Orthopaedic Case of the Month

Ossified Calf Mass in a 32-year-old Woman

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History and Physical Examination

A 32-year-old woman presented with onset of progressive left leg pain and swelling after minor trauma 8 weeks previously. Approximately 6 weeks after injury, she was evaluated by a physician at an outside hospital and was prescribed crutches. Imaging studies at that time revealed a calf mass. She then was referred to our orthopaedic clinic for further evaluation. The pain was aggravated by walking, prolonged sitting, or standing. On physical examination, her left calf was tender to palpation and asymmetrically enlarged relative to the contralateral side. A calf mass was palpated. The mass extended from the proximal popliteal fossa down to the lower distal third of the tibia, clinically measured approximately 18 cm, was firm throughout its length, not freely movable from the tibia, and appeared to be deep to the subcutaneous tissue. Radiographic and MR evaluations of the patient's left calf were performed. Based on the history,

physical examination, and imaging studies, what is the differential diagnosis? A biopsy of the mass initially was performed 2 months after the onset of symptoms. The initial biopsy sections were interpreted as showing heterotopic ossification without evidence of malignant cells.

Imaging Interpretation

AP (Fig. 1A) and lateral (Fig. 1B) radiographs showed an extensively calcified mass in the left calf. The mass consisted of numerous calcifications with central lucencies that were similar in size but did not appear to involve or erode the adjacent tibia. Axial T1 (Fig. 2A), axial fat-suppressed T2 (Fig. 2B), axial fat-suppressed postcontrast T1 (Fig. 2C), and sagittal T1 (Fig. 3) MR images showed a heterogeneous mass located deep to the gastrocnemius muscle and adjacent to the posterior surface of the tibia. The mass appeared well-circumscribed. The calcifications seen on the radiograph contained fat signal (high signal on T1 images, which suppresses with fat-saturation technique), consistent with innumerable fragments of mature bone. The tissue between the ossifications showed predominantly intermediate T1 and intermediate to high T2 signal, compared with the surrounding musculature. Only minimal surrounding edema was present. Contrast enhancement was limited to the internal septa and the lining of the mass. The proximal margin of the mass was at the level of the proximal tibiofibular joint, but there was no visible extension to the knee or tibiofibular joint and no bone invasion.

After discussion of the physical findings, imaging, and initial biopsy, the decision was made not to surgically excise the mass. However, the patient continued to have persistent discomfort and difficulty ambulating and therefore we proceeded with excision of the mass 9 months after the onset of

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Fig. 1A–B (A) AP and (B) lateral radiographic views of the calf show numerous calcifications with central lucencies that were similar in size but did not appear to involve or erode the adjacent tibia.

symptoms. A 30-cm incision was made along the postero-medial calf and the fascia of the superficial posterior compartment was split to unroof the tibialis posterior muscle. Adjacent to this muscle was a 22 cm × 7 cm × 6 cm, heavily calcified mass with a thin, fibrous membrane that was adherent to the tibia proximally. Distally, the mass was peeled off the muscle and removed en bloc (Fig. 4).

Differential Diagnosis

Lipoma with chondroosseous metaplasia
 Myonecrosis with calcification
 Synovial chondromatosis
 Heterotopic ossification/myositis ossificans

Because the apparent radiographic calcifications were proven to be mature bone on the MR scan, lesions that show chondroid calcifications (soft tissue chondroma, extraskeletal chondrosarcoma) or dystrophic calcifications (synovial sarcoma, undifferentiated pleomorphic sarcoma) were excluded from the differential diagnosis. One always must consider soft tissue sarcomas, such as synovial sarcoma, in the differential for a large soft tissue mass. However, given the appearance of this mass, specifically the presence of innumerable well-defined fragments of mature bone and the absence of osseous erosion and periosteal reaction, soft tissue sarcomas were thought to be extremely unlikely and therefore were not included in the

Fig. 2A–C (A) Axial T1, (B) fat-suppressed T2, and (C) fat-suppressed postcontrast T1 MR images show a heterogeneous well-circumscribed mass located deep to the gastrocnemius muscle and adjacent to the posterior surface of the tibia. The mass contains numerous foci showing high signal on T1 images, which suppresses with the fat-saturation technique, consistent with innumerable fragments of mature bone. The tissue between the ossifications shows predominantly intermediate T1 and intermediate to high T2 signal, compared with the surrounding musculature. Only minimal surrounding edema and contrast enhancement limited to the internal septa and the lining of the mass are present.

differential diagnosis for this calf mass. Based on the history, physical examination, laboratory studies, imaging studies, and histologic picture, what is the diagnosis and how should the patient be treated?

Fig. 3 A sagittal T1 MR image through the calf show the numerous ossifications in the posterior calf. There is intermediate T1 signal between these ossifications, compared with the surrounding musculature.

Fig. 4 An intraoperative photograph shows the well-encapsulated mass during excision.

Histology Interpretation

The resected specimen consisted of a 460 g tan, firm mass (Fig. 5) covered by a thin, fibrous capsule consisting of uniform grey-white and glistening osteocartilaginous nodules embedded in fibrous stroma. A small amount of mature adipose tissue was seen on one end (Fig. 5). Histologic examination showed numerous, ovoid nodules of

Fig. 5 A photograph of a cross-section gross specimen of the mass shows numerous osteocartilaginous nodules embedded in fibrous stroma. A small amount of mature adipose tissue is seen on one end (arrow).

mature hyaline cartilage with central ossification, 0.2 cm to 1 cm in greatest dimension, separated by fibromyxoid stroma (Fig. 6). The latter was hypocellular except for the areas where spindle and stellate fibroblasts gradually progressed to form cartilage. The cartilage was mildly hypercellular with haphazard arrangement and clustering of chondrocytes and without significant nuclear atypia or pleomorphism. Rare binucleated cells were seen (Fig. 6C). Mitoses, myxoid change, sheet-like growth, and necrosis were not identified. Nearly every nodule showed a transition to lamellar bone via enchondral ossification, with fatty marrow between mature osseous trabeculae. The intervening tissue showed myxoid change. Rare islands of mature adipocytes were present (Fig. 7).

Diagnosis

Synovial chondromatosis of the calf.

Discussion and Treatment

This unusual case of synovial chondromatosis features an enlarging calf mass associated with progressive pain and difficulty walking, with onset after minor trauma. Although the imaging features of the calf mass are not specific, they are characteristic of the diagnosis: a collection of innumerable fragments of bone and cartilage with no substantial enhancing tissue. The imaging and pathologic features of this mass (particularly the presence of numerous ossified ovoid cartilage nodules) led to the correct diagnosis of extraarticular synovial chondromatosis, specifically tenosynovial chondromatosis, which is thought to have arisen from the tibialis posterior tendon sheath.

Fig. 6A–C (A) A low power microscopic view shows numerous round to ovoid nodules of mature hyaline cartilage with central ossification (Stain, hematoxylin & eosin, original magnification, $\times 20$). (B) Mildly hypercellular cartilage with a haphazard arrangement and clustering of chondrocytes can be seen. Enchondral ossification is evident (Stain, hematoxylin & eosin, original magnification, $\times 200$). (C) The chondrocytes show minimal cytologic atypia. Rare binucleated cells are seen (arrows) (Stain, hematoxylin & eosin, original magnification, $\times 400$).

Although the patient had a known history of trauma, which can serve as an inciting factor for osseous metaplasia of a lipoma [12], the minimal amount of fat component in the mass would be unusual for this diagnosis. Calcific myonecrosis is characterized by the replacement of muscle with central liquefactive necrosis and typically results in an intramuscular mass with central cystic features [11], which

Fig. 7 There is intervening fibromyxoid stroma with myxoid degeneration and rare islands of mature adipocytes (Stain, hematoxylin & eosin, original magnification, $\times 100$).

were not seen in this case. In myositis ossificans, a specific form of heterotopic ossification, the imaging appearance reflects the stage of maturation of the lesion. A soft tissue mass with circumscribed or peripheral calcification and a relative lucent center typically is seen on radiographs in the subacute phase, whereas the chronic phase often reveals a more densely ossified mass with a cortex surrounding central trabecular bone [1, 10], unlike the small uniform ossifications observed in this case. Synovial chondromatosis is an uncommon benign condition, which involves the synovial lining of joints, tendon sheaths, or bursae [8, 9]. It is divided into two main types: primary and secondary forms. Secondary synovial chondromatosis results from mechanical injury to the intraarticular hyaline cartilage and often is related to preexisting joint disease, such as prior trauma, infection, osteoarthritis, osteochondritis dissecans, or inflammatory arthritis [9]. Primary synovial chondromatosis most commonly affects adults in their third to fifth decades of life. The etiology is unknown, but it is thought to occur owing to synovial irritation and hyperplastic metaplasia of the synovial connective tissue [9, 13]. The natural history of the disease involves growth of hyaline cartilage in the subsynovial connective tissue and eventually results in the formation of cartilaginous loose bodies, which subsequently may ossify [8].

Primary synovial chondromatosis can be intraarticular or extraarticular. The extraarticular form involves the synovial lining around tendons or bursae and frequently is called tenosynovial or bursal chondromatosis, respectively. Tenosynovial chondromatosis is much less common than the intraarticular form of this disease and commonly involves the hands and feet [2, 3]. The presence of mineralization can suggest the diagnosis. Typical signs and symptoms of synovial chondromatosis include pain, swelling, restricted ROM of the affected joint, joint tenderness, palpable nodules or masses, joint crepitus, and

locking. Symptoms often have a slow, insidious onset and gradually progress with time [9]. Clinical symptoms often are present for a long time before the diagnosis is made, with an average duration of 5 years [9]. The most frequent site involved is the knee, followed by the hip [8]. Polyarticular involvement is rare [9]. Synovial chondromatosis is a slowly progressive condition, although it eventually may resolve without therapy [4]. Surgery often is indicated owing to associated symptoms and functional impairment [4].

Radiographic evaluation of patients with intraarticular synovial chondromatosis often shows multiple intraarticular calcifications, which are characteristically numerous and similar in size and shape [9]. Other radiographic features include a chondroid ring-and-arc pattern of mineralization and loose bodies with a peripheral rim of calcification [3]. In tenosynovial and bursal chondromatosis, radiographic features are similar to those seen in the intraarticular form of the disease, with the location of the calcifications reflecting the anatomic location of a known tendon sheath or bursa. The lesion shape and location are important in suggesting the diagnosis of tenosynovial chondromatosis [15]. CT is useful for detecting and characterizing the associated calcifications. MR appearance can be variable, depending on the degree of calcification and the stage of formation [6]. The most common MRI appearance of synovial chondromatosis is a lobulated T1-isointense and T2-hyperintense mass, with foci of low signal intensity on all sequences corresponding to the calcifications seen on radiographic and CT imaging [7]. Foci of peripheral low signal surrounding central fat-like signal also may be seen, corresponding to areas of ossification [7].

The main form of treatment for primary synovial chondromatosis is surgical resection. Recurrence is estimated to occur in 3% to 23% of cases, and may be related to incomplete resection [9]. There is some controversy regarding the extent of surgical resection required [14]. However, open synovectomy with removal of chondral bodies is considered to be the standard for best surgical treatment [9]. Arthroscopic synovectomy and removal of loose bodies also may be performed [5].

Malignant transformation of primary synovial chondromatosis to chondrosarcoma can occur, but is considered a rare event [9]. Patients with malignant transformation may present with pain and swelling, which are nonspecific symptoms similar to those seen with primary synovial chondromatosis [9]. Malignant transformation should be suspected in a patient with known primary synovial chondromatosis, if there is rapid increase in the size of the lesion, or rapid clinical deterioration [9]. Although there is some overlap between the radiographic appearance of recurrent primary synovial chondromatosis and malignant transformation to chondrosarcoma, evidence of cortical

destruction, bone marrow invasion, and permeation should raise concern for malignancy [9]. Treatment of malignant transformation to chondrosarcoma typically requires amputation.

After surgical resection of the calf mass, the patient's symptoms resolved. At the last followup, 14 months after excision of the mass, the patient reported no pain, swelling, weakness, or difficulty ambulating. Physical examination at that time showed a well-healed surgical incision and no palpable masses. Radiographs also were performed, which showed no evidence of a soft tissue mass or calcifications.

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