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CASE REPORT

Jennifer Rodenberg
Olaf Myhre Jensen
Johnny Keller
Ole S. Nielsen
Cody Bünger
Anne G. Jurik

Fibrous dysplasia of the spine, costae and hemipelvis with sarcomatous transformation

J. Rodenberg · O.M. Jensen · J. Keller
O.S. Nielsen · C. Bünger · A.G. Jurik
The Centre for Bone
and Soft Tissue Tumours,
University Hospital of Aarhus, Denmark

A.G. Jurik, M.D., D.M.Sc. (✉)
Department of Diagnostic Radiology R,
Aarhus Kommunehospital,
Noerrebrogade 44, DK-8000 Aarhus C,
Denmark

Abstract We describe a patient with polyostotic fibrous dysplasia and secondary malignant fibrous histiocytoma in a spinal lesion.

Key words Fibrous dysplasia · Malignant fibrous dysplasia

Introduction

Malignant transformation in fibrous dysplasia is uncommon and occurs predominantly in lesions of the femur, maxilla and mandible. It has not previously been reported in spinal lesions.

Case report

A 62-year-old woman with suspected disc herniation was admitted to the University Hospital of Aarhus. She had experienced back pain for many years and as a result, had been on a disability pension for 10 years. For the 2 months preceding her admission, she suffered severe back pain radiating to the right groin and thigh occurring relatively acutely, but had no history of trauma. Clinical examination revealed slight scoliosis and tenderness of the lumbar spine and paravertebral muscles, in addition to decreased sensitivity on the lateral side of the femur. There was no abnormal skin pigmentation. Results of laboratory examinations, including alkaline phosphatase, were normal.

Radiography of the spine showed lytic destruction of the third lumbar vertebral body, in addition to well-demarcated lytic expansive lesions in the two upper lumbar and the T8–12 vertebrae (Fig. 1). A supplementary skeletal survey revealed large expansive lytic areas in the posterior part of the sixth and eight to tenth costae on the right side and the tenth costa on the left side (Fig. 1). It also revealed hyperostosis and irregular sclerotic bone structure corresponding to the left-sided pelvic bones, but no peripheral lesions. CT of the chest, spine and pelvis revealed a large soft-tissue mass related to osseous destruction at the right side of the L3 vertebra. The other spinal, pelvic, and costal lesions were well-demarcated without accompanying soft-tissue masses or other signs suggesting malignancy (Fig. 2). The chest CT revealed multiple, small pulmonary metastases.

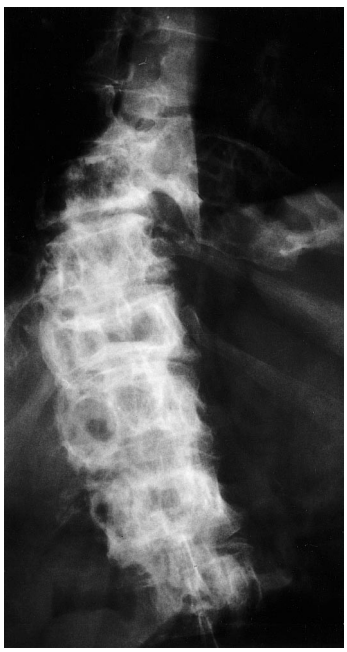

Fig. 1 Anterior-posterior (AP) view of the upper lumbar and lower thoracic spine, showing lytic destruction of the third lumbar vertebral body on the right side, in addition to well-demarcated expansive lytic areas in the other vertebrae and in the posterior part of the left tenth costa

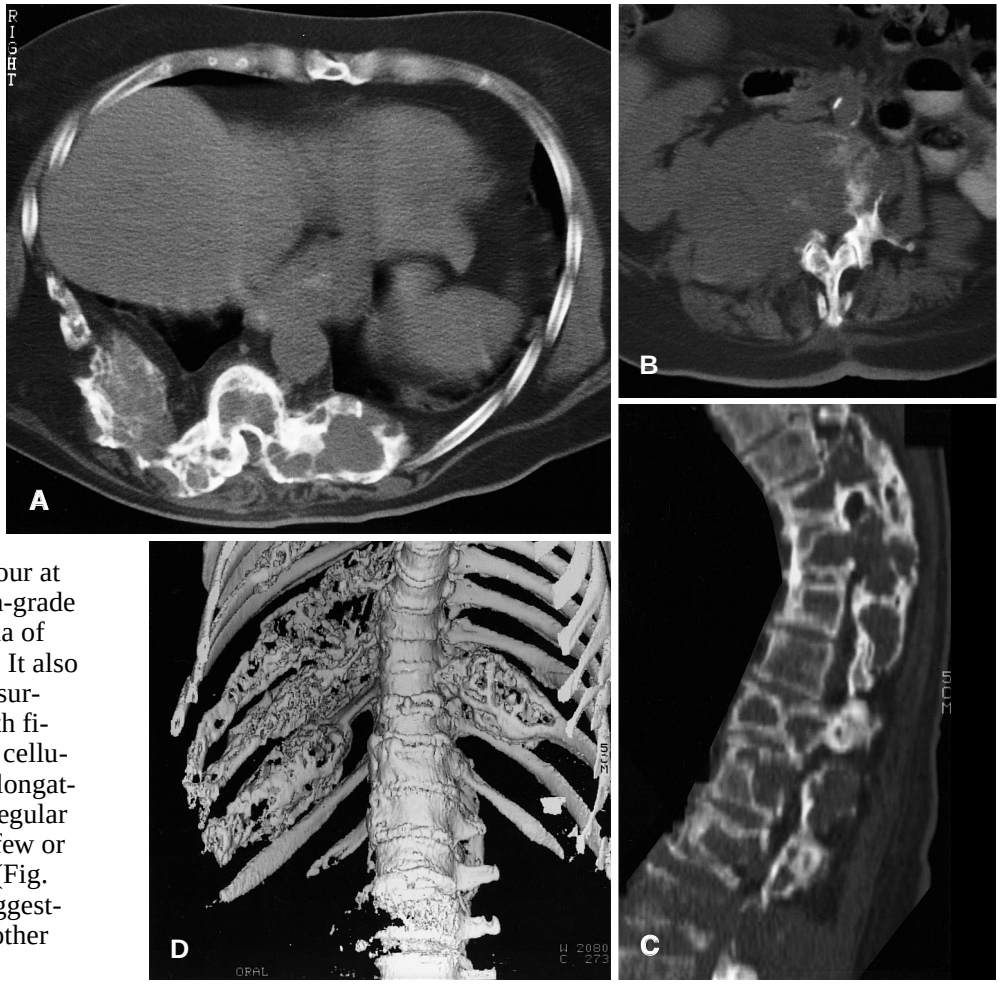

Fig. 2 CT scans at the level of the tenth costae (**A**) and L3 vertebra (**B**), in addition to a sagittal view (**C**) and 3D reconstruction (**D**) of the lower thoracic and upper lumbar region, demonstrating lytic destruction of the L3 vertebra with a soft tissue mass at the right posterior aspect. Also apparent are lytic well-demarcated expansive lesions in T8–12 and L1–2, in the posterior part of the sixth and eight to tenth costae on the right side, and the tenth costa on the left side

Surgical biopsy of the tumour at the L3 vertebra showed a high-grade malignant fibrous histiocytoma of the pleomorphic type (Fig. 3). It also revealed abnormalities of the surrounding bone compatible with fibrous dysplasia, consisting of cellular fibrous tissue containing elongated cells, intermingled with irregular spiculae of woven bone with few or no osteoblasts on the surface (Fig. 4). There were no changes suggesting osteosarcoma. Biopsy of other lesions was not performed.

Palliative lumbar radiotherapy was planned, but the patient suddenly died of pulmonary insufficiency.

Discussion

Fibrous dysplasia is a benign fibro-osseous aberration of the skeletal system, in which medullary bone is replaced by one or more areas of fibrous tissue, within which an osseous element may develop. It may affect single segments (monostotic) or cause widespread, diffuse and generalized skeletal involvement (polyostotic). Polyostotic fibrous dysplasia accounts for about 20% of all fibrous dysplasia. Vertebral lesions are uncommon and are usually associated with polyostotic fibrous dysplasia [1, 2]. The frequency of involvement of the cervical and lumbar vertebrae, as part of polyostotic fibrous dysplasia, has been reported to be 7% and 14%, respectively [3]. It may result in progressive debilitation and deformity [4].

The radiographic features depend upon the phase of evolution. In the bone-destroying or bone-replacing phase, radiolucent, often cyst-like, areas surrounded by a sclerotic border are characteristic [2, 3, 5]. The spongiosa is widened and the cortex thinned and expanded. Blunt septae and small flecks of calcification may be identified occasionally within the lucent zones, which may make the differential diagnosis against enchondroma difficult. In the bone-forming phase, zones of new bone are distributed irregularly, but are mainly confined to the spongiosa. Bone-forming and bone-destroying lesions are often visible simultaneously. The presence of large lytic lesions in the costae and thoracic vertebrae may be considered pathognomonic for fibrous dysplasia. The absence of mottled calcifications differentiate them from enchondromas.

A histologically diverse mixture of fibrous connective tissue and new bone is seen [5].The bone trabeculae are thin and irregular, and the fibrous tissue consists of spindle cells in a whorled arrangement that may be interspersed with trabeculae of new bone, changes consistent with those of the present patient.

Malignant transformation is a distinctly unusual complication. The incidence of malignant metaplasia in fibrous dysplasia may be difficult to determine, but has been estimated to

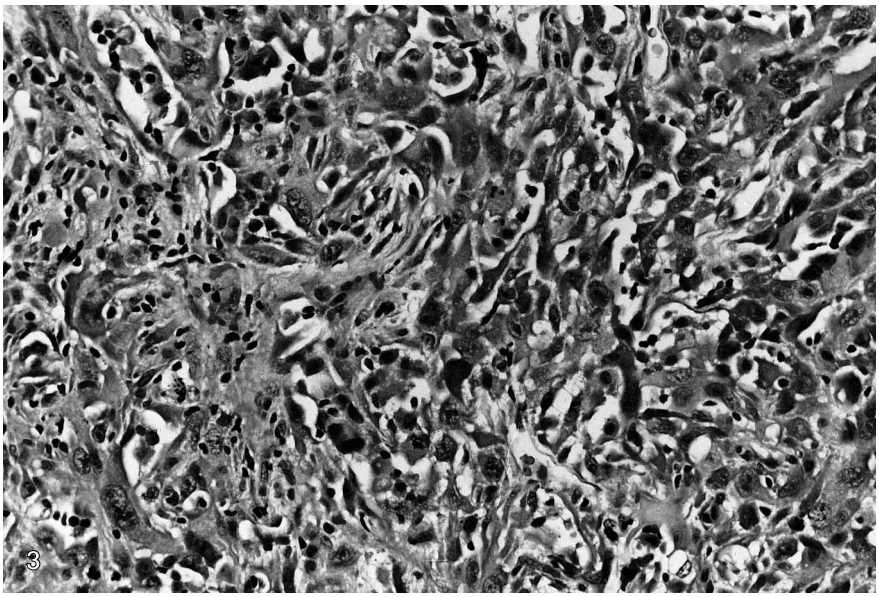

Fig. 3 Biopsy from tumour in relation to L3: pleomorphic and anaplastic sarcoma without formation of osteoid or chondroid compatible with malignant fibrous histiocytoma (haematoxylin-eosin, ×1000)

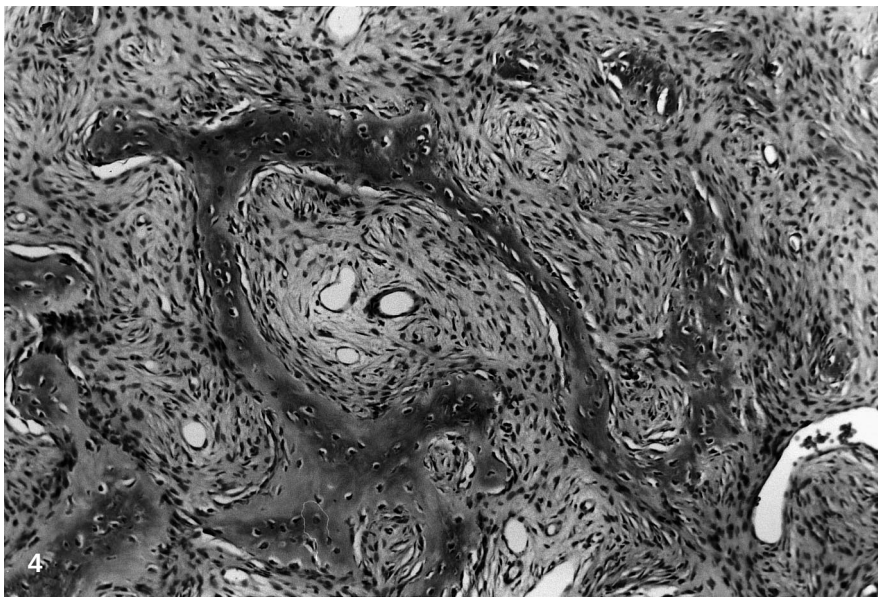

Fig. 4 Biopsy from L3 showing fibrous dysplasia: cellular fibrous stroma with metaplastic bone formation. Primitive trabeculae of woven bone without osteoblastic rim can be seen (haematoxylin-eosin, ×500)

occur in less than 1% of the cases [6, 7]. Malignancies occur predominantly in lesions of the femur and maxilla/mandible [6–11], sometimes following radiotherapy for fibrous dysplasia [6, 7, 9, 11]. Malignant transformation in a spinal lesion has to our knowledge not been reported previously. The most common malignant tumour is osteosarcoma, followed by fibrosarcomalike tumours, as seen in the present patient, and then chondrosarcoma [6, 7, 10–12]. Malignant transformation must be suspected in the case of endosteal and cortical erosion, especially if there is cortical destruction with associated soft-tissue mass, as seen in the present patient. CT or MRI can help in the evaluation of a clinically or radiographically suspicious lesion, as well as in planning the therapeutic approach and mapping the extent and location of the tumour, if malignant transformation has taken place.

In summary, a case of fibrous dysplasia involving the spine, costae and hemipelvis with malignant transformation of a vertebral lesion is described.

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