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CASE REPORT

Extensively calcified synovial sarcoma

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Abstract Synovial sarcoma is an uncommon fibroblastic soft tissue neoplasm, commonly arising near, but not necessarily from, the synovium of joint capsules, bursae or tendon sheaths. The radiological diagnosis is difficult. We present a case of synovial sarcoma studied with plain film radiography, CT, and pathology that had an unusual extensive calcification, which complicated its radiological diagnosis.

Key words Synovial sarcoma · Soft tissue tumors · Calcification

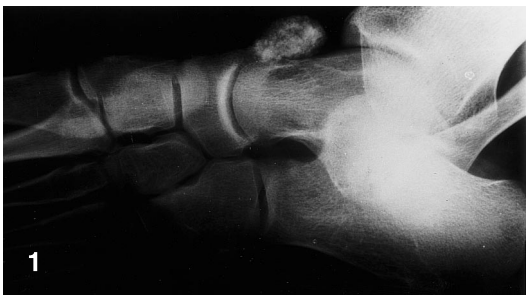
Introduction

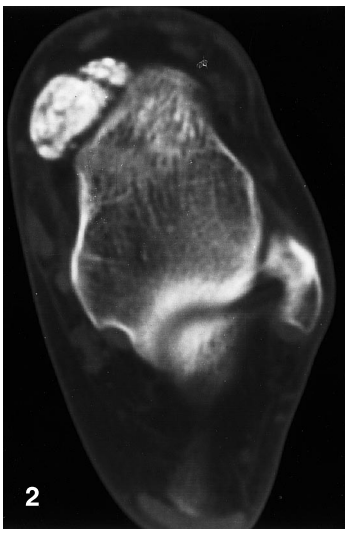
Synovial sarcomas are well known for their radiological presentation, with punctate diffuse calcifications occuring in 30% of cases. However, extensive calcification simulating cartilaginous or bony tumors is exceptional. We present a juxta-articular parosteal tumor that did not have a clear connection to the adjacent bone, did not present radiologic signs of malignancy and, on pathology, was a densely calcified synovial sarcoma. We review the features encountered on the usual radiologic examinations (plain films, CT, and MR) and emphasize features related to calcifications.

Case report

A 20-year-old man presented with a palpable painless tumor in the forefoot. The lesion had slowly grown over 7 years, not producing any other symptoms. There was no history of local trauma or systemic disease.

Oblique view plain film radiographs demonstrated a calcified soft tissue mass overlying the anteromedial astragalus. The mass was ovoid





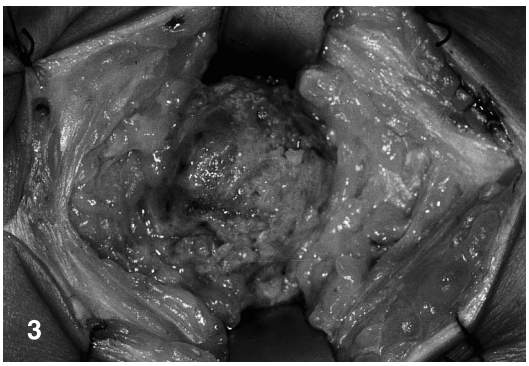
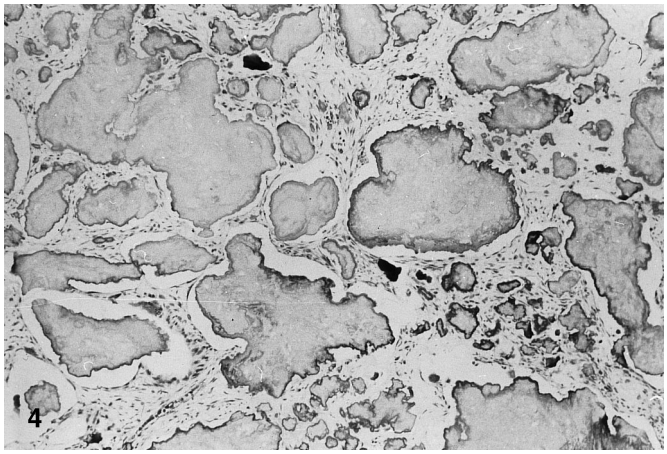


Fig. 1 Oblique roentgenograph of the forefoot depicting a well-demarcated, densely calcified soft tissue mass adjacent to the anteromedial astragalus

Fig. 2 CT scan (axial plane) of the forefoot showing the mass with punctate calcifications mixed with areas of hypodensity; no bone lesion was present

Fig. 3 Photograph of the gross specimen shows the mass not connected to bone and firmly adherent to the articular capsule



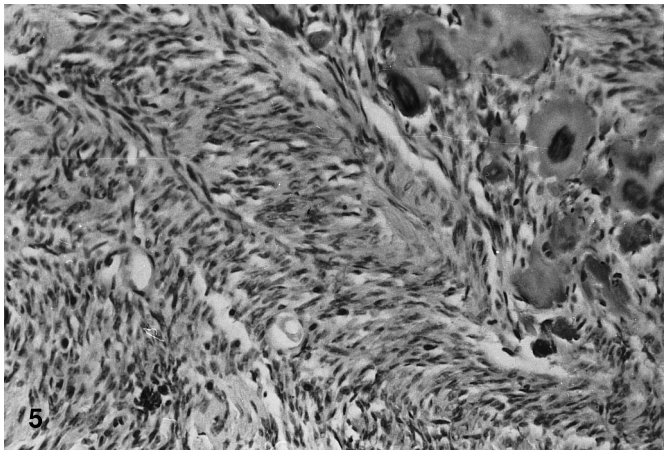


Fig. 4 Photomicrograph (×100; H&E stain) shows multiple foci of irregular and extensive calcification in a fibrous stroma

Fig. 5 Photomicrograph (×250; H&E stain) shows proliferation of spindle cells with few mitoses and infiltration with mastocytes

in shape with a maximum diameter of 2 cm. The calcification was coarse and berry-like, resembling chondroid matrix (Fig. 1).

CT scans were obtained in the axial and coronal planes. The tumor was densely calcified with well-defined margins and marked peripheral calcification. Centrally, the lesion was heterogeneous with both punctate and annular calcifications, suggesting cartilaginous calcifications, mixed with areas of low attenuation in the range of fat. A cleavage plane could be identified between the lesion and the astragalus in all images, and no bony erosion was present (Fig. 2). The lesion was located next to the anterior insertion of the ankle's articular capsule. The anterior border contacted with the insertion of the anterior tibialis tendon.

Radiologic differential diagnosis included several benign osteocartilaginous soft tissue tumors, chondroma being the most likely; extra-articular synovial chondromatosis; calcifying myositis; tumoral calcinosis; and, less probably, synovial sarcoma.

The patient underwent open excisional surgery of the forefoot lesion. The tumor was firmly adherent to the articular capsule and not connected to bone (Fig. 3). The pathologic report described an extensively calcified tumor, with dense monomorphic cellularity, few mitoses or atypia and extensive infiltration with mastocytes (Figs. 4, 5). Phenotypic studies demonstrated expression of vimentine diffusely and some areas of keratin (Ae1–Ae3). The articular capsule was infiltrated. The final pathologic report was monophasic synovial sarcoma.

Discussion

Synovial sarcoma is a malignant mesenchymal neoplasm of unclear pathogenesis representing 10% of malignant soft tissue neoplasms. It usually occurs in young adults or children (15–35 years old). Seventy percent are located in the extremities, particularly around the knee [1]. Although most are juxta-articular, only 10% occur in the articular capsule, the rest arise from tendons, tendon sheaths, and synovial bursa [1, 2].

Although synovial sarcomas are malignant and may metastasize (mainly to the lung), usually they have a slow growth and are localized, thus being easily mistaken for benign lesions [2, 3]. Clinically they present with few symptoms, usually local discomfort, pain, or mass.

Three histologic patterns have been described on pathologic examination: biphasic with epithelial and fusiform cells (33%); monophasic (31%); and poorly differentiated (36%) [2].

On plain film radiographs and CT they have been described as juxta-articular soft tissue tumors, usually lobulated [2]. They tend to involve the underlying bone in 70% of cases, producing osteoporosis, superficial erosions, periosteal reactions, or invasion [3]. The main diagnostic features are diffuse punctate radiopacities, which represent calcification. Although these calcifications are only present in 25–30% of cases, they should raise suspicions of this tumor [2, 3]. Extensive or massive calcification may occur occasionaly, as in our case. Sometimes osteoid matrix or bone is seen [2, 4], resembling osseous or cartialaginous lesions such as soft tissue chondroma, extra-articular synovial chondromatosis; ossifying myositis; tumoral calcinosis; or osteosarcoma, both parosteal and extraskeletal [3, 5, 6]. Curiously, cases with extensive calcification have been reported to have a better prognosis, with higher survival rates [2, 4].

On MRI the tumor usually appears as a septated heterogeneous mass, well marginated and next to or invading bone. On T1-weighted images there may be areas of hyperintensity representing hemorrhage [1, 7]. T2-weighted images may have liquid-liquid levels and the sign of the "triple signal": areas of hypo-,

iso-, and hyperintensity related to fat, which represent a mixture of solid, cystic, and fibrous elements and areas of hemorrhage [1, 7, 8]. Calcified areas will appear hypointense on both T1- and T2-weighted images.

In conclusion, extensively calcified synovial sarcoma is rare, but this entity should be considered since it may be mistaken for benign lesions and it may have a better prognosis than other synovial sarcomas.

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