Intramuscular Hemangioma Causing Periosteal Reaction and Cortical Hypertrophy: A Frequently Missed Radiological Diagnosis

B Palumbo, E Henderson, G Marulanda, D Cheong, G Letson

Citation

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Abstract

Intramuscular hemangiomas are benign tumors that consist of a heterogeneous mixture of various blood vessel types and stromal parenchyma. They are among the most common soft tissue tumors and may occur in deep structures such as muscle, tendon, connective tissue, and synovium. The most common location for these tumors is intramuscularly. The varied clinical and radiographic presentations of these tumors often lead to misdiagnosis by both radiologists and Orthopaedic surgeons. The phenomenon of hemangiomas causing osseous change in neighboring bone is not unfounded however it continues to be overlooked radiographically. Cortical, medullary, and periosteal bone changes are associated with regional hemangiomas. The purpose of this report is to present a patient with a hemangioma of the lower extremity who was originally misdiagnosed after undergoing thorough assessment with multiple radiographic modalities. We performed a literature review of the clinical and radiographic manifestations of hemangiomas.

INTRODUCTION

Hemangiomas are one of the most common soft tissue tumors and account for 7% of all resected benign soft tissue masses [1,2,3] . Histologically, hemangiomas consist of intermixed vascular and pleomorphic stromal tissues. Proposed etiopathogenesis have included congenital vascular malformations, benign vascular neoplasms, and hamartomas. An association to minor trauma as an initiating event has been discussed [4]. When hemangiomas are located within deep structures they are found within muscle, tendon, connective tissue, and synovium. Tang and coinvestigators reported the most common anatomic presentations of hemangiomas. They showed that thirty-six percent of hemangiomas are located in the thigh, seventeen percent in the lower leg, and twelve percent in the forearm [2]. The mean age at presentation was thirty years of age (Range,7-71)[2] . Common clinical findings include pain over the mass, minor neurological deficits, and decreased range of motion. They occur 1.5 times more frequently in women than in men [3,5,6].

Orthopaedic surgeons should be aware that skeletal muscle is the most common site for deeply located hemangiomas. This is important so to avoid misdiagnosis and to understand the clinical and radiographic presentations. Here is a case of a patient referred to our tertiary care oncologic institution to be evaluated for a suspected primary malignancy of the right femur. Through a careful process of clinical and radiological analysis by our team we were able to appropriately diagnose and treat the patient despite misinterpretations that had been previously made by an outside musculoskeletal radiologist.

CASE PRESENTATION

HISTORY OF PRESENT ILLNESS

A twenty-nine year old caucasian gentleman was referred to our institution with a four-year history of a painful mass on the anterior aspect of his right thigh. The referring physician, after clinical and radiological assessment, was concerned that the patient's symptoms were due to a primary malignancy of the right femur.

Pain was controlled initially by ibuprofen, however, in the weeks prior to presentation the pain became unremitting and worsened with activity. No change in size had been noted since discovery of the mass. The patient's medical history was otherwise unremarkable.

On physical exam the patient had a small, palpable mass underlying the right rectus femoris. There was no obvious skin change overlying the mass however a significant amount of pain was elicited with palpation. There were no additional pertinent physical findings.

RADIOLOGICAL ASSESSMENT

PLAIN RADIOGRAPH

A sclerotic lesion extends contiguously from the anterior aspect of the mid-diaphyseal cortex of the right femur. This mass is not well visualized on images in the anteroposterior plane although it is well visualized on the lateral projection. There is no osseous destruction or fracture associated with this lesion. There are no calcifications noted adjacent to the mass either. Soft tissue abnormalities are not identified within the femur and the visualized hip and knee joints appear normal. (See figure 1)

Figure 1

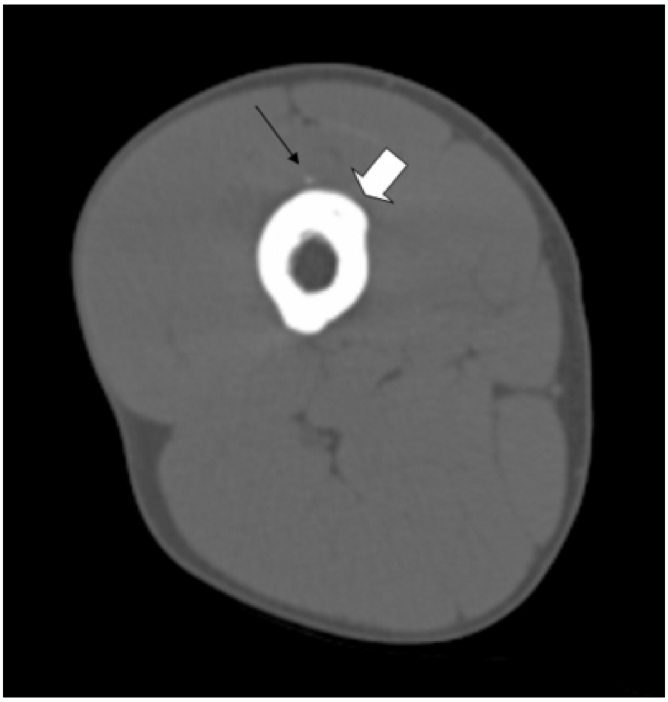
Figure 1: Plain radiograph demonstrates a region of cortical hypertrophy in the anterior- middle diaphyseal region of the right femur.

COMPUTERIZED TOMOGRAPHY WITH CONTRAST

At the right femoral there is a cortically-based lesion which is sclerotic and emanates from the anterior aspect of the femoral diaphysis (2.01 cm x 4.79 cm x 0.67 cm in the sagital, coronal, and axial planes respectively). There is no cortical breakthrough identified. A single punctate focus of high density is seen immediately anterior to this lesion within the adjacent musculature likely representing a plebolith. A lucent focus within the lesion is not seen to suggest an osteoid osteoma. (see figure 2)

Figure 2

Figure 2: Computed tomography demonstrates a cortically-based lesion which is sclerotic and emanates from the anterior aspect of the femoral diaphysis (indicated by the short, white arrow). Note the single punctate focus of high density is seen immediately anterior to the lesion within the adjacent musculature likely representing a phlebolith (indicated by the black arrow).

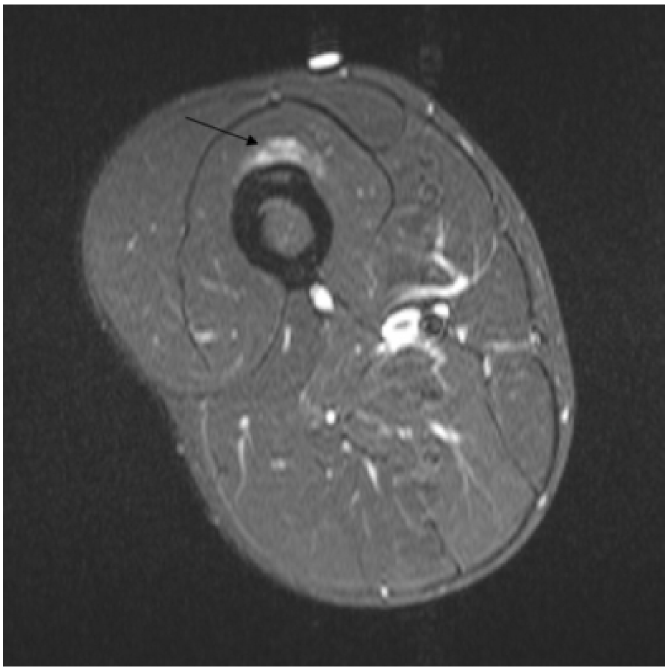


MAGNETIC RESONANCE IMAGING WITH AND WITHOUT CONTRAST

A focal area of periosteal thickening on the right proximal femur with adjacent vastus intermedius muscle enhancement was found. (See figure 3)

Figure 3

Figure 3: T2-weighted fat suppressed magnetic resonance imaging reveals a focal area of periosteal thickening and heterogeneous enhancement anterior to the mid diaphysis of the femur demonstrated by the arrow.



SURGICAL PROCEDURE

Under general anesthesia and with the use of a tourniquet the affected extremity was aseptically prepped in the usual way [7]. Palpable soft-tissue fullness in the anterior aspect of the right thigh was observed preoperatively. A longitudinal incision of eight centimeters was made over the mass. Subcutaneous tissues were divided using electrocautery splitting the quadriceps muscle atraumatically. Palpation and dissection continued down to the vastus intermedialis where the soft-tissue component was firmly located. Resection of the soft-tissue mass was performed using electrocautery down to the femoral periosteal surface. The specimen was excised and properly tagged with silk suture and sent for pathology analysis. The actual tourniquet duration was twenty-two minutes. The extensor mechanism was re-approximated and the fascia closed using Vicryl suture. The subcutaneous tissues were closed in an interrupted way with a running absorbable suture used for skin.

POST-OPERATIVE PLAN

The patient was involved in light exercises for three weeks and then allowed to increase activity level as tolerated. Physical therapy was focused on muscle strengthening, propioception, and range of motion exercises.

PATHOLOGY REPORT

The specimen consisted of an irregular, dark red-brown soft tissue mass measuring 3.0 x 2.5 x 1.5 centimeters. A diagnosis of intramuscular hemangioma with negative margins was made.

DISCUSSION

HISTOLOGICAL/PATHOLOGICAL DISCUSSION

Hemangiomas are composed of intermixed vascular and pleomorphic stromal tissues in various degrees which define there subtype. Prior to Allen and Enzinger's study of intramuscular hemangiomas and their histiologic subclassifications in 1971, it was thought that these vascular abnormalities exhibited variations of vessel morphology that distinctly subclassified them as being of venous or capillary origin [8]. Their work, however, revealed hemangiomas to be a continuum between the two vessel types rather than forming discrete diagnostic camps. In this study of 89 cases of intramuscular hemangiomas 49% were predominately small-vessel type, 29% were predominately large-vessel type, and 21% were predominately mixed-vessel type [8]

MECHANISM LEADING TO OSSEOUS CHANGE

The mechanism for boney changes due to regional hemangiomas remains unknown. It is likely that multiple factors are responsible for these changes [9,10,11]. Pathophysiologic mechanisms that have been put forth are direct pressure applied by the hemangioma, hyperemia due to increase blood flow surrounding the lesion, and stretching or irritation of the adjacent periosteum by the mass.

Studies have shown that all three categories of osseous change (periosteal, cortical, and medullary) demonstrate a significant relationship with the proximity of the hemangioma to the bone [12,13,14]. Ly and coinvestigators studied the correlation of vascular malformation size and their proximity to bone. They found that thirteen of fourteen cases of observed osseous changes the vascular lesion was in direct contact with the adjacent bone [13]. A relationship of the hemangioma size and the presence of medullary bone changes was demonstrated in this study. However, there was no relationship between hemangioma size and cortical or periosteal changes [13]. Goto and coinvestigators found a relationship between the level of the patient's pain and osseous change [14]. Sung reported that the length of bone changes and the maximum dimension of the vascular tumor was related[12]. He also reported that in a significant amount of cases the hemangioma was juxtacapsular but no osseous changes were noted on radiograph or magnetic resonance imaging. It was hypothesized that a thin plane of soft tissue can separate the vascular lesion from bone and inhibit formation of osseous changes [12].

PLAIN RADIOGRAPH

Radiographs attempting to evaluate a lesion clinically suspicious for a hemangioma often may show no abnormalities. Well-delineated, rounded calcifications called phleboliths occur commonly and raise the suspicion of a vascular tumor; especially when they are located in an area distant to venous plexuses [15]. Fulton and Sosman considered this finding pathognomonic for the presence of a venous angioma when they studied this phenomenon in 1942 [16]. Phleboliths in the presence of a deep soft tissue hemangioma is present radiographically in twenty to seventy percent of cases [12,17,18]. Other forms of calcification such as amorphous calcific deposits and curvilinear shadowing patterns can also be observed [15]. The appearance of a "calcific sponge" on radiographs has been described and correlates strongly with the presence of a cavernous hemangiomas [15].

Regional bone changes are also an indicator of a potential vascular malformation and the suspicion of a vascular tumor should increase when this finding is present along with areas of calcification and/or the presence of phleboliths. Ly and coinvestigators reviewed thirty five patients with hemangiomas and found osseous change in thirty seven percent of plain radiographs [13]. In this study three categories of osseous change were described: periosteal, cortical, and medullary. The specific morphologies of osseous changes are not discussed in this paper but are discussed in detail in the studies listed [12,13]. Thirty-seven percent of patients in Ly's study demonstrated one type of osseous change, twenty-percent exhibited two categories, and eleven-percent demonstrated all three [13]. Of the three categories of boney changes noted on plain film, seven of thirty five cases (20%) showed evidence of periosteal change, nine of thirty five (26%) showed evidence of cortical change, and seven of thirty five (20%) demonstrated medullary change [13]. Phleboliths were noted in fifty percent of these cases and they were the lone finding twenty six percent of the time [13].

COMPUTER TOMOGRAPHY (CT)

Computer tomography is able to demonstrate periosteal thickening, calcifications, and soft tissue masses associated with vascular lesions however it is not conclusively diagnostic. In Tang's review of eighty nine cases of vascular lesions, twenty four percent of patients received CT imaging yet only eighteen percent of these images were read by radiologists as being diagnostic for hemangiomas without further need for radiographic workup. Diagnostic modalities such as ultrasound, angiogram, magnetic resonance imaging, and bone scans were additionally used to form a final diagnosis in most cases [2]. CT imaging of hemangiomas with the use of contrast demonstrates only mild enhancement [18]. Computed tomography may help to further define bony changes not seen on plain radiograph however it does not offer an advantage over the combination of plain radiograph and MRI in conclusively diagnosing the presence of a vascular lesion.

MAGNETIC RESONANCE IMAGING (MRI)

Magnetic resonance imaging is highly sensitive for the detection of soft tissue masses and is useful for the detection of vascular malformations. Deep soft-tissue hemangiomas appear as heterogeneous masses with intermixed areas of low signal intensity representing fibrous septa, calcifications, and high flow vessels. Areas of high T1 signal intensity signify fat. T2 weighted imaging demonstrates a homogenous lesion which is hyperintense to fat. Memis and coinvestigators reported that in cases of small, localized hemangiomas contrast enhanced MRI produces a serpentine pattern and increases the specificity of the study [19]. Sung's review of radiographic changes in 115 cases of soft tissue hemangiomas demonstrated that twenty-nine percent of MRIs obtained detected regional bone changes [12]. MRI did not detect any cases of subtle osseous change that was missed on plain radiograph however. Tang found that among patients who had an MRI, ninety four percent revealed bone changes. Yet only twenty-five percent of these cases were read by radiologists as being diagnostic for a hemangioma [2].

ANGIOGRAPHY

Angiograms are commonly utilized to diagnose suspected vascular tumors. Such studies are able to outline vessels supplying the tumor and can demonstrate the pathognomonic feature of contrast pooling within dilated vascular spaces [18]. Greenspan found this feature in one hundred percent of the deeply located hemangiomas he studied [18]. Angiography, although effective, is an invasive modality and should be used for patients requiring more detailed surgical planning.

SONOGRAPHY

Ultrasound can be useful for identifying soft tissue masses. It does not exhibit the sensitivity and specificity that magnetic resonance imaging and plain radiograph have shown in diagnosing intramuscular hemangiomas. Vascular lesions are depicted as hyperechogenic masses on sonographic imaging. Their appearance is variable however and there is no pathognomonic representation of hemangiomas with this imaging modality [20]. In cases where magnetic resonance imaging is unavailable, ultrasound imaging may be a useful tool in aiding the diagnosis of vascular tumors.

CONCLUSION

Diagnosing deeply located soft tissue hemangiomas can prove challenging. In the case presented, a misdiagnosis was made prior to our team's intervention; we find this to be a common mistake with regard to these tumors. The purpose of this report and literature review is to demonstrate that diagnosing these vascular lesions can be accomplished utilizing careful clinical judgment and thorough radiological assessment. We have outlined important findings that may be present using many of the imaging tools that are currently available. In the pursuit of making the diagnosis multiple diagnostic modalities are likely to be used concomitantly. However, plain radiograph presents vital clinical clues that should not be ignored although are often missed by Radiologists as well as Orthopedists. Plain radiograph and magnetic resonance imaging with contrast is the most sensitive and specific combination of imaging modalities and appears to be a reasonable first line approach in diagnosing these tumors.

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Author Information

Brian T. Palumbo, M.D.
Department of Orthopaedics and Sports Medicine, University of South Florida

Eric R. Henderson, M.D.
Department of Orthopaedics and Sports Medicine, University of South Florida

German A. Marulanda, M.D.
Department of Orthopaedics and Sports Medicine, University of South Florida

David H. Cheong, M.D.
Interdisciplinary Oncology , H. Lee Moffitt Cancer Center and Research Institut

G. Douglas Letson, M.D.
Interdisciplinary Oncology, H. Lee Moffitt Cancer Center and Research Institute