

Synovial Sarcoma of the Foot Mimicking Acute Gouty Arthritis

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ABSTRACT. Synovial sarcoma is a slow-growing soft tissue sarcoma that mainly affects young adults. Patients commonly present with a slowly enlarging mass in the paraarticular regions of extremities. We describe a case of synovial sarcoma with an unusually acute presentation near the first metatarsophalangeal joint that resembled acute gouty arthritis. (*J Rheumatol* 2005;32:2006–8)

Key Indexing Terms:
SYNOVIAL SARCOMA

ACUTE GOUTY ARTHRITIS

Synovial sarcoma is a rare soft tissue tumor that accounts for 5% to 8% of soft tissue sarcomas. It is the fourth most common type of sarcoma after malignant fibrous histiocytoma, liposarcoma, and rhabdomyosarcoma. It is the most common pediatric nonrhabdomyosarcomatous soft tissue malignancy. The histogenesis of synovial sarcoma remains unclear. Synovial sarcoma is so termed because it primarily occurs in the vicinity of joints and histologically resembles developing synovium. However, immunohistochemical and ultrastructural studies have revealed significant differences between synovial sarcoma tumor cells and the true synovial lining cells. In addition, synovial sarcoma can arise from organs where synovial tissue is absent, suggesting that synovial sarcoma may originate from cells that are widely distributed in a variety of tissues. The term synovial sarcoma is now considered a misnomer. The tumor is a true carcinosarcoma of soft tissue, despite the classic biphasic histological appearance that prompted early investigators to believe it had a synovial origin¹.

Synovial sarcoma usually occurs in the vicinity of a joint, although it has been reported in the head and neck region, lung, pleura, mediastinum, cardiac tissue, kidney, retroperitoneum, skin, abdominal wall, chest wall, and bone marrow,

etc¹⁻⁶. Ironically, despite its name, synovial sarcoma almost never occurs inside joints.

Most commonly, the tumor manifests as a slowly enlarging deep-seated mass that is painful in slightly over half of the cases. Patients may also present with symptoms related to mass effect on adjacent structures, such as dyspnea, dysphagia, hoarseness, and headaches. Occasionally, a patient may present with symptoms secondary to pulmonary metastases such as hemoptysis. Uncommon manifestations, including cerebral hemorrhage caused by metastasis of a previously unidentified synovial sarcoma that was thought to be a Baker's cyst, and a foot mass that was misdiagnosed as an arteriovenous malformation on the basis of magnetic resonance imaging (MRI) and angiogram, have been described^{7,8}.

We describe a case of synovial sarcoma with an unusual presentation that mimicked acute gouty arthritis.

CASE REPORT

A 45-year-old African American man presented to the emergency department with acute pain and swelling of right medial forefoot around the first metatarsophalangeal (MTP) joint. Two months before onset, he had had a minor trauma to this area in a car accident. There had been no pain until 2 months later, when he developed a rather acute onset of pain in the right first MTP area. It was described as a constant pain exacerbated by weight-bearing. Swelling of this area was noted, although he was not sure how long the swelling had been present. There were no fevers, chills, or morning stiffness. He denied pain or swelling in other joints. He had been taking over the counter ibuprofen for about 2 months before seeking care at the emergency department for persistent symptoms. Complete blood count and metabolic panel were normal. Uric acid was 7.1 mg/dl (normal range 4.8–8.7 mg/dl). Radiograph showed moderate soft tissue swelling over the medial forefoot (Figure 1). He was referred to the rheumatology clinic with a clinical diagnosis of acute gouty arthritis.

He denied personal or family history of gout or other types of arthritis. He had hypertension, which was controlled with felodipine. He smoked a half pack of cigarettes per day for 10 years and drank four 16 ounce bottles of beer every night. Examination revealed swelling, tenderness, and fluctuation in the area proximal to the medial aspect of the right first MTP joint. There was no warmth, erythema, or pain with range of motion in the first MTP joint.

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Figure 1. Radiograph of right foot shows soft tissue swelling over the medial forefoot. There are no bony or articular abnormalities.

Aspiration of the swollen area was attempted. Only a small drop of blood was obtained. No crystals were identified.

MRI of the right foot was performed. A large lobulated soft tissue mass was identified. It measured 8 × 5 × 3 cm, centered beneath the right first metatarsal, and straddled the first and second metatarsals. The mass was isointense to muscles on T1 images, bright on T2, and enhanced with gadolinium (Figure 2).

An incision biopsy was performed. The histology was consistent with monophasic spindle-cell synovial sarcoma (Figure 3). The tumor cells stained positively for epithelial membrane antigen (EMA) and cytokeratins 7 and 19. Neither vimentin nor *bcl-2* stain was performed. Reverse transcriptase polymerase chain reaction yielded positive result for *SYT-SSX1* fusion transcripts.

A diagnosis of synovial sarcoma was made. Search for metastases included a chest radiograph that was normal, and a chest computerized tomography (CT) scan that showed a 2.1 cm nodular density at the right lung base.

The patient underwent an amputation of the right mid- and forefoot. Postoperatively, he received adjuvant chemotherapy with adriamycin and ifosfamide. He was lost to followup after receiving one course of chemotherapy.

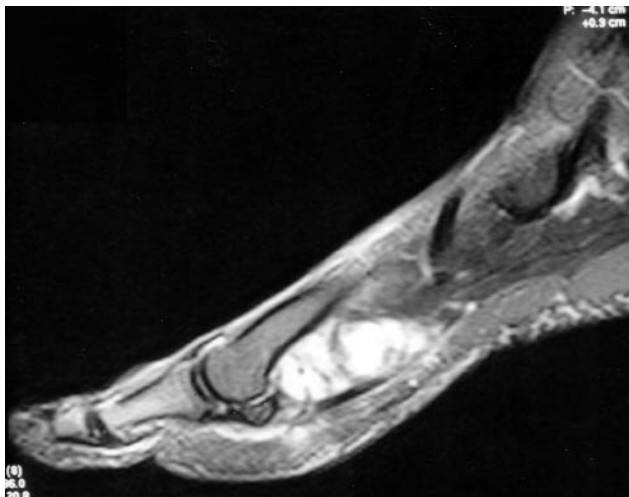


Figure 2. T2 weighted sagittal MRI scan of right foot showing a lobulated mass with moderately high signal intensity beneath the first metatarsal and proximal to the first MTP joint.

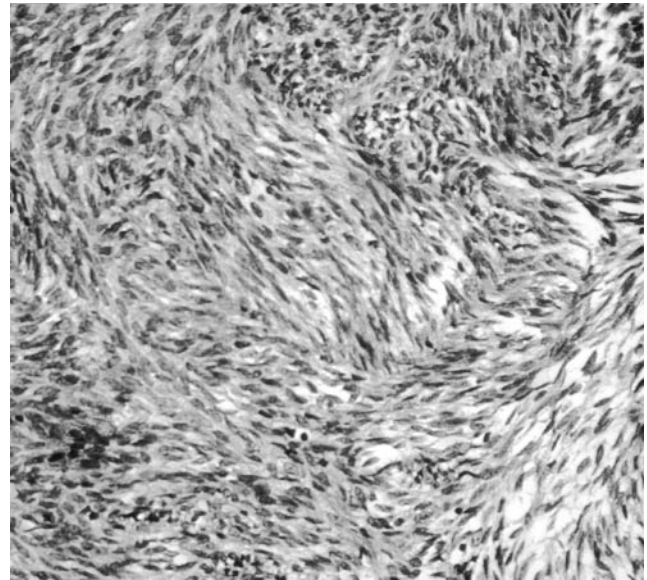


Figure 3. Histology of the tumor showing monophasic spindle cells (H&E, original magnification ×400).

DISCUSSION

Synovial sarcoma is frequently misdiagnosed as benign because of its slow growth and well defined appearance. Patients commonly present with a slowly enlarging mass that is painful in slightly over half of the cases. Our patient had an unusual presentation, with acute pain and swelling of the first MTP area, which could easily be confused with a gout attack. Indeed, he was clinically diagnosed with gout in the emergency department and was referred to the rheumatology clinic. Upon further interview and examination, we did identify several features atypical for acute gouty arthritis, including the persistent pain and swelling, which is unusual for an acute gouty attack, the location of the swelling, adjacent to but not centered around the MTP joint, the absence of erythema and warmth, and the absence of pain when moving the first MTP joint, indicating an extra-articular process. Bleeding into the tumor and trauma are common causes of acute presentations of slow-growing tumors. In our case, no massive bleeding was identified on MRI. He did have a minor trauma to the area 2 months prior to the onset of pain. It is not clear whether this injury had triggered the acute presentation.

Synovial sarcoma is usually well circumscribed, but can be infiltrative, as we saw in our patient. The tumor mass may be present for an extended period of time before medical evaluation is sought, averaging 2.5 years. Invasion of bone is seen in about 11%–20% of cases. Typical morphology is that of 2 strikingly distinct, well differentiated cell populations. One population is that of large polygonal epithelioid cells that secrete hyaluronic acid and show an organization suggestive of microscopic joint spaces. These cells are surrounded by spindle cells that simulate subsyn-

ovial mesenchymal cells. Depending on which cell type predominates, the overall histological appearances can be described as biphasic, monophasic spindle-cell (as in our case), monophasic epithelioid, or poorly differentiated types. The tumor cells usually stain positively for cytokeratins, EMA, vimentin, and *bcl-2*. Almost all patients have a t(X;18) translocation that is rarely associated with other types of sarcomas. The translocation involves the *SYT* gene on chromosome 18 and the *SSX1*, *SSX2*, or *SSX4* gene on chromosome X^{9,10}. Treatment requires wide local resection or amputation, often followed by radiation or chemotherapy. Local recurrence is common, usually within 2 years of surgery, but can be delayed for up to 10 years. The tumor commonly metastasizes to lung and occasionally to bone. About one-fourth of cases with synovial sarcoma are associated with pulmonary metastases at the time of initial presentation. Our patient had a 2.1 cm lung nodule on chest CT, which is likely to be metastatic, although no further investigation was pursued. Overall, the survival rates of patients with synovial sarcoma vary from 23.5% to 64% at 5 years and 11.2% to 34% at 10 years.

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