

# Paraspinal mass in a child

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A nine-year-old girl was admitted to our hospital with difficulty in walking in September 1993. On physical examination there was a solid mass of 10 × 10 cm in the lumbosacral region and the patient was unable to walk or stand. Bilateral deep tendon reflexes of the lower extremities were hypoactive and muscle strength was also decreased. There was hypoesthesia at L4, L5 and S1 on both legs. Babinsky test was negative bilaterally. On lumbar spine radiographs there were lytic and sclerotic lesions on the corpus of the fifth lumbar vertebra and sacrum. Results of bone scintigraphy showed intense tracer accumulation at the L5 and L5-S1 level on both blood pool and delayed images.

Magnetic resonance imaging (MRI) of the lumbosacral spine showed a large mass which invaded the multifidus and both psoas muscles. The mass extended caudally to the pelvis. Both intervertebral foramina were dilated at L4-5, L5-S1, and S1-2 levels and the right intervertebral foramen on S2-3 level (figure 1). The mass extended into the spinal canal through these foramina and filled the spinal canal between L5 and S3 (figure 2). The mass showed high-signal intensity on T2-weighted images and isointensity with adjacent muscles on T1-weighted images. After contrast enhancement with gadolinium DTPA the mass showed enhancement almost homogeneously (figure 3). The soft tissue component of the mass extended between L4 and S3 levels. The fifth lumbar and the first two sacral vertebrae were of low signal intensity on T1-weighted and of high signal intensity on T2-weighted images. Both L5 and S2 vertebral bodies showed inhomogeneous contrast enhancement but S1 vertebra showed only a few enhanced regions (figure 3). Left L5 hemilaminectomy, left laminotomy and biopsy from the epidural mass were performed.

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## Questions

- 1 What is the diagnosis?
- 2 What are the treatment options?



**Figure 1** T1-weighted right paramedian sagittal MRI. The intervertebral foramina are dilated and filled with mass on L4-5 and L5-S1 levels (arrows). Note the intermediate signal intensity (SI) of the mass and lower SI of the L5 vertebra



**Figure 2** Median T2-weighted sagittal MRI. The hyperintense mass fills the spinal canal between L4-S3 levels (arrowheads). L5, S1 and S2 vertebral bodies show high SI



**Figure 3** Contrast-enhanced T1-weighted median sagittal MRI. The mass shows contrast enhancement (arrows). L5 and S2 vertebral bodies are also enhanced

## Answers

### QUESTION 1

Pathological examination revealed mesenchymal chondrosarcoma characterised by undifferentiated mesenchymal cells and islands of malignant cartilage differentiation (figures 4 and 5).

### QUESTION 2

Surgery is the major method of treating the disease. Chemotherapy and radiotherapy may be used as additional therapeutic modalities.

## Discussion

Chondrosarcoma is primarily a tumour of adulthood. Its occurrence in children is uncommon.<sup>1</sup> Mesenchymal chondrosarcoma is a rare variant of chondrosarcoma which acts biologically and, in its response to treatment, in a completely different way from the more common differentiated forms.<sup>2</sup> It is about 70 times less common than the intramedullary osteosarcoma, and five times less common than fibro- or osteosarcomatous transformation of chondrosarcoma.<sup>3</sup>

### CLINICAL FEATURES

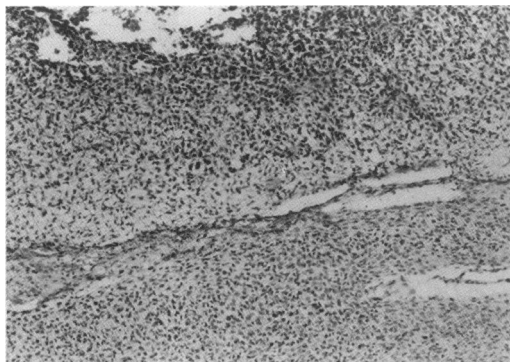
About 80% of patients are younger than age 40 years. There is no gender predilection. Approximately 0.5% of biopsy-analysed primary bone tumours are mesenchymal chondrosarcomas. It is about one tenth as common as conventional chondrosarcomas. Lesions range from 1 to 40 cm (average about 10 cm) in longest dimension. The most common sites of involvement are shown in box 1. The long bones are affected in only 28% of cases. Most

#### Mesenchymal chondrosarcoma: sites of involvement

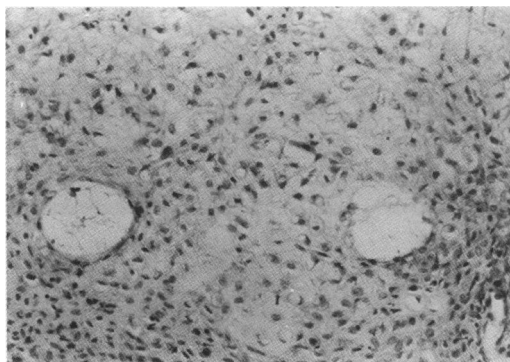
- femur 18%
- ribs 18%
- jaw 16%
- spine 8%
- pelvis 8%

Box 1

**Figure 4** Mesenchymal chondrosarcoma rich with spindle tumour cells. (H&E, original  $\times 230$ )



**Figure 5** Photomicrograph showing the chondroid areas in the mesenchymal chondrosarcoma. (H&E, original  $\times 460$ )



lesions begin as solitary bone lesions, although lesions of soft tissue and multifocal osseous origin have been reported.<sup>3,4</sup> About 66% of lesions originate from bones or meninges and the remaining from the soft tissues.<sup>3</sup>

### SIGNS AND SYMPTOMS

Pain (80% of patients) and/or swelling (70%) are the dominating clinical stigmata. Pain may be of a month to five or more years duration. More than 50% of patients have experienced pain for less than six months when first evaluated. If the tumour is near a joint, instability, limitation of motion, stiffness, and effusion are possible. When located in the meninges, the symptoms are headache (intracranial), cranial nerve palsies and visual symptoms (base of skull), and pain, stiffness, sensory and motor deficits, and subarachnoid haemorrhage (spinal).

### RADIOLOGIC FEATURES

The radiologic manifestations (box 2) are not diagnostic. Definitive diagnosis depends on biopsy. In about two-thirds of cases, radiographs of lesions exhibit stippled densities, which are highly suggestive of the presence of cartilage. The radiologic appearance usually prompts a diagnosis of chondrosarcoma or osteosarcoma, as mesenchymal chondrosarcoma has no pathognomonic radiologic features, and is less common than the former tumours.

### DISEASE COURSE

This tumour is fully malignant, and carries a less favourable prognosis than conventional chondrosarcomas.<sup>3</sup> Fewer than 30% of patients treated by surgical methods are alive after 10 years. Patients must undergo radical surgical

#### Mesenchymal chondrosarcoma: radiologic appearance

- intraosseous primary lesions: most often associated with large, lobulated soft tissue mass; remaining lesions are predominantly lytic
- periosteal lesions: most often demonstrate a large, juxtacortical, lobulated soft tissue mass with prominent coarse, stippled or short linear densities; remaining lesions are predominantly lytic

Box 2

#### Mesenchymal chondrosarcoma: histologic findings

##### Constant findings

- undifferentiated mesenchymal cells
- multifocal differentiation into malignant cartilage

##### Variable histologic findings

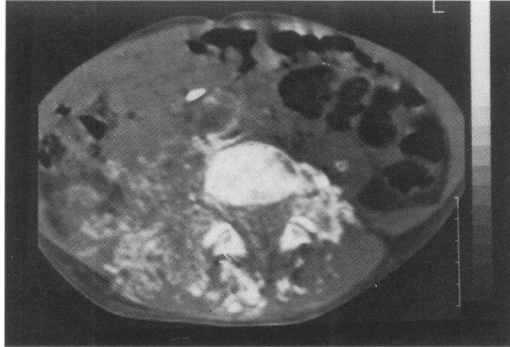
- haemangiopericytoma-like features
- herringbone spindle cell pattern
- 'pushing' versus permeative borders
- necrosis and haemorrhage
- osseous metaplasia

Box 3

### Mesenchymal chondrosarcoma: differential diagnosis

- chondrosarcoma with fibrosarcomatous transformation
- osteosarcoma
- fibrosarcoma
- chondroblastoma
- chondromyxoid fibroma
- Ewing's sarcoma

#### Box 4



**Figure 6** Axial CT image at L5 level. The paravertebral mass, and the intraspinal component show speckled calcification. The right L5-S1 intervertebral foramen is enlarged by the mass

procedures to reduce the rather high incidence of local recurrence. Metastases to the lungs and other bones (predominantly spine, skull and ribs) may be seen as late as 10 years after surgery. Metastases to lymph nodes rarely occur.<sup>3</sup> Some variants of the tumour respond favourably to chemotherapeutic agents.

In our patient, radiotherapy was administered to L3-S2 region to a 11 × 8 cm area, with Co-60 at a dose of 4680 cGy in October 1993. Four courses of chemotherapy with cisplatin (120 mg/m<sup>2</sup>) and doxorubicin (30 mg/m<sup>2</sup> daily for two days every three weeks) were administered.

In January 1994, on control examination, plain abdominal X-ray showed paraspinal heavy calcification. On computed tomograph (CT) examination the mass was enlarged into the abdomen and pelvis and showed heavy calcification with a speckled pattern (figure 6). The intraspinal component of the mass showed the same pattern of calcification. Lytic lesions

were present in the fifth lumbar and first two sacral vertebrae. On thoracic CT, 1 × 2 cm mass was observed at the right paracardiac region. She died of progressive disease in March 1994.

In our patient MRI clearly showed the extension of the tumour with its multiplanar imaging capability and good soft tissue resolution. Plain abdominal X-rays and especially CT showed heavy calcifications of the tumour. MRI failed to show these calcifications, but after contrast enhancement, nonenhanced irregular foci, which were thought to belong to these calcifications, were present. MRI was better than CT in showing the three-dimensional localisation of the mass because of its multiplanar imaging capability. The intraspinal component of the tumour could be better evaluated with MRI, especially because of its sagittal images. MRI was therefore a better imaging modality for evaluation of the tumour extension but failed to show the calcification pattern that can be a clue for differential diagnosis.

Although surgery is the major method of treating the disease, radical surgery could not be performed because of the vertebral localisation of the tumour. In the literature it was reported that unresectable tumours responded to irradiation and chemotherapy. In our patient, however, local and systemic metastases were detected during chemotherapy and radiotherapy was also ineffective. Poor prognostic factors, such as pain and the central location of the tumour might be the cause of this rapid progression, as discussed in the published data.<sup>3</sup>

In conclusion, although rare in children, mesenchymal chondrosarcoma may be cured with radical surgery, chemotherapy and radiotherapy if the disease has not progressed. Radiologic examination, especially with MRI, is useful in showing the tumour extension.

#### Final diagnosis

Paraspinal mesenchymal chondrosarcoma.

**Keywords:** MRI, paraspinal mesenchymal chondrosarcoma

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