

REVIEW

Imaging of chondrosarcomas

L Ollivier, D Vanel and J Leclère

Department of Radiology, Institut Curie, Paris, France

Corresponding address: Dr Liliane Ollivier, Department of Radiology, Institut Curie, 26 Rue D'Ulm, Paris 7005, France. Tel.: +0033 1 44 32 40 15; fax: +0033 1 44 32 40 15. E-mail: liliane.ollivier@curie.net

Abstract

Chondrosarcoma is the commonest primary sarcoma of bone in adults, with a male predominance. Patients are usually between 30 and 70 years old. Clinical symptoms are pain and tenderness with or without a mass, the average duration of symptoms being 1–2 years, but growth may be very slow, especially for pelvic tumours^[1]. Chondrosarcomas characteristically produce coalescent cartilage lobules of various size. The center often becomes necrotic or cystic^[1].

Skeletal distribution

The commonest sites are the pelvic bones, the femur and humerus. Other sites are the trunk, the skull and facial bones. Involvement of the hands and feet is rare. Peculiar forms develop on laryngeal cartilage, base of the skull, or in soft tissue. Chondrosarcomas may occur on pre-existing lesions.

Central chondrosarcoma predominates in long bones and peripheral tumours in the pelvis and vertebrae.

Imaging

Plain films allow the depiction and location of the lesion, identify its cartilagenous nature and its aggressiveness. Central chondrosarcoma is the most frequent type of lesion. The tumour begins in the metaphysis and extends to the diaphysis. The lytic lesion usually appears well-defined, associated with endosteal scalloping, and cortical thinning or thickening. High-grade tumours show irregular margins. Calcifications of the tumoral matrix may be punctate, flocculent, or have a ring-like pattern, they can be small, or disseminated, dense or subtle. Their absence is frequent in aggressive types. When the tumour extends into the soft tissue the mass is frequently huge and palpable. CT-scan has a diagnostic role as it shows the bony destruction, the small calcifications, and the intra and extra-osseous extent.

In typical forms, MRI shows a lobulated lesion with a low or intermediate signal on T1-weighted images and a high-signal intensity on T2^[2]. MRI allows for the precise staging of the medullary involvement and the soft-tissue mass, and helps in diffusion: low-grade lesions show a lobulated pattern with enhanced septations after intravenous injection of contrast media. High-grade tumours do not have septations and show a more diffuse, heterogeneous enhancement. Benign and low-grade tumors cannot be differentiated by the MRI appearance of the matrix alone.

Differential diagnosis

The main differential diagnosis is chondromas, especially in the differentiation between a benign chondroma and a low-grade central chondrosarcoma^[3]. Pain, a proximal location or a location on the axial skeleton, size being greater than 5 cm, a lobulated aspect, an ill-defined margin, endosteal erosion and bone destruction with an extra-osseous component all suggest a malignant lesion^[4,5]. Biopsy is necessary to make the diagnosis. A metaphyseal lesion could suggest a chondromyxoid fibroma, an epiphyseal lesion a chondroblastoma or a giant cell tumour. Fibrous dysplasia or a bone infarction can be misdiagnosed as chondrosarcomas, the lack of cortical erosion or of soft-tissue mass would suggest something other than a chondrosarcoma.

This paper is available online at <http://www.cancerimaging.org>. In the event of a change in the URL address, please use the DOI provided to locate the paper.

New immunohistochemistry techniques contribute to the differentiation of malignant lesions^[6,7]. More rarely, a lytic lesion can be considered with a lytic form of osteosarcoma or fibrosarcoma, a plasmocytoma or a metastasis.

Chondrosarcomas: variants

Periosteal chondrosarcomas^[8]

This is a rare form representing 1–2% of all cases of chondrosarcomas. The growth of the tumour begins at the surface of the bone (usually the metaphysis of the distal femur or proximal humerus) and develops in the soft tissues as a lobulated mass. It is revealed by long-term indolent tenderness. The lesion is usually well-differentiated and grows slower than central forms. The cortex is never normal, either eroded or often thickened by the tumour, but never destroyed. Ring-like calcifications can be disseminated or localised within the mass. Medullary involvement evaluated on CT or MRI is rare and limited. Tumoral nodules not calcified are hypodense on CT and have a high signal on T2 weighted MRI. Satellite nodules can be depicted and separated from the principal lesion. The outcome is generally favourable after an appropriate surgical resection.

The differentiation from an osteochondroma is generally easy. The diagnosis of periosteal chondroma can be made by histology alone. Patients are younger, lesions are smaller, not painful and are located more distally on the skeleton. The periosteal osteosarcoma is more often located on the diaphysis and has reactional cortical spiculations.

Mesenchymal chondrosarcoma

This entity, representing 2–3% of all chondrosarcomas, combines an undifferentiated cell component with well-differentiated cartilaginous areas^[9]. The diagnosis is only made on this biphasic aspect. The average age of the patients is 26 years. Common skeletal sites are the femur, pelvic bones, ribs and vertebrae. About one-third of cases involve the extraosseous sites: brain, meninges, or soft tissues. The prognosis is poor, with early pulmonary, bony and lymph nodes metastases. The tumours are large and destructive lesions, some have a purely lytic pattern. Calcified masses can be found. The lesion appears of low signal intensity on T1 weighted MRI and heterogeneous high signal on T2 images^[10]. The lesion is aggressive. Multidrug chemotherapy used in osteosarcomas can be combined with surgery and radiotherapy, but the 10-year survival is only 28%.

Clear cell chondrosarcoma

A rare form (2%) of chondrosarcoma, these lesions are distinguished by their cytology, epiphyseal location in

long bones, and slow evolution^[11]. There is a male predominance and most patients are in their third to fifth decade. Clinical symptoms are pain and swelling, and may last from 1 to 23 years. Some tumours may be an incidental finding, pathological fractures have been reported in one-quarter of cases. The commonest sites are the femur, humerus and tibia. This low-grade tumour shows a geographic lytic epiphyseal lesion with extension to the metaphysis. The margins can be well-defined, but indistinct or sclerotic margins have also been described. Calcifications of the tumoral matrix are not always present. There is no extension to the soft tissues. Periosteal reactions are unusual. CT may be useful to depict lobulated margins and calcified matrix. MRI shows well-delineated low signal on T1 weighted images and heterogeneous high signal on T2 images. The main differential diagnosis is the chondroblastoma, which is a smaller lesion in younger patients. The other differential diagnoses include giant cell tumours and other epiphyseal tumours.

The treatment is radical surgery. The prognosis is good with a 5-year survival of 92%^[11], even though metastases are found in 15% of cases (lung, brain and bones).

Dedifferentiated chondrosarcoma

This form represents 10–12% of all chondrosarcomas^[1]. It is characterised by a special histology and very poor prognosis. The biphasic tumour associates a low-grade chondrosarcoma with a non-chondroid high-grade sarcoma^[1]. Pain and swelling are the usual clinical symptoms as well as pathological fractures^[12]. The commonest locations are the femur, the acetabular regions and the proximal humerus.

These metaphyseal or diaphyseal lesions are rapidly destructive. The osteolytic lesion may predominate, but usually the lytic area is associated with calcifications resulting in the bimorphic pattern^[1]. A huge soft-tissue mass without calcifications, seen on CT or MRI is also indicative for this diagnosis. Imaging helps to direct biopsy of the lytic area in order to improve the histological diagnosis. The treatment involves surgery and adjuvant chemotherapy or radiotherapy. The prognosis is very poor, with an overall 5-year survival rate of only 8.5–13%. The metastases appear in the lungs but also in unusual sites such as the adrenal gland, brain and liver.

Secondary chondrosarcoma

Twelve per cent of all chondrosarcomas are developed in a pre-existing lesion. It may be secondary to: a solitary osteochondroma, osteochondromatosis, enchondromatosis (Ollier's disease), fibrous dysplasia, Paget's disease, irradiated bone or synovial chondromatosis^[1]. In osteochondromatosis the risk of sarcomatous transformation is 5–25%, it is 25–50% in enchondromatosis and nearly 100% in Maffucci's syndrome^[12].

The increased size of an enchondroma, the appearance of a lytic area with cortical destruction, associated with pain or fracture all suggest transformation. An enlarging exostosis associated with pain, the appearance of a less mineralized zone in the cartilage cap, calcifications in the soft tissues, the thickening of the cap (>1 cm) on CT and MRI suggest sarcomatous transformation^[13].

Conclusions

Radiologists should be aware of the different aspects of chondrosarcomas because they are the commonest malignant bone tumour in adults, and if detected early the prognosis can be much improved. The treatment is radical surgery. These lesions are not sensitive to the chemotherapy used in dedifferentiated, mesenchymal or metastatic forms. Radiotherapy is an alternative therapy to incomplete surgery. The staging must be accurate in order to delineate the limits of the tumour. Follow-up is done by plain films and MRI, even if the metallic prosthesis may generate artifacts.

References

- [1] Krishnan K. Dahlin's Bone tumors: General Aspects and Data on 11087 cases, 5th edn. Philadelphia: Lippincot-Raven publishers, 1996: 71–115.
- [2] Varma DG, Ayala AG, Carrasco CH, Guo SQ, Kumar R, Edeiken J. Chondrosarcoma: MR imaging with pathologic correlation. *Radiographics* 1992; 12: 687–704.
- [3] Welkerling H, Kratz S, Delling G, Ewerbeck V. Differentiation of enchondroma and low-grade chondrosarcoma-clinical and radiological findings in 34 cases. *Sarcoma* 2002; 6: S9–EMSOS abstract A09
- [4] Geirmaerd MJA, Hermans J, Bloem JL *et al*. Usefulness of radiography in differentiating enchondroma from grade I chondrosarcoma. *AJR* 1997; 169: 1097–104.
- [5] Murphey MD, Flemming DJ, Boyea SR, Bojescul JA, Sweet DE, Temple HT. Enchondroma versus Chondrosarcoma in the appendicular skeleton: differentiating features. *Radiographics* 1998; 18: 1213–37.
- [6] Eefting D, Geirmaerd MJA, Le Cessie S, Taminiu AHM, Hogendoorn PCW. Diagnostic impact of histologic parameters in differentiating enchondroma from grade I central chondrosarcoma. *Sarcoma* 2002; 6: S1–EMSOS abstract A02
- [7] Bovee JV, Kok P, Van den Broek LJ, Hogendoorn PCW. Immunohistochemistry as a tool to distinguish osteochondroma and grade I peripheral chondrosarcoma. *Sarcoma* 2002; 6: S1–EMSOS abstract A03
- [8] Vanel D, De Paolis M, Monti C, Mercuri M, Picci P. Radiological features of 24 periosteal chondrosarcomas. *Skeletal Radiol* 2001; 30: 208–12.
- [9] Forest M, Tomeno B, Vanel D. Orthopedic surgical pathology: diagnosis of tumors and pseudotumoral lesions of bones and joints, Edinburgh: Churchill Livingstone, 1998: 233–91.
- [10] Justin Q. Mesenchymal Chondrosarcoma of the Maxilla. *AJR* 2002; 179: 1077–8.
- [11] Fechner RE, Mills EM. Cartilaginous lesions. In: Tumors of the Bones and Joints, 3rd edn., vol. 8, Fechner R E, Mills E M, eds. Washington, DC: Armed Forces Institute of Pathology, 1993.
- [12] Anract P, Tomeno B, Forest M. Chondrosarcomes différenciés. Etude de treize cas cliniques et revue de la littérature. *Rev Chir Orthop* 1994; 80: 669–80.
- [13] Hudson TM, Springfield DS, Spanier SS, Enneking WF, Hamlin DJ. Benign exostoses and axostosis chondrosarcomas, evaluation of cartilage thickness by CT. *Radiology* 1984; 153: 595–9.