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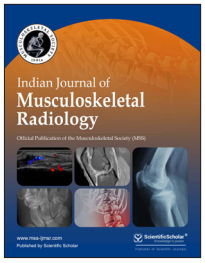

Case Report

Intramuscular hemangioma presenting as pathological femur fracture: A case report

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Received: 31 October 2022
Accepted: 06 February 2023
EPub Ahead of Print: 28 February 2023
Published: 29 June 2023

DOI
10.25259/IJMSR_39_2022

Quick Response Code:

ABSTRACT

Hemangioma is a common benign soft-tissue tumor; however, intramuscular hemangioma presenting with a pathological fracture is an infrequent occurrence. Imaging plays a key role in diagnosing this entity. We report a case of a 30-year-old male who presented with a pathological fracture of the left femur following trivial trauma. Clinical examination was remarkable for a significant soft-tissue swelling which on further radiological investigation revealed a soft-tissue mass lesion in the left thigh muscles having multiple vascular channels and phleboliths extending into femoral cortex and medulla causing a pathological fracture. Diagnosing it as an aggressive hemangioma, the main feeder artery was embolized by polyvinyl alcohol particles. The patient is on traction and follow-up. Intramuscular hemangioma causing pathological fracture is extremely rare. Understanding the typical and atypical radiological findings of an intramuscular hemangioma will impede misdiagnosis, and unnecessary tissue sampling, and allow for expedited and appropriate management.

Keywords: Vascular malformations, Intramuscular hemangioma, Pathological fracture, Embolization, Magnetic resonance imaging, Ultrasound, Phlebolith

INTRODUCTION

Hemangioma is the most common soft-tissue tumor which is histologically benign and has a vascular origin. Intramuscular hemangioma presenting with a pathological fracture is extremely rare, with only a few cases reported so far.[1] Imaging plays a key role in diagnosis and management and includes conventional radiography, ultrasonography, and magnetic resonance imaging (MRI). A histological diagnosis is rarely needed. The treatment must be individualized and includes options such as conservative management, embolization, sclerotherapy, and, infrequently, surgical excision.[2] We report a case of a 30-year-old male who presented with a pathological fracture of the femur. Radiological investigations helped diagnose the underlying cause as an atypical intramuscular hemangioma extending into the femur, causing pathological fracture, which was then managed by embolization of the feeder arteries. The fracture was managed conservatively by traction.

CASE REPORT

A 30-year-old male presented to the emergency department with profound pain in the left thigh with an inability to move his lower limb. There was a history of insidious onset mild pain in the left thigh for the past few months, followed by severe pain after a trivial trauma

2 days back. Systemic examination was unremarkable. Local examination revealed soft-tissue swelling over the left thigh, which was out of proportion to expected post-traumatic edema with severe tenderness in the lower half of the thigh. There was no neurovascular deficit. His routine blood tests were unremarkable. A radiograph of the left thigh was performed, which showed a displaced comminuted fracture through the distal diaphysis of the left femur with marked surrounding soft-tissue swelling. Few calcified foci were seen in the intramuscular plane of the thigh [Figure 1].

As there was marked soft-tissue swelling out of proportion to the expected posy-traumatic edema, an MRI of the left thigh was performed. MRI revealed a large ill-defined T2/short tau inversion recovery hyperintense and T1 isointense lesion infiltrating into and involving all of the muscles of the anterior compartment of the thigh and extending superficially into the skin and subcutaneous plane. The lesion was also extending into the gluteus maximus muscle. There was significant edema in the rest of the muscles of the thigh and intermuscular fascial planes. There was an associated femoral cortical breach with an extension of the lesion into the medullary cavity of the mid and distal femur with periosteal thickening. Multiple flow voids were seen within the lesion. The T1-weighted sequence showed interspersed fatty areas within the mass. Associated comminuted pathological fracture of femur is shown [Figure 2]. The first radiological impression from the MRI findings was some kind of vascular malformation (most likely hemangioma) in the thigh muscles extending into the femur with pathological fracture.

To further confirm the MRI findings, an ultrasound examination of the thigh was performed with the application of color Doppler.

Ultrasonography demonstrated an ill-defined heterogeneous lesion with areas of hyperechogenicity predominantly seen within the muscles of the thigh with multiple vascular channels within. On color Doppler, the vascular channels showed predominantly monophasic venous flow [Videos 1 and 2]. After using both imaging modalities, the combined impression was hemangioma/slow flow venous malformation of the thigh muscles.

Computed tomography angiography (CTA) was performed to delineate arterial supply and venous drainage of the lesion. CTA revealed an ill-defined heterogeneous hypodense lesion infiltrating into the muscles of the thigh with intense enhancement. The arterial feeders were seen arising from the profunda femoris artery. The venous drainage was seen going into the common femoral vein. The lesion was extending into the femoral cortex and medulla. On axial cuts in the bone window setting, coarsened

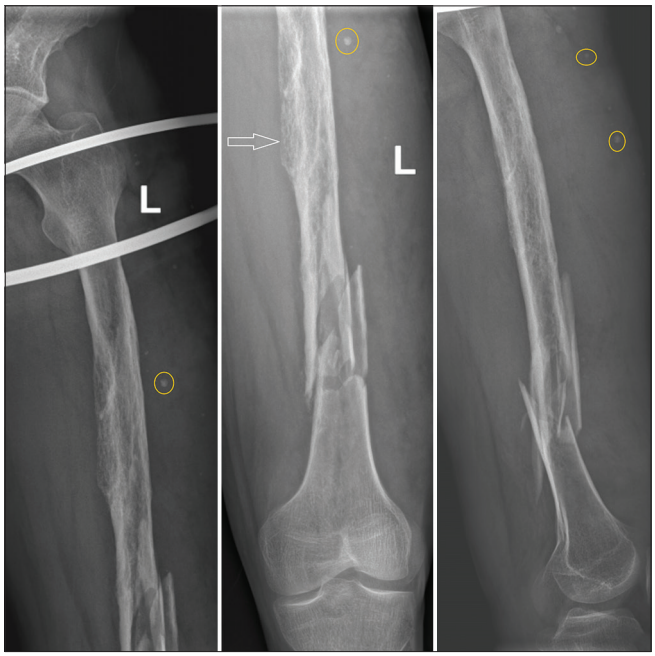


Figure 1: Radiograph of the left thigh (AP and Lateral view), showing a displaced comminuted fracture through the distal diaphysis of the left femur with marked surrounding soft-tissue swelling. Few calcified foci were seen in the intramuscular plane of the thigh (yellow circle) representing phleboliths. Upper diaphysis of the femur is showing cortical thickening, coarsened trabecula, and mild expansion (arrow).

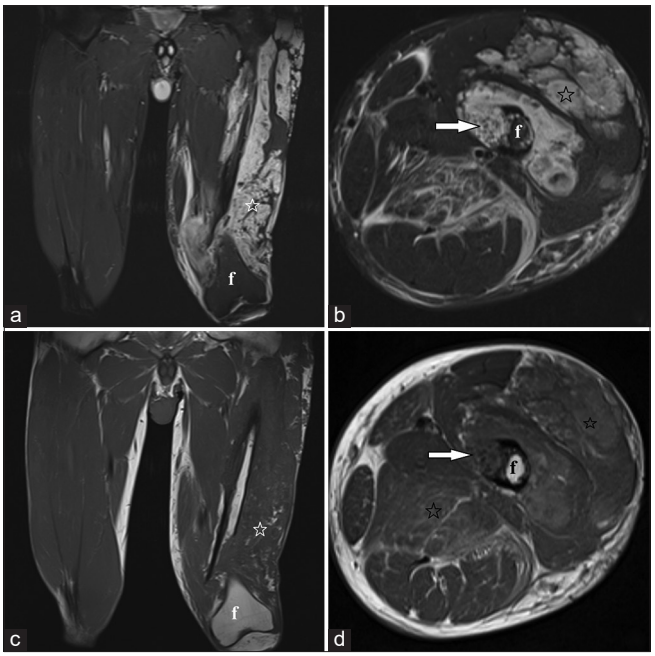
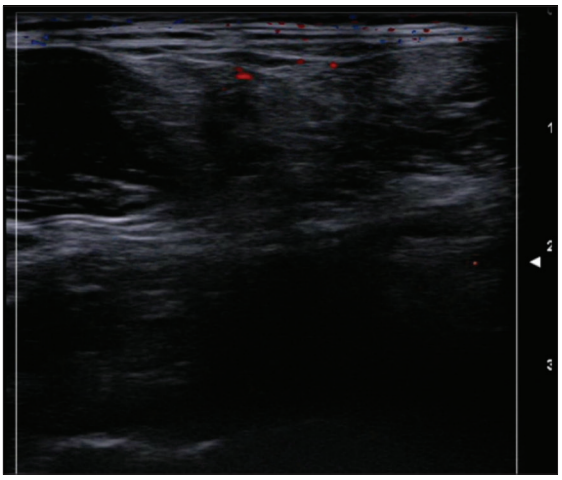
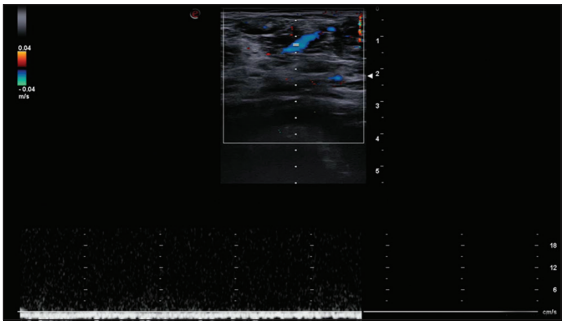


Figure 2: Magnetic resonance imaging of the left thigh showing a large ill-defined short tau inversion recovery hyperintense (a and b) and T1 isointense (c and d) mass lesion (star) infiltrating into and involving all of the muscles of the anterior compartment of the thigh and extending superficially into the skin and subcutaneous plane. There was an associated femoral cortical breach with an extension of the lesion into the medullary cavity of the mid and distal femur with cortical thickening (arrow). Associated comminuted pathological fracture of femur (f) is also seen.



Video 1: Ultrasonography video clip showing an ill-defined heterogeneous lesion with areas of hyperechogenicity predominantly seen within the muscles of the thigh with multiple vascular channels within which on Color Doppler application demonstrated predominantly monophasic venous flow.



Video 2: Spectral Doppler video clip showing monophasic venous flow in one of the vascular channels.

trabecular appearance was noted, giving a typical "polka dot appearance" (similar to vertebral hemangioma), which further confirmed our diagnosis of hemangioma [Figure 3].

The patient was planned for an interventional radiological procedure. Embolization of the feeder arteries supplying the lesion was done using polyvinyl alcohol particles under fluoroscopic guidance to occlude the vascular supply of the lesion. The fracture was managed conservatively by traction as the extension of hemangioma into the entry point of the intramedullary implant prompted the surgeon to manage him on a bohler braun frame [Figure 4]. The alignment on the frame was acceptable and maintained on regular follow-ups. The patient is advised to undergo a repeat ultrasound after 3 months.

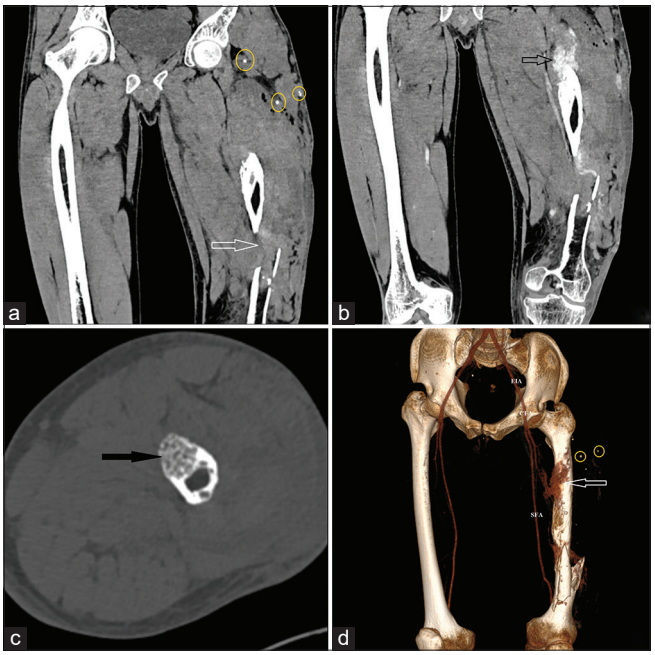


Figure 3: Computed tomography angiography (CTA) showing comminuted displaced fracture of the left femur (open white arrow in a) with an ill-defined heterogeneous hypodense lesion infiltrating into the muscles of the thigh with intense enhancement (open black arrow in b) multiple calcified phleboliths are also seen (yellow circles). On axial cuts in bone window setting (c), coarsened trabecular appearance is noted, giving typical "polka dot appearance" (similar to vertebral hemangioma) (solid black arrow in c). Volumetric data acquired from CT (d) is showing major arterial feeders arising from the profunda femoris artery. EIA: External iliac artery, CFA: Common femoral artery, SFA: Superficial femoral artery.

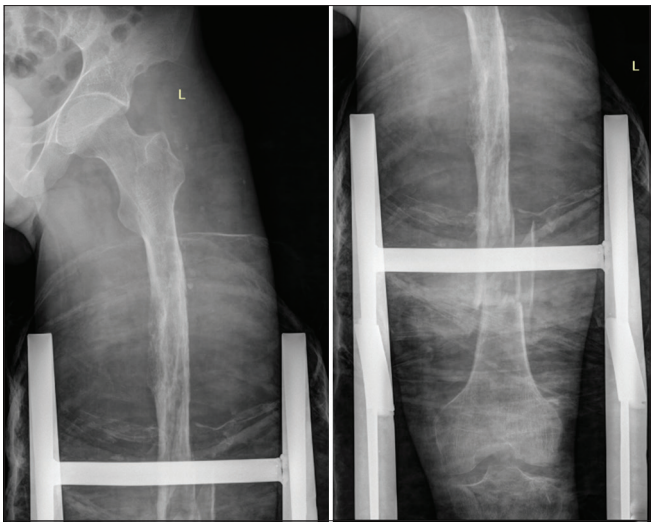


Figure 4: Anteroposterior radiograph of the left femur acquired after managing him conservatively on bohler braun frame.

DISCUSSION

Hemangiomas are benign lesions characterized by abnormal proliferation of the blood vessels.[3] They are common soft-tissue tumors, accounting for 7% of all benign soft-tissue tumors.[4] They can be cutaneous, subcutaneous,

intramuscular, or synovial.[2] Intramuscular hemangiomas are rare, and the estimated prevalence is 0.8–1.8% of all hemangiomas.[4] The most common site of intramuscular hemangioma is the thigh followed by the calf muscles.[5]

Patients with intramuscular hemangioma usually present with chronic pain, which aggravates with exercise due to the dilatation of vessels and increased regional blood flow.[2] Superficial hemangiomas are usually associated with discoloration of the skin, bruit, or pulsations. However, deeper lesions do not cause skin discoloration and are hence diagnosed late.

Histologically, hemangiomas are classified into five categories: Capillary, cavernous, arteriovenous, venous, and mixed hemangiomas.[2] Capillary hemangiomas are superficial hemangiomas originating in the skin or subcutaneous tissue. They usually present in the first decade of life and show spontaneous regression. Cavernous hemangiomas are deeper hemangiomas and are larger. They do not show spontaneous regression and surgical treatment is usually required. Arteriovenous hemangiomas have both arterial and venous components with shunt, and they may be superficial or deep. Venous hemangiomas are composed of large venous channels with thick walls. Mixed hemangiomas are composed of both capillary and cavernous components.

A radiograph will demonstrate the presence of calcified phleboliths within the lesion.[6] Ultrasonography can demonstrate the presence of soft-tissue mass with vascular channels and phleboliths. Doppler helps in delineating the vascularity of the mass. Arterial feeders and venous drainage can also be identified on an ultrasound Doppler study.

With its superior soft-tissue resolution and multiplanar capability, MRI is the diagnostic modality of choice, and most cases do not require histopathological confirmation.[3] The lesion appears iso to hyperintense on T1 and hyperintense on T2-weighted sequence with few T2 hypointense foci within due to the presence of phlebolith.[7] There is often the presence of a variable amount of fat in intramuscular hemangioma, and larger hemangiomas may be confused with lipoma.[4] Deeper lesions can cause changes in the adjacent bone which can be categorized into three categories: periosteal, cortical, and intramedullary, which may occur in the form of cortical erosion, tunneling, osteosclerosis, or coarsening of the trabeculae.[4]

Yu *et al*. reported a similar case of pathological femoral fracture in a patient with intramuscular hemangioma[1] Takeuchi *et al*. reported a pathologic femoral shaft fracture associated with a large arteriovenous malformation managed by a three-dimensional external fixator and teriparatide.[8]

The treatment includes embolization/sclerotherapy with or without surgical excision.[9] Surgical excision is indicated if there is rapid growth, intractable pain, cosmetic or functional limitation, or suspicion of malignant transformation. Embolization can decrease pain and reduce the size and vascularity of the lesion, thereby reducing intraoperative blood loss. There is a high risk of recurrence; hence, close follow-up is required.

CONCLUSION

We report a case of a 30-year-old male who presented with a pathological fracture of the femur with underlying intramuscular hemangioma, which is uncommonly reported in the literature. Understanding the typical and atypical radiological findings of an intramuscular hemangioma will impede misdiagnosis, and unnecessary tissue sampling, and allow for expedited and appropriate management.

Intramuscular hemangiomas are generally benign slow-growing tumors; however, an aggressive hemangioma can extend into the adjacent bone causing its remodeling, leading to pathological fracture. Early identification of this entity with the help of typical radiographic features (presence of phlebolith, coarsened trabecula, and thickening of cortex), ultrasound findings (presence of hyperechogenicity and multiple vascular channles), and MRI findings can help in early diagnosis, thereby preventing catastrophe associated with biopsy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Singhal S, Saran S, Agarwal A, Dhingra M. Intramuscular hemangioma presenting as pathological femur fracture: A case report. Indian J Musculoskelet Radiol 2023;5:47-51.