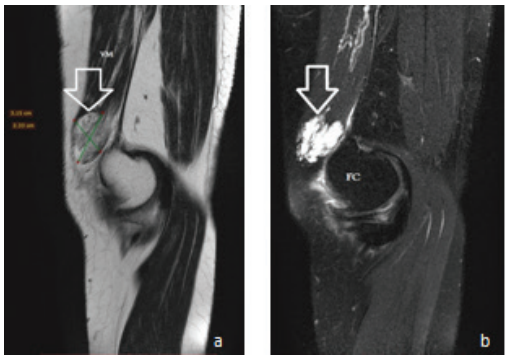

Figure 3. *Sagittal section of the affected leg. (a) T2W image shows the lesion of medium to high intensity at the lower end of the VM (inverted arrow). (b) T2W wzith fat saturation MRI image revealswell localizedpathology (white-inverted arrow) above the femoral condyle condyle.*

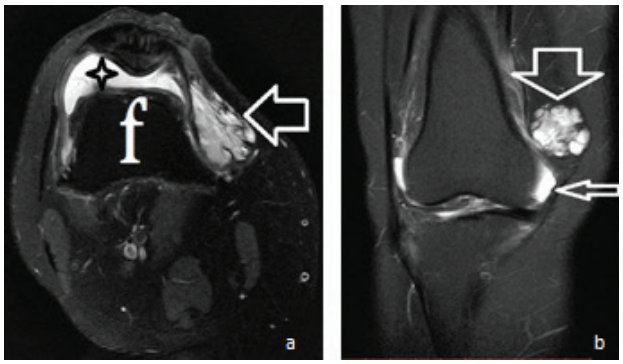

Figure 4. *STIR images. (a) Axial image revealing the site of pathology in the form of mixed intensity (white arrow). The lesion shows well demarcated lobulation adjacent to femur (f). (b) Coronal image shows the same lesion with surrounding hypointense muscles.*

falls in the last category [2]. These can either be of congenital or of acquired background. VLMs are mixture of dilated lymphatic and venous channel along with proteinaceous fluid. The lymphatic channels do not have any communication with the main lymphatic sytem. These are very low-flow channels. The lower limb is slightly uncommon site as craniofacial region is the most common area involved. There are following two processes for the development of the vascular system:

A) Vasculogenesis

B) Angiogenesis

Endothelial precursor leads to the formation of primitive vascular complexes [3]. This unites with the developing heart tube in the third week of intrauterine life. Angiogenesis is the next step which makes communication of peripheral and central circulation. Multicystic appearance is because of the dilated lymph channels. Larger veins are also present within these lesions. The presence of phleboliths in the lesion add pointer toward venous malformations. The majority of these types of lesions are found in head and neck regions. These can either be congenital or that of acquired in origin. There had been instances where spontaneous regression had been observed [4].

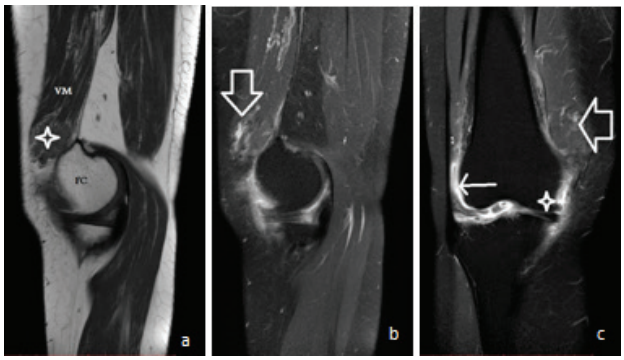

Figure 5. *Plain T1W and contrast enhanced T1W sequences of left knee. (a) Sagittal plain T1W shows subtle changes (white star) at the inferior aspect of the VM. The lesion lies adjacent to the femoral condyle (FC). (b) Contrast T1W sagittal section shows subtle marginal enhancement. (c) Coronal T1W (FS) contrast shows the lesion (wide arrow) with minimal effusion (small arrow) with the normal underlying bone (white star).*

The appearance and the palpation can either be sponge like or cystic in either of the cases. This can be of mixed consistency as was in our case. This had been shown in diagrammatic presentation with the underlying malformation. The overlying skin may be of normal looking. This could become ulcerated with superadded infection (Figure 6).

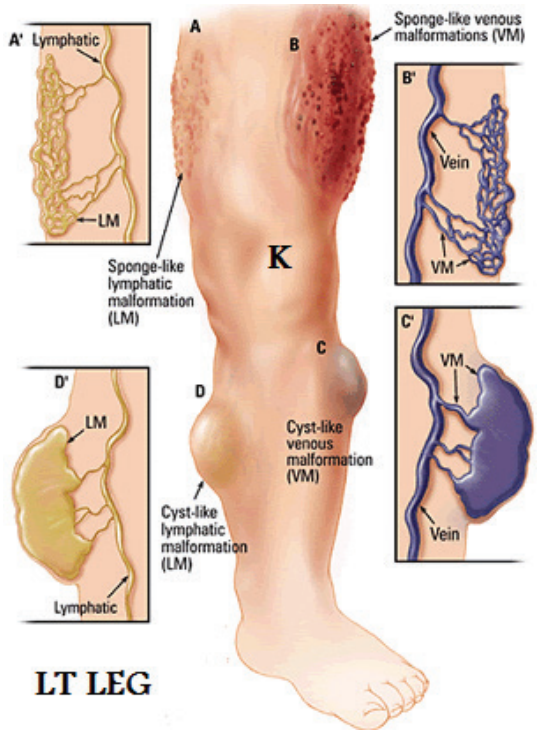

Figure 6. *Diagrammatic representation of VLMs of left lower limb (LT LEG) with knee (K) in centre. (A and B) Sponge like lymphatic and venous malformations. (C and D) Cystic type lymphatic and venous malformations (borrowed).*

Table 1. Table depicts the differentiating features of various types oflympho-vascular malformationslesions.

FEATURES	HEMANGIOMA	LYMPHATIC	VENOUS	ARTERIAL
Bruit	–	–	–	+
Overyling skin coloration	Blue–Red	No color	Blue	Blue–Red
Trans illumination	Blue–Red	No color	Blue	Blue–Red
Deflate	+	–	+	+
Refill	Rapid	Slow	Slow	Rapid

The characteristic of malformations can be differentiated on the basis of their morphological features. The features are dependent on the underlying slow flow vessels or simply lymphatic malformations. The exact aetiopathological grounds can be made after knowing the underlying involvement. This could be deep seated or superficial. These have been shown in Table 1.

The Hamburg classification is the most recent one updated for the classification of these type of malformations [5]. The main stay of the diagnosis is by Doppler US and magnetic resonance imaging (MRI). The later modality can identify the lesion with the relation and depth prospective. Computerized tomography is not helpful except where bone involvement is suspected. In rare cases, angiography may be used as adjunct to other modalities [6]. Sixteen percent of the malformations do not show any flow on Doppler US [7]. Conventional MRI has got 100% sensitivity and without any ionizing radiation. This has 24%–33% specificity in delineating the lesions. Dynamic MR study increases the specificity to the tune of 95% [8].

The management can either be with surgery, sclerotherapy, laser therapy, or combination of these [9]. The surgical management is contemplated by radiotherapy, electrocoagulation, ligation, cryotherapy, or embolization [10]. Embolization is minimally invasive surgical procedure and can be done in some case before the surgery. A microcatheter is introduced as a pre-requisite for finding out the feeder before embolisation. This will save the blood loss during the procedure [11,12].

Conclusion

The diagnosis of VLMs is benign condition and requires complete radiological evaluation for the confirmatory diagnosis. The management can only be decided, once the structural formation is confirmed. The appropriate management option can lead to full recovery. The follow-up should be done to rule out any recurrence.

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List of Abbreviations

CFI Color flow imaging
FC Femoral condyle
MRI Magnetic resonance imaging
VM Vastus medialis

Consent for publication

Informed consent was obtained from the participants.

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Summary of the case

Patient (gender, age)	1	Male, 55-years
Final diagnosis	2	VLM of vastus medialis
Symptoms	3	Pain and swelling on the medial part of left upper leg
Medications	4	Only symptomatic
Clinical procedure	5	Surgical management following embolization was advised
Specialty	6	Radio-diagnosis and Orthopedics