Intramuscular Arteriovenous Hemangioma of Thigh: A Case Report and Review of Literature

Sanjeev Patnaik[1], Praveen Kumar[2], Biswaranjan Nayak[1], Nachiketa Mohapatra[3]

What to Learn from this Article?

Diagnostics dilemmas of intramuscular arteriovenous hemangiomas involving skeletal muscles of thigh and its judicious management.

Abstract

Introduction: Skeletal muscle hemangiomas are uncommon soft tissue tumors; more than 90% are misdiagnosed initially. They present as chronic pain and swelling in a muscle with or without a history of trauma. Magnetic resonance imaging is the investigation of choice. Many treatment modalities for the symptomatic hemangiomas are available of which surgical excision is the most preferred.

Case Report: We present an unusual case of pain, swelling, and restriction of movements in the right knee following an episode of trauma in an 8-year-old boy diagnosed to have intramuscular arteriovenous hemangioma in the vastusmedialis and vastusintermedius for which he was treated by surgical excision and followed for 2 years and found to have no recurrence.

Conclusion: Skeletal muscle hemangiomas are completely treatable; the knowledge of their natural history, clinical findings, and imaging appearances are of great importance for proper diagnosis and treatment.

Keywords: Intramuscular, hemangioma, surgical excision.

Introduction

Hemangiomas constitute 7-10% of all soft tissue tumors [1]. Intramuscular hemangioma is a rare entity accounting for 0.8% of all hemangiomas [2]. There is a general agreement that females are more commonly affected than males [3]. Intramuscular hemangiomas may present as a cause of persistent pain, swelling and may present as a perceived injury [2]. Diagnostic ultrasound is an appropriate initial imaging modality for suspected hemangioma although magnetic resonance imaging (MRI) is the investigation of choice [4]. They are of interest to the surgeon because their location may present diagnostic and therapeutic dilemma.

This study reports intramuscular cavernous hemangioma of the vastusmedialis and vastusintermedius in an 8-year-old boy for which surgical excision was performed. The patient was followed for 1 year and found to have no signs of recurrence.

Case Report

An 8-year-old boy presented to us in our OPD with a swelling in the anterolateral aspect of right lower thigh since 1 year. It was gradually increasing in size and for the last 2 months was painful. There was no significant history of preceding trauma. His medical history was unremarkable, no associated constitutional symptoms or bleeding

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Author's Photo Gallery


Dr. Sanjeev Patnaik


Dr. Pravin Kumar


Dr. Biswaranjan Nayak


Dr. Nachiketa Mohapatra

[1]Department of Orthopedics, Apollo Hospital, Bhubaneswar, Odisha, India, [2]Department of Neurosurgery, Apollo Hospital, Bhubaneswar, Odisha, India, [3]Department of Pathology, Apollo Hospital, Bhubaneswar, Odisha, India.

Address of Correspondence
Dr. Sanjeev Patnaik,
Department of Orthopaedics, Apollo Hospital, Bhubaneswar, Odisha, India. Phone: +91-8093060267.
E-mail: sportho1973@yahoo.co.in

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tendencies. He had received symptomatic treatment for the last 2 months without any improvement.

Examination revealed an asymmetric, boggy, non-compressible, and non-pulsatile swelling in the anteromedial aspect of right lower thigh measuring about 10 cm × 7 cm. The overlying skin was normal without any discoloration, venous engorgement or inflammatory signs. There was fullness of medial para-patellar fossa without any joint effusion. There was tenderness over the quadriceps muscle more on the anteromedial aspect. There was no distal neurovascular deficit or bruit. Limb length was equal to the contra-lateral limb. Knee exhibited full range of motion.

With the differential diagnosis of lipoma, cold abscess, suprapatellar bursitis, synovitis, sarcoma, fibroma or other extra-articular soft tissue tumors, the patient was subjected to further investigations.

MRI of the thigh was done which revealed soft tissue mass of about 14 cm × 5 cm in the vastusmedialis and intermedius which was of high-signal intensity on T1, T2, and PD FAT-SAT images, indicating the mass could be an intramuscular hemangioma (Fig. 1).

The patient underwent surgical excision of the tumor under general anesthesia. An anterolateral approach to the distal thigh was used. A vascular mass involving the lower half of vastusmedialis andintermedius muscles of size 14 cm × 8 cm × 5 cm was encountered with bluish discoloration, feeder vessels from the profunda femoris artery were ligated, and total excision of the tumor was done (Fig. 2) and sent for histopathological examination. The biopsy revealed dilated interconnecting thin-walled vascular channels with a combination of arteries and veins. Many of these vascular spaces were filled with blood, some of the which were thrombosed and organized and others show features of recanalization, suggestive of arteriovenous (AV) hemangioma. Many of these vascular spaces were filled with blood, some of the which were thrombosed and organized and others show features of recanalization, suggestive of arteriovenous (AV) hemangioma (Fig. 3).

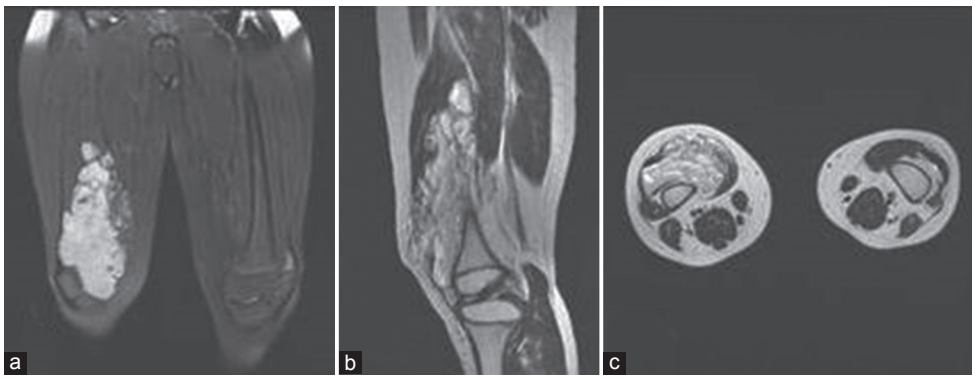

Figure 1: Pre-operative magnetic resonance images of arteriovenous hemangioma involving vastusintermedius and vastusmedialis of right thigh. (a) Coronal short tau inversion recovery image, (b) sagital T2-weighted image, (c) axial T2-weighted image.

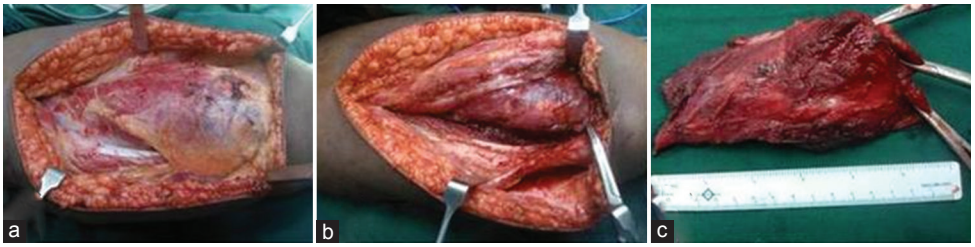

Figure 2: (a-c) Intraoperative pictures of arteriovenous hemangioma involving the vastusmedialis and vastusintermedius muscle of thigh and its excision.

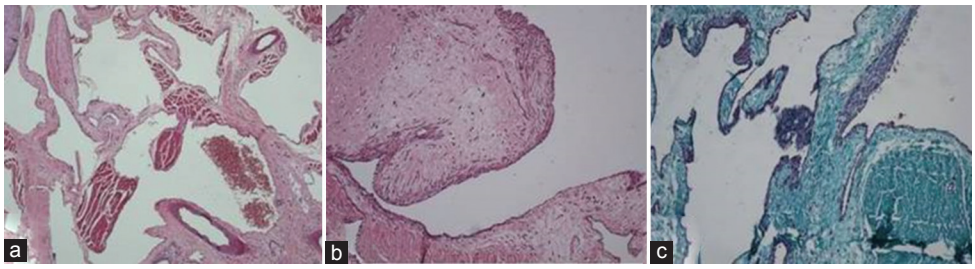

Figure 3: Microscopic picture showing both arterial complete and venous incomplete muscular lining suggestive of arteriovenous hemangioma. (a) H and E stain ×4, (b) H and E stain ×10, (c) Masson trichome stain ×4.

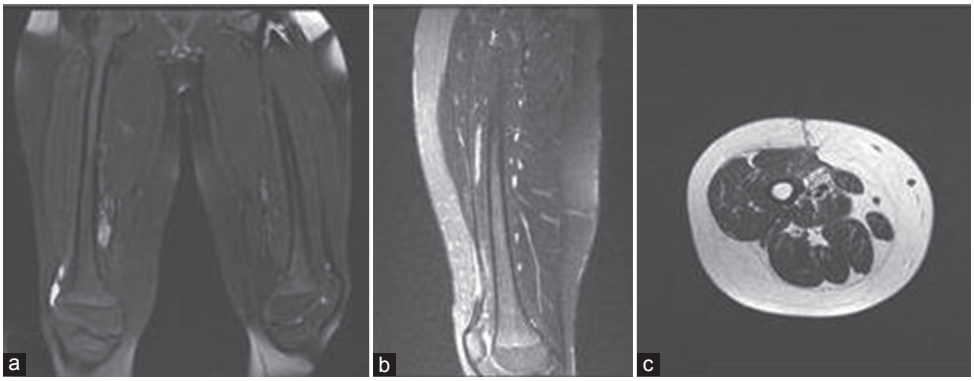

Figure 4: Post excision magnetic resonance images showing near total removal of the arteriovenous hemangioma. (a) Coronal short tau inversion recovery image, (b) sagital T2-W FAT-SAT image, (c) axial T2-weighted image.

The patient had an uneventful post-operative period. Repeat MRI was done after 1 year showed no evidence of recurrence (Fig. 4). He was followed up for 2 years and was found to be free of symptoms.

Discussion

Hemangiomas are either a neoplasm or a hamartoma. Most believe that hemangiomas are hamartomas, which are developmental anomalies, the cells being natural to the area and presenting in abnormal numbers. Hemangiomas can be divided into seven types - cavernous, capillary, venous, arteriovenous, epitheloid, granulation tissue type and miscellaneous [1] and may be localized or diffuse (more common) [5].

Hemangiomas usually grow slowly and can spontaneously regress. Their growth may be accelerated with a growth spurt or trauma. Malignant transformation is rare [1]. The younger age groups are most frequently affected with 85% of cases under the age of 30 years, among which 30% are seen in lower extremities with quadriceps being the most common muscle involved [6].

Hemangiomas can present with swelling and pain, with or without a history of trauma. They usually present as a non-compressible mass with bluish discoloration of overlying skin and engorged veins without bruit. Because of intermittent painless periods, the diagnosis may be delayed. Aspiration of the tumor yields blood, which might be confused with hematoma in the presence of history of trauma. Complications of the hemangioma include functional impairment, skin necrosis of the overlying skin, bone erosion, entrapment of vessels and nerves, cardiac failure, thrombocytopenia, and consumptive coagulopathy (Kasabach-Merritt syndrome) [6].

Plain radiographs can occasionally pick up calcified phleboliths which may be present in 25% of cases aiding in the diagnosis but may be uncommon. Ultrasonograms may reveal a complex hypoechoic mass. MRI is the investigation of choice, which helps in diagnosis and defining the location and extent of the hemangioma. Hemangiomas appear as areas of high signal intensity on both T1 and T2 images in contrast to most soft tissue tumors, which show intermediate signal intensity on T1 and high signal intensity on T2 [2, 6].

Intramuscular AV hemangiomas do not undergo spontaneous regression; hence in symptomatic cases, optimal treatment is the complete surgical excision of the tumor with a surrounding margin of the normal muscle [5, 6]. The 17-20% of local recurrence rate reported in the literature which is due to inadequate primary surgical excision rather than histological subtype [7]. Compression sclerotherapy, radiotherapy, embolization, and laser ablation [6, 8, 9] have also been suggested as treatment options in large diffuse lesions where surgical excision is impractical.

AV hemangiomas can rarely present with normal skin without any discoloration venous engorgement or inflammatory signs. Keeping this in mind any history of pain with swelling should be investigated for radiological findings before ordering a FNAC. This will also give a preliminary idea to the pathologist as to what he is dealing with - whether solid, cystic, or a vascular mass as in our case. Thus, a delay in diagnosis can be prevented and so also the complications associated with it.

Conclusion

Skeletal muscle AV hemangiomas are uncommon soft tissue tumors that are completely treatable, the knowledge of their natural history, clinical findings, and imaging appearances are of great importance for proper diagnosis. These lesions are often diagnosed late or misdiagnosed as abscess, lipoma, sarcoma, fibroma, muscle hernia, dermoid cyst, lymphatic glands, or even synovitis when presenting close to a joint. MRI is preferred for identifying the exact location, extent, and size of the lesion for planning surgical excision, which is the most preferred treatment for these lesions.

Clinical Message

AV hemangiomas although uncommon should be included in the differential diagnosis of soft tissue tumors involving the skeletal muscles. The uniqueness about this case is that Hemangiomas can rarely present with normal skin without any discoloration venous engorgement or inflammatory signs as in our case. Keeping this in mind any history of pain with swelling should be investigated for radiological findings before ordering a FNAC. This will also give a preliminary idea to the pathologist as to what he is dealing with - whether solid, cystic or, a vascular mass as in our case. Thus, a delay in diagnosis can be prevented and so also the complications associated with it.

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