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CASE REPORT

Heavily Mineralised Malignant Synovial Sarcoma Mimicking a Benign Extraskeletal Chondroma

HT Tam,[1] WH Ng,[1] KC Wong,[2] JF Griffith[1]

[1]Department of Diagnostic Radiology and Organ Imaging, and [2]Department of Orthopaedics and Traumatology, Prince of Wales Hospital, Shatin, Hong Kong

ABSTRACT

A 15-year-old boy presented with a 6-month history of painful wrist swelling. Radiographs, computed tomography, and magnetic resonance imaging revealed a heavily calcified extra-osseous soft tissue mass considered to be a benign chondroma. In view of the heavy calcification, the patient proceeded directly for excisional rather than percutaneous biopsy. The final diagnosis was synovial sarcoma. The pertinent imaging features of this lesion and its differential diagnosis are reviewed.

Key Words: *Bone neoplasms; Chondroma; Magnetic resonance imaging; Sarcoma, synovial; Tomography, X-ray computed*

中文摘要

擬似良性骨外軟骨瘤的嚴重鈣化惡性滑膜肉瘤

譚曉東、伍永鴻、黃國全、高士進

一名15歲男孩出現手腕疼痛及腫脹的症狀6個月。X光、電腦斷層、磁共振均顯示病人有嚴重鈣化的骨外軟組織腫塊，遂被診斷為良性軟骨瘤。由於有嚴重鈣化的情況，於是替病人進行切除式切片，而非經皮穿刺活檢。最終的診斷為滑膜肉瘤。本文會探討有關此病症的影像學特徵及鑒別診斷。

INTRODUCTION

Synovial sarcoma is an uncommon malignant soft tissue tumour typically occurring in close proximity to joints, particularly the larger joints of the lower extremity and foot. Although less than 10% of these tumours are intra-articular, nevertheless they are termed synovial sarcomas, because histologically the tumour tissue resembles synovium.[1] Radiographically, these tumours typically manifest as para-articular soft tissue masses with faint calcification, although more unusual forms can occur as illustrated in this report describing a synovial sarcoma of the wrist initially considered to be a benign chondroma.

CASE REPORT

A 15-year-old boy presented with a slowly enlarging painful wrist swelling over 6 months in September 2008. There was no history of trauma. On physical examination, an ill-defined mildly tender soft tissue mass on the dorsoradial aspect of the wrist was present.

Radiography of the wrist revealed a mass with coarse and mixed-type matrix calcification, adjacent to the carpal bones (Figure 1). Computed tomographic (CT)

Correspondence: *Dr HT Tam, Department of Diagnostic Radiology and Organ Imaging, Prince of Wales Hospital, 30-32 Ngan Shing Street, Shatin, Hong Kong.*
Tel: 852 2632 2892; Fax: 852 2648 4122; Email: tht126@yahoo.com.hk

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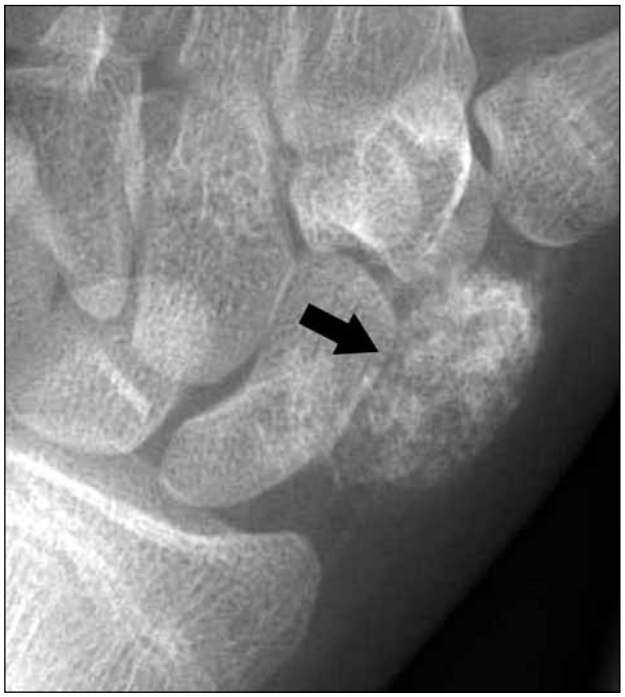

Figure 1. Radiograph showing quite a large heavily calcified mass (arrow) with mixed-type calcification adjacent to the scaphoid bone. There was no erosion or adjacent carpal bones.

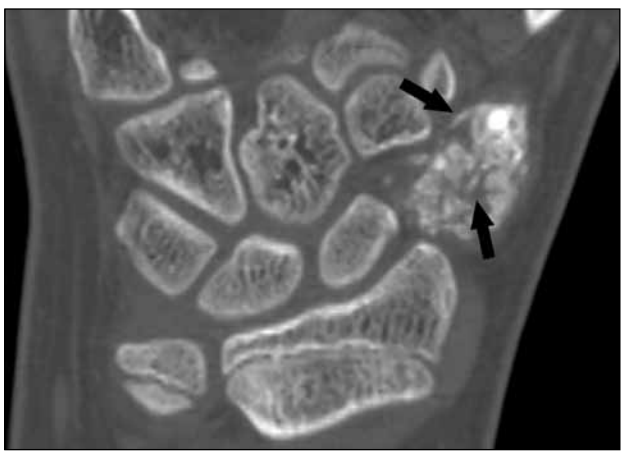

Figure 2. Computed tomographic coronal reformat image showing the heavily mineralised mass separated from the adjacent carpal bones. Some areas show curvilinear-type calcification suspicious of chondroid calcification (arrows). No bony erosion is evident.

scan revealed it to be oval-shaped with a smooth margin (Figure 2), and being heavily calcified with mixed-type calcification, though with some areas a 'ring and arcs' appearance suggestive of a chondroid matrix. A distinct plane between the mass and the adjacent bones was present with no visible continuity with the intercarpal or radiocarpal joints. No bony erosions or osteolysis were present.

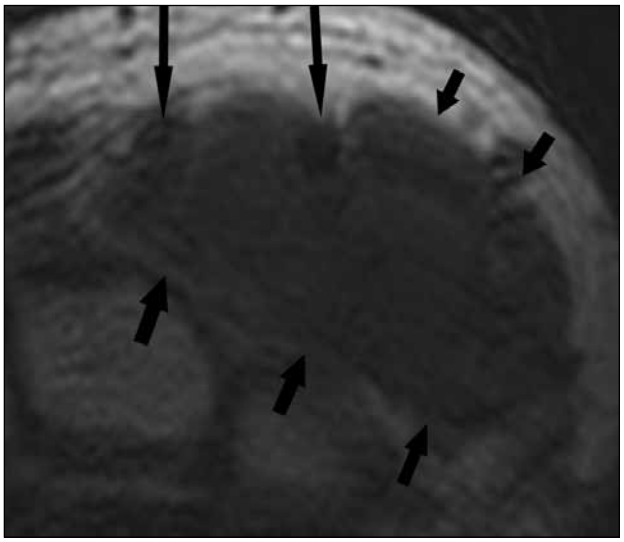

Figure 3. T1-weighted axial image showing the mass on the dorsum of the radial aspect of the wrist. It has low-to-intermediate T1 signal intensity (short arrows). The mass partially envelops the extensor carpi radialis longus and brevis tendons (long arrows).

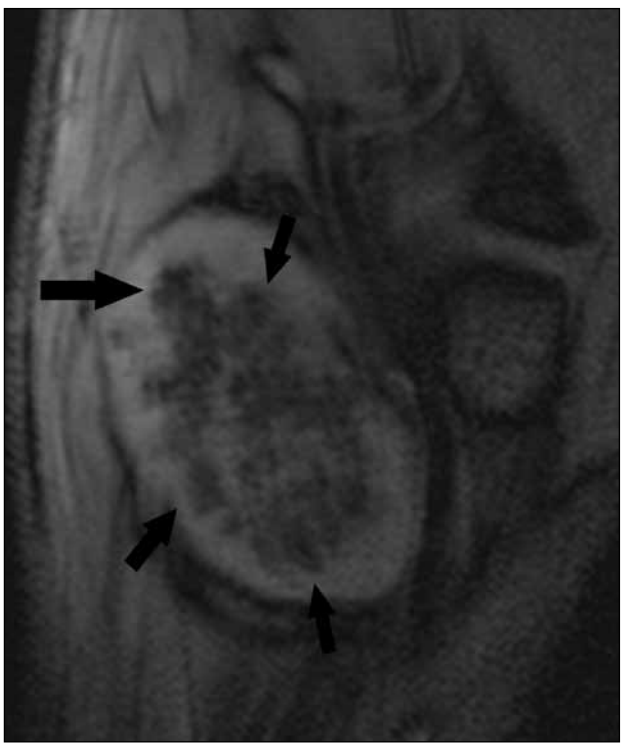

Figure 4. T2-weighted sagittal images showing the mass has multiple small hypointense nodular areas due to matrix mineralisation (arrows).

On magnetic resonance imaging (MRI), the mass was predominantly T1-hypointense (Figure 3) with several small nodular T2-hypointense foci consistent with calcification (Figure 4). The soft tissue component showed moderate contrast enhancement (Figure 5). The

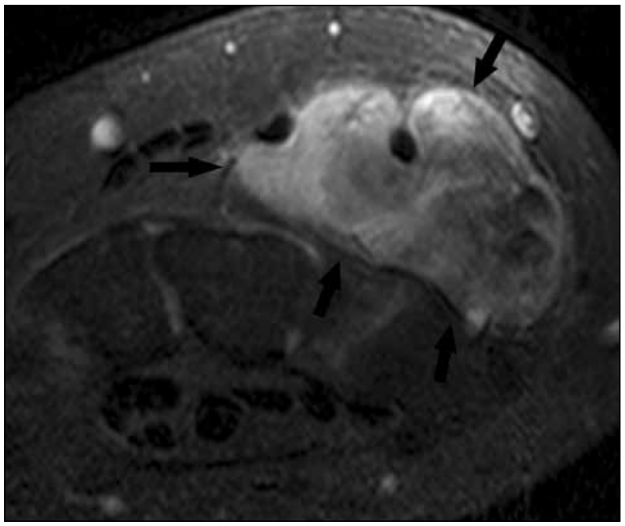

Figure 5. Post-contrast T1-weighted fat saturated axial image showing marked enhancement of the non-mineralised component of the tumour (arrows).

extensor carpi radialis longus and brevis tendons were partially enveloped although the radial artery, which was immediately adjacent to the mass, was not encased. Overall radiological features were considered most representative of an extraskeletal soft tissue chondroma. Malignant soft tissue sarcoma was considered unlikely. Percutaneous biopsy of the lesion was considered but was not performed in view of the relatively small size and extensive calcification and as all imaging findings favoured a non-aggressive chondroid tumour. In view of persistent symptoms, excisional biopsy was performed. Histology revealed a synovial sarcoma. The patient and relatives declined the offer of another operation with a wider resection margin. Postoperative radiotherapy was administered. There has been no evidence of local recurrence 12 months after surgery.

DISCUSSION

The two rather unusual features about this patient at presentation were that he was relatively young, and that the tumour was extensively mineralised.

Extraskeletal chondroma is a benign, cartilaginous, mainly encapsulated tumour, with a lobulated growth pattern. It occurs most commonly in the hands and feet.[2] Treatment entails total excision without adjunctive therapy. Synovial sarcoma is an uncommon malignant soft tissue lesion. In the hand, it comprises a very small proportion of extremity tumours, with an incidence of around one in 1.25 million inhabitants per year.[3] It is nevertheless one of the two most common soft tissue sarcomas of the hand and wrist, the other being epithelioid sarcoma. Synovial sarcomas are usually small and painless at presentation, and the duration of symptoms is reported to be 2 to 364 (mean, 98) weeks.[4] These tumours tend to be aggressive, with higher incidence of local recurrence.[5] In one series, for 50% of the patients a definitive diagnosis was only obtained after unplanned excision.[4]

Synovial sarcomas should be distinguished from benign masses, as the ideal treatment is wide local excision with or without postoperative chemotherapy / radiotherapy. Even with adjunctive treatment, estimated 5-year survival rate is about 83% in children and 62% for adults.[6] However, a significant proportion of tumours in this series underwent unplanned surgery without adequate resection of the margins. In another series, compared to patients in whom a clear margin is achieved, those in whom surgical margins were compromised had a 12-fold greater risk of local recurrence, a 3-fold greater risk of developing metastasis, and a 5-fold greater risk of death.[7]

The radiological features of synovial sarcoma are variable. On radiographs and CT, 25% of synovial sarcomas are calcified (typically at the periphery),[8] while 33 to 70% of soft tissue chondromas contain calcification (typically centrally).[2] Chondroma calcification can be curvilinear, punctuate, focally dense or mixed.[2] Rarely (as in this case), synovial sarcoma can present with heavy calcification, which resembles an osseous or cartilaginous lesion. Other calcified tumours occurring around the wrist include synovial osteochondromatosis, crystal tophi, myositis ossificans or tumoural calcinosis, though when imaged none of these mimick the appearances noted in this case. Bizarre parosteal osteochondromatous proliferation and periosteal chondroma are not included in the differential, since these tumours arise from the cortical surface.[9]

On MRI imaging, synovial sarcoma may show more specific features, including areas of T1 hyperintensity due to haemorrhage and fluid-fluid levels. Notably, the 'triple signal' sign includes T2 hyperintensity due to fluid, intermediate signal intensity equal to or slightly hyperintense relative to fat, and low signal intensity closer to that of fibrous tissue on T2-weighted images.[4,10] However, largely dependent on the degree of calcification, the MR signal is variable and as in the case it was largely T1-hypointense with multiple small T2-hypointense areas due to matrix mineralisation. In

retrospect, the best clue to this tumour being a malignant was the rapidity of onset, and the degree of tendon envelopment. This degree of tendon envelopment would be unusual for a benign chondroma, though it can occur in other benign tumours as can tendon encasement.

In conclusion, synovial sarcoma is an uncommon tumour with a variable imaging appearance, and may mimic a benign tumour. Clinico-radiological correlation is, as always, crucial though particularly so in young patients with calcified soft tissue masses. Image-guided needle biopsy should ideally be performed in all musculoskeletal tumours, so as to facilitate optimal surgical planning except in situations where the clinical and imaging appearances appear entirely benign. Use of 18F-fluorodeoxyglucose positron emission tomography may have a role in such cancer risk assessment, the prediction of patient prognosis, and treatment planning.[11]

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