Case Reports

Angiomatous Malformation: A Rare Cause of Recurrent Swelling of the Knee Joint

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Angiomatous malformation of the muscle around the knee joint is an extremely uncommon cause of recurrent swelling of the joint. We report a case of a 13-year-old boy who presented with recurrent episodes of knee joint swelling. Though the history and physical examination was typical, magnetic resonance imaging clinched the diagnosis.

A 13-year-old boy presented with recurrent swelling of the right knee joint since the age of five years. The right knee pain and swelling was not associated with any trauma, fever, early morning stiffness or pain, swelling in any other joint. Each episode used to last for 7-10 days and would occur once in 1 to1.5 year and resolve without any residual swelling. In the subsequent years the episodes became more frequent and he also noticed that they were precipitated by prolonged standing or strenuous physical activity. Arthrocentesis done outside during one of episodes revealed hemorrhagic fluid. He had received anti-tubercular treatment for about 6 months without any benefit.

His general and systemic examination was unremarkable except for the presence of fullness in the right supra-patellar area and medial aspect of the right knee joint. There was no deformity, limitation of movement or crepitus but emptying sign could be demonstrated. There was no leg length discrepancy. The investigations were all normal including radiograph of bilateral knee joints. Based on the history and examination, a diagnosis of vascular malformation around the knee joint was suspected and magnetic resonance angiography was done which revealed hemangiomatous malformation in the lower part of the right thigh (*Fig.1*). The patient was advised conservative management with avoidance of prolonged standing.

Discussion

Intramuscular hemangiomas constitute less than 1% of all hemangiomas. They occur most often in children and young adults, with 85% presenting before the age of 30 years(1,2). Lower extremity, especially the thigh as was seen in our patient is commonly involved. They can be asymptomatic or present with increased girth of the extremity, increased temperature in the area, discoloration of the overlying skin, and pain. Exercise often exacerbates the symptoms of pain and swelling due to increased blood flow as was seen in this child. Larger hemangiomas may be associated with a bruit or thrill. Often, it is difficult to differentiate them from soft tissue sarcomas(2) and other masses on clinical

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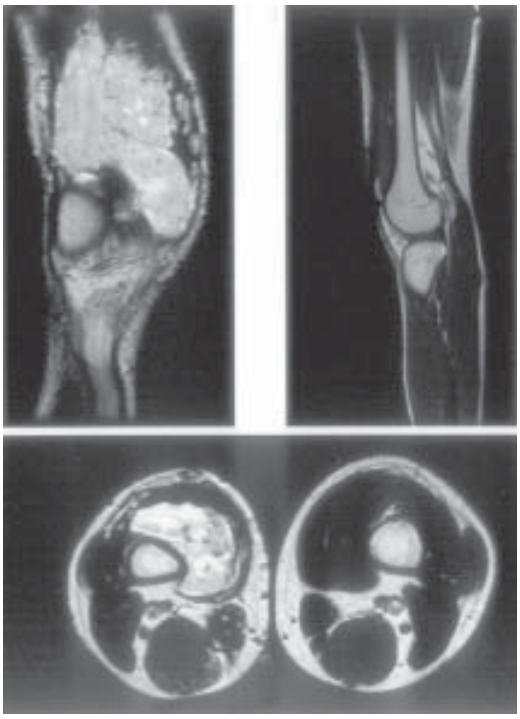

Fig. 1. T2 weighted coronal (A) and axial (B) MRI images show the lesion as hyperintense region just anterior and medial to the right lower femur. T1 weighter sagittal image (C) shows the isodense lesion anterior to the femur around the patello-femoral joint.

examination. The natural history is that of gradual fatty replacement, atrophy, and involution over time.

Bleeding into the joint is a rare phenomenon. In our case even though we failed to see any extension of hemangioma into the joint, it is possible that there is a small communication between the hemangioma and joint cavity leading to recurrent hemarthrosis. The other possibility is that the swelling is periarticular and the so-called arthrocentesis done outside was actually done through the hemangioma leading to hemorrhagic aspirate. Absence of blood in the synovial cavity on MRI favors this possibility.

Soft tissue hemangiomas may be seen on radiographs as soft tissue shadows, although typically they are isodense with muscle. An MRI(3) is the imaging modality of choice for soft tissue hemangiomas, including those of muscle(4) and synovium. Typical MRI findings are increased signals on both T1 and T2 weighted images frequently with areas of signal void, which are indicative of dense fibrous tissue or thrombi. A characteristic serpentine pattern of vascular structure is often visible(2). Gadolinium induced enhancement helps in differentiating them from other soft tissue masses.

Treatment may be needed if there is significant pain or a large mass lesion causing pressure symptoms, but due to the poor success rate of treatment and the good natural history of the process, indications for treatment are limited. Observation is appropriate for asymptomatic or mildly symptomatic hemangiomas of skeletal muscle and bone(5). If symptoms cannot be adequately managed by activity modification and non-narcotic analgesics, embolization may be used to provide symptomatic relief. The more limited the disease, the better the results with surgical excision. When surgical excision is planned, embolization may be used preoperatively to decrease intraoperative blood loss and postoperative recurrence. Complete resection is not always possible, and when not completely resected, hemangiomas may recur in more than 50% cases(1,6). Laser knife excision provides better control of intraoperative bleeding.

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REFERENCES

1. Beham A, Fletcher CD. Intramuscular angioma: a clinicopathological analysis of

74 cases. Histopathology 1991; 18: 53-59.

2. Hein KD, Mulliken JB, Kozakewich HP, Upton J, Burrows PE. Venous malformations of skeletal muscle. Plast Reconstr Surg 2002; 110: 1625-1635.
3. Buetow PC, Kransdorf MJ, Moser RP Jr, Jelinek JS, Berry BH. Radiologic appearance of intramuscular hemangioma with emphasis on MR imaging. Am J Roentgenol 1990; 154: 563-567.
4. Maki DD, Craig-Mueller J, Griffiths HJ. Radiologic case study. Intramuscular hemangioma. Orthopedics 1996; 19: 916, 907-909.
5. Pritchard DJ: Surgical management of common benign soft-tissue tumors. In: Simon MA, Springfield D, eds. Surgery for Bone and Soft-tissue Tumors. Philadelphia, Lippincott-Raven Publishers, 1998. p. 534-537.
6. Trout HH 3rd, McAllister HA Jr, Giordano JM, Rich NM. Vascular malformations. Surgery 1985; 97: 36-41.

Acute Myeloid Leukemia after Intensive Immunosuppressive Therapy in Aplastic Anemia

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A 10-year-old boy was admitted with complaints of fever, pallor, fatigue and skin bleeds of 10 days duration and diagnosed as very severe aplastic anemia. He was given intensive immunosuppressive therapy but showed no response to therapy. He later evolved into acute myeloid leukemia. The occurrence of AML is reviewed and possible pathogenesis is discussed.

Key words: *Aplastic anemia., Immunosuppressive therapy, Myeloid leukemia.*

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Acquired aplastic anemia is an uncommon but potentially life threatening hematological disease in children. The ideal treatment is HLA matched allogenic bone marrow transplant (BMT) but the high cost involved, non-availability of histocompatible sibling donor or age restrictions and nonavailability of expertise has limited its use. Intensive immunosuppressive therapy (IIST) comprising lymphoglobulin, cyclosporine A with or without steroids has achieved cure rates similar to that of BMT. IIST however is not without its side effects. The causes of concern include serum sickness, infections, increased risk of solid tumors and clonal hematological complications such as paroxysmal nocturnal hemoglobinuria (PNH), myelodysplastic syndrome (MDS) or acute myeloid leukemia (AML)(1).

We report first Indian pediatric case of aplastic anemia that evolved into AML after IIST.

Case Report

A 10-year-old boy was admitted with complaints of fever, pallor, fatigue and skin bleeds of 10 days duration. He had no other positive medical or family history of any