

A rare appendicular skeleton chondrosarcoma

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Introduction

Chondrosarcoma is a malignant bone tumour containing tumour cells that produce cartilage. This tumour may form *de novo* as a primary lesion or from malignant transformation of a pre-existing benign condition or cartilage lesion, for example enchondroma or osteochondroma. There are numerous types of primary chondrosarcomas, including conventional intramedullary, clear cell, juxtacortical, myxoid, mesenchymal, extraskeletal, and dedifferentiated chondrosarcomas. The primary conventional chondrosarcoma, also known as central or medullary chondrosarcoma, is more commonly seen in adults 30 years and older, with the most frequently affected sites being the pelvis and long bones, especially the femur and humerus in up to 65% of cases.^{1,2} Secondary chondrosarcomas usually develop at a younger age, 20 - 40 years, and are usually of low grade malignancy.¹

Enchondromas are benign cartilaginous neoplasms that are usually solitary lesions in intramedullary bone. These benign lesions are most frequently found in the small bones of the hands and feet. These lesions replace normal bone with hyaline cartilage, and arise from cartilaginous rests displaced from the growth plate. Malignant transformation is very rare in solitary enchondromas of the hands and feet.³

We present a rare case of appendicular skeleton chondrosarcoma which left us asking the age-old question: 'What came first?' That is, was this a case of a rare primary appendicular chondrosarcoma, or the rarer transformation of an appendicular enchondroma into a secondary chondrosarcoma? The usual 'chicken or the egg' conundrum.

History and clinical findings

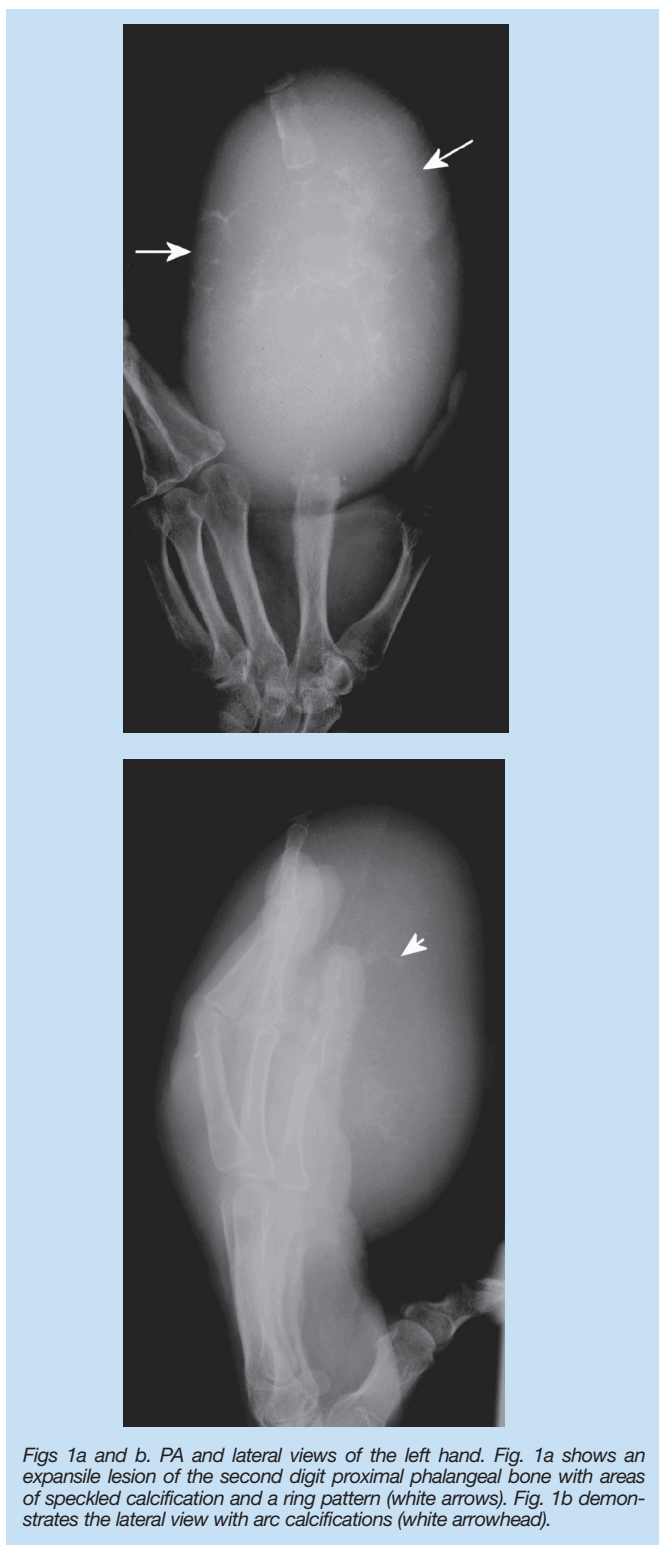
A 38-year-old woman presented with a 2-year history of a large progressive swelling of the left index finger, with occasional pain. The patient had recently developed bleeding from ulceration of the mass.

Radiological investigations

On plain radiography, the left hand index finger showed expansile destruction of the proximal phalangeal bone with areas of speckled calcification showing a ring-and-arc pattern, a known classical pattern of enchondroma calcification.² In addition, a lytic permeative component as seen in chondrosarcomas, especially higher-grade lesions, was noted. Extension of the lesion into the soft-tissue mass involving most of the left index finger was demonstrated. There was erosion of the middle and distal phalangeal bones at their bases and erosion of the distal end of the 2nd metacarpal bone (Fig. 1).

The features were suggestive of a secondary bone malignancy of cartilage origin. Thus, to exclude further cartilaginous lesions, radiographs of the right hand and both feet were also done. No enchondromas were demonstrated in the bones of either feet or bones of the right hand.

A CT scan with contrast of the left hand was performed which



Figs 1a and b. PA and lateral views of the left hand. Fig. 1a shows an expansile lesion of the second digit proximal phalangeal bone with areas of speckled calcification and a ring pattern (white arrows). Fig. 1b demonstrates the lateral view with arc calcifications (white arrowhead).

demonstrated a low attenuation mass, reflecting the high water content of hyaline cartilage, containing central speckled calcifications with a ring-and-arc pattern in the central region of the mass. Post intravenous contrast-enhanced CT showed inhomogeneous mild peripheral rim

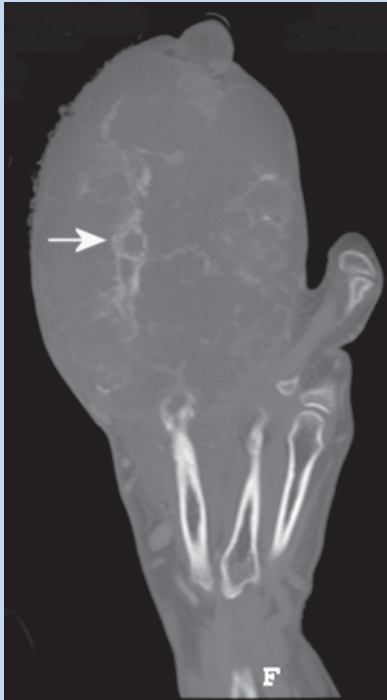


Fig. 2a. Coronal reconstructed image in bone window setting showing the left hand index finger mass with central speckled calcifications with ring-and-arc pattern in the central region of the mass (white arrow).

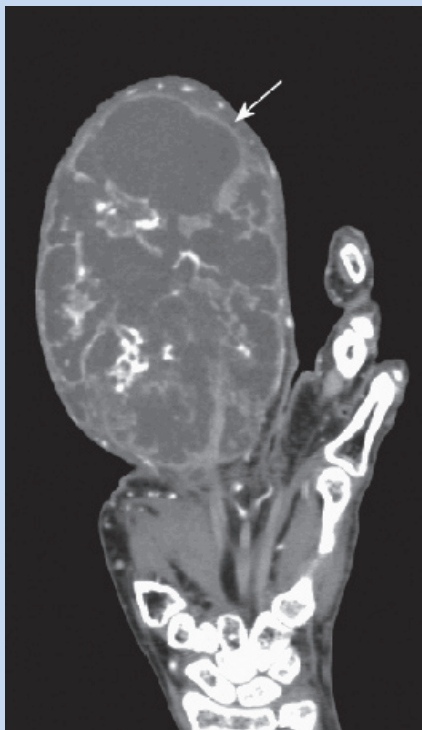


Fig. 2b. Post intravenous contrast-enhanced CT in the coronal view with inhomogeneous mild peripheral rim and septal enhancement of the mass (white arrow).

and septal enhancement of the mass. Lytic bone erosion of the proximal phalanx and middle phalanx of the index finger with erosion of the distal lateral end of the 3rd metacarpal bone was also shown. The mass

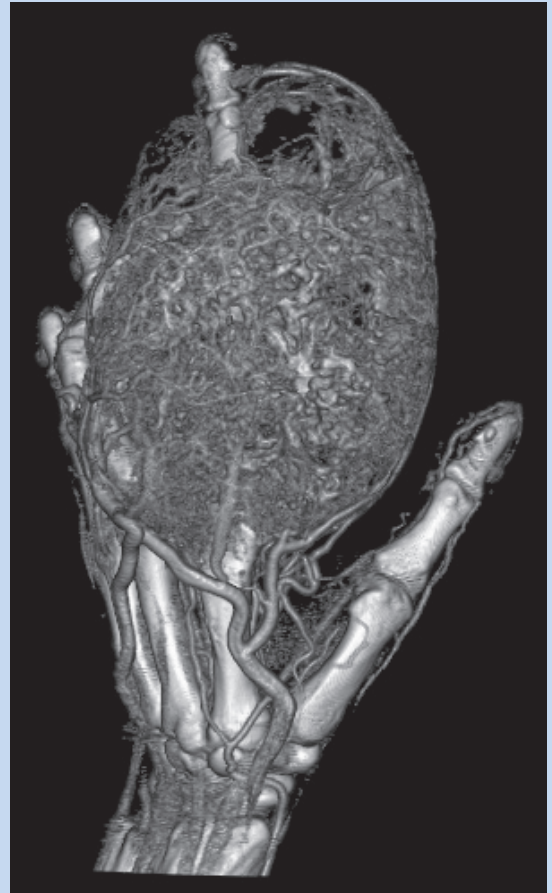


Fig. 3. CT volume-rendered representation of the elaborate vascular supply to the lesion.

encased the ligaments of the left index finger and involved the 2nd digit interosseos and lumbrical muscles. A prominent vascular supply of the lesion with neovascularisation pattern was seen (Figs 2a and b and Fig. 3).

MRI of the left hand in T1-weighted axial and coronal sequences, pre and post gadolinium enhancement confirmed involvement of the left index finger proximal and middle phalangeal bones as well as distal ends of the 2nd and 3rd metacarpal bones. Non-contrast T1-weighted axial and coronal images showed low to intermediate signal intensity of the mass indicating marrow replacement. Matrix mineralisation was seen as low signal intensity. Post-gadolinium enhancement T1-weighted images showed intense peripheral and septal enhancement of the lesion, with involvement of the ligaments of the index finger and 2nd digit muscles distally (Fig. 4).

The patient underwent a ray amputation of both the index and middle fingers of the left hand. The histology result showed a chondrosarcoma grade 3 which had been adequately excised.

Discussion

Chondrosarcomas and enchondromas

Primary chondrosarcoma is the third most common primary malignant tumour of bone, representing 20 - 27% of primary malignant osseous neo-

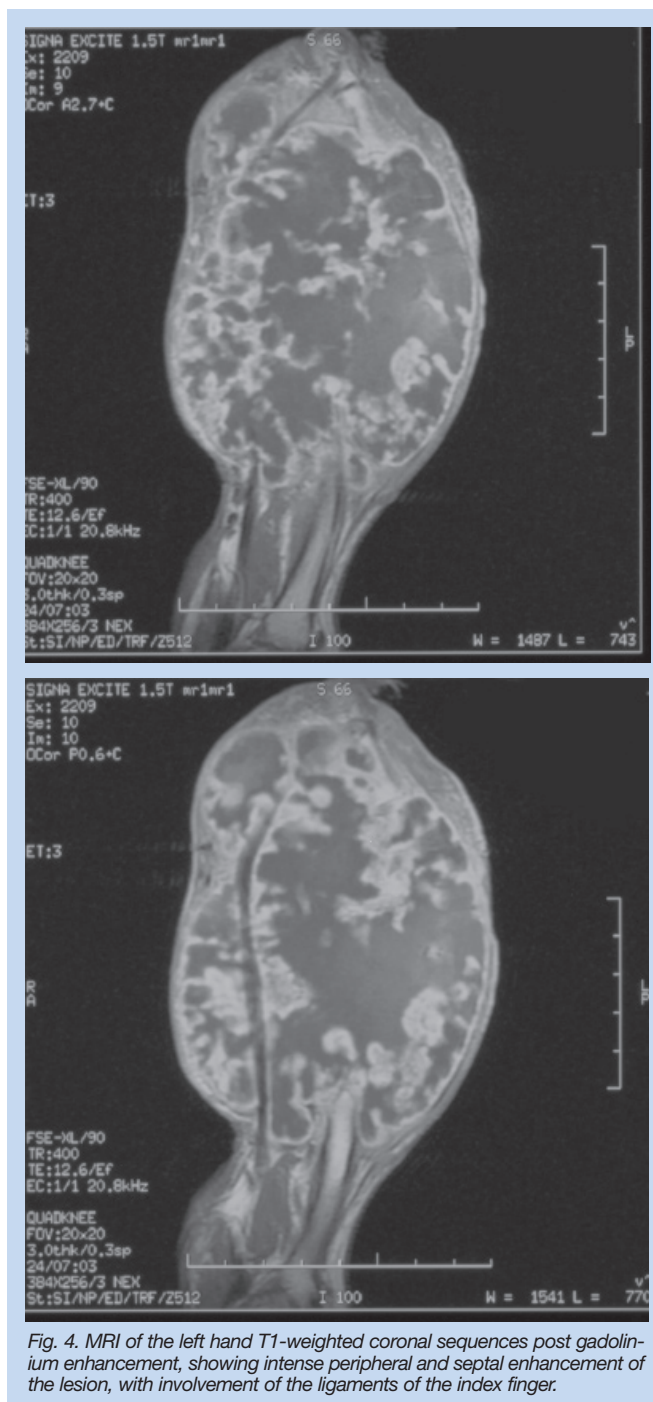


Fig. 4. MRI of the left hand T1-weighted coronal sequences post gadolinium enhancement, showing intense peripheral and septal enhancement of the lesion, with involvement of the ligaments of the index finger.

plasm.² Most frequently affected sites in primary chondrosarcoma include the pelvis and long bones in up to 65% of cases, with the short tubular bones of the hand and feet being rarer sites (1 - 4% of all cases), with less than 200 cases having been described.^{2,4-6}

Chondrosarcomas are categorised as central, peripheral, or juxtacortical (periosteal) lesions depending on their osseous location. Central chondrosarcomas are intramedullary in origin, and large tumours may erode the cortex and invade the surrounding soft tissue.² Bovée *et al.*⁴ reported 35 cases of chondrosarcoma of the phalanx, with the range of patient age at the time of diagnosis being 21 - 87 years. There was a slight female predominance. Occurrence in the hand was noted more commonly than in the foot,

with the proximal phalanx affected most often.

Histological grading of conventional intramedullary chondrosarcomas correlates with clinical behaviour and prognosis. A three-grade system is commonly used.

Grade 1 lesions - low grade, with a predominantly chondroid stroma, with distinction of grade 1 chondrosarcoma from enchondroma often proving difficult.²

Grade 2 chondrosarcomas - intermediate grade, have less chondroid matrix and are correspondingly more cellular. Necrosis may be seen.²

Grade 3 chondrosarcomas - high grade, show greater cellularity and nuclear pleomorphism than grade 2 tumours. Chondroid matrix is sparse or absent. Foci of necrosis are seen and are frequently extensive.²

Enchondromas are usually solitary lesions in intramedullary bone and are commonly seen in the hands and feet, while hands and feet are rare sites for intramedullary chondrosarcomas. The primary significance of enchondromas is related to their complications, being pathological fractures, and a small incidence of malignant transformation.³ Many texts have noted the rarity of an enchondroma progressing to become a malignant chondrosarcoma.

Enchondromatosis is a condition where multiple enchondromas are present. Three distinct disorders associated with multiple enchondromas are known: Ollier disease, Maffucci syndrome and metachondromatosis.

Solitary enchondromas are intramedullary lesions; they may expand enough to cause endosteal scalloping of the cortex. They have a predilection for the small bones of the hands and feet, with half involving the proximal phalanx, followed in frequency by the metacarpal, middle phalanx and distal phalanges and carpus. Approximately 50% of solitary enchondromas are found in the hands. Other locations include the shoulder, pelvis, and long bones. Enchondromas tend to occupy the diaphyseal region in the short tubular bones and the metaphyseal region in the longer bones.

Multiple clinical and imaging parameters have demonstrated statistically significant differences between enchondroma and chondrosarcoma, particularly pain related to the lesion, deep endosteal scalloping of more than two-thirds of cortical thickness, cortical destruction and soft-tissue mass seen on CT or MRI and periosteal reaction seen at radiography. All of these features strongly suggest the diagnosis of chondrosarcoma and thus allow distinction of appendicular enchondroma and chondrosarcoma in at least 90% of cases.⁷ In a study of 30 cases of chondrosarcoma lesions in the hands and feet Dahlin and Salvador⁵ noted that the lesions produced lytic areas of destruction that lacked, at least in part, well-defined margins commonly seen in enchondromas as well as causing apparent expansion of a portion or all of the bone of origin. CT and MRI demonstration of soft-tissue extension of the lesion with mass formation essentially excludes the diagnosis of enchondroma.

The typical radiological findings on plain film, CT and MRI, of both enchondromas and chondrosarcomas are represented in Table I.

Treatment and prognosis

For conventional intramedullary chondrosarcoma there are two choices of surgical treatment in the case of 'low-grade' type (grade 1). The first is intra-lesional curettage, adjunct chemical or thermal ablation, and cementation or bone grafting of the defect. The second surgical option is wide excision with structural graft or metal reconstruction. With obvious histological grade 2 or 3 features aggressive surgical management is necessary to optimise local disease control and reduce the frequency of

Table I. Common radiological features of enchondromas and chondrosarcomas on plain film, CT and MRI

	Enchondroma	Conventional intramedullary chondrosarcoma
Plain film radiography	<ol style="list-style-type: none"> 1) Classic ring-and-arc pattern of calcifications, being pathognomonic when seen in the hands. 2) Endosteal scalloping greater than two-thirds of the normal thickness of the long bone cortex is strong evidence of chondrosarcoma versus enchondroma.⁷ 	<ol style="list-style-type: none"> 1) Mixed lytic and sclerotic appearance. 2) Characteristic appearance of ring-and-arc pattern. 3) Sclerotic areas represent chondroid matrix mineralisation and are seen in 60 - 78% of lesions.² 4) Aggressive pattern of bone lysis with a moth-eaten and permeative pattern may be seen with higher-grade conventional chondrosarcomas grade 3.
Computed tomography	<ol style="list-style-type: none"> 1) Mineralisation in the form of rings and arcs, which correspond to calcification around lobules of cartilage. 2) Endosteal scalloping may be present, and may represent degeneration of the enchondroma to a chondrosarcoma. 3) Should not penetrate the cortex or extend into the soft tissues 4) Neither the extent nor the presence of matrix mineralisation identified on CT scans helps distinguish between long bone enchondroma and chondrosarcoma. 	<ol style="list-style-type: none"> 1) In chondrosarcoma of long bones calcification throughout the lesion is seen in 94% of cases. 2) The soft-tissue component frequently reveals typical punctate or ring-and-arc matrix mineralisation and a lobular growth pattern. 3) Higher-grade lesions may show higher CT attenuation, caused by increased cellularity and resultant reduced water content.
Magnetic resonance imaging	<ol style="list-style-type: none"> 1) Lobulated borders with a cluster of numerous tiny locules of high-signal-intensity foci on T2-weighted images that appear to coalesce with one another and reflect the high fluid content of hyaline cartilage.³ 2) On T1-weighted images, enchondromas demonstrate low-to-intermediate signal intensity.³ 	<ol style="list-style-type: none"> 1) On T1-weighted MR images, marrow replacement appears as low to intermediate signal intensity. 2) On T2-weighted MR images the non-mineralised components of chondrosarcoma have high signal intensity, a reflection of the high water content of hyaline cartilage. 3) The cartilaginous lobules may be surrounded by low-signal-intensity septa. 4) Areas of matrix mineralisation are common in intramedullary chondrosarcoma (79% of long bone lesions on MR images) and have low signal intensity with all MR pulse sequences. This feature often creates marked heterogeneity on T2-weighted MR images.

metastatic spread of disease. The local recurrence rate of grade 3 chondrosarcoma is 47%, while the reported rate of distant metastases is 10 - 50% for grade 2 lesions and 50 - 71% for grade 3 lesions.²

Metastases most commonly involve the lung, regional lymph nodes, and liver.

The overall 5-year survival rates for chondrosarcoma are 90 - 94% (grade 1), 61 - 81% (grade 2), and 43 - 44% (grade 3), whereas the 10-year survival rates are 83 - 87% (grade 1), 41 - 64% (grade 2), and 27 - 29% (grade 3).²

Phalangeal chondrosarcoma behaves as a locally aggressive lesion, and in contrast to chondrosarcomas located elsewhere, rarely metastasises.³

Conclusion

Primary chondrosarcoma is the third most common primary malignant tumour of bone, and most commonly affects the pelvic bones and long bones, with the bones of the hands and feet being rare sites for primary involvement.

Enchondromas are benign cartilaginous neoplasms that are usually solitary lesions in intramedullary bone, with malignant transformation

being very rare in solitary enchondromas of the hands and feet. Since no previous imaging was available on our case we are still faced with the dilemma as to whether the lesion was a primary or a secondary chondrosarcoma. However, since the literature points to the rarity of primary chondrosarcomas in the hand and feet, we suggest that this case may represent that rare transformation of a solitary enchondroma into a chondrosarcoma.

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