Malignant fibrous histiocytoma arising from a calcifying enchondroma – a case report

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Accepted: 9 November 1996

Summary. A man, 79 years of age, developed a malignant fibrous histiocytoma which arose in relation to a calcifying enchondroma of the distal femur. Radiographs showed a fracture through an intensely calcified bony tumour with no bony destruction. The clinical diagnosis was a dedifferentiated chondrosarcoma. Histology showed an anaplastic sarcoma with calcified tissue showing extensive areas of necrosis and degeneration. No chondrosarcomatous foci were found.

Resumé. Nous rapportons le cas d'un homme de 79 ans, qui a développé un histiocytofibrome malin à partir d'un enchondrome calcifié du fémur distal. L'examen radiologique a montré une fracture au sein d'une tumeur très calcifiée sans association de destruction osseuse. Cette lésion apparaissait cliniquement être un chondrosarcome dédifférencié. Mais l'étude histologique a montré un sarcome anaplasique avec un tissu cartilagineux calcifié présentant des plages étendues de nécrose et de dégénérescence. Il n'y avait pas de foyer chondrosarcomateux visible.

Introduction

Secondary sarcomas arising in relation to a solitary enchondroma are uncommon [5, 6, 8], and it is not known whether such lesions are dedifferentiated chondrosarcoma [5, 6, 8].

We report a case of malignant fibrous histiocytoma arising from a calcifying enchondroma and review the literature.

Case report

A Japanese man, 79 years of age, presented with a pathological fracture of the distal end of his right femur. Laboratory investigations were normal, except for a positive HBs antigen.

Radiographs showed that the fracture was through a calcified lesion, 10 cm×4 cm in size (Fig. 1). There was no cortical destruction or periosteal reaction. CT showed a zone of soft tissue density in the bone marrow with scattered calcification in its centre and a swelling of the soft tissue surrounding the fracture (Fig. 2). A bone scan revealed a hot spot at the site. Angiography was normal.

At operation, the tumour was curetted and the fracture fixed with a condylar plate. Histology showed a large amount of necrotic or degenerate cartilaginous tissue with foci of irregular calcification and focal proliferation of anaplastic spindle-shaped cells without an osteoid or chondroid matrix (Fig. 3). The anaplastic tumour was next to viable cartilage with no evidence of nuclear atypism (Fig. 4).

Disarticulation was carried out at the hip joint, followed by radiotherapy with a dose of 20 Gy. Histology of the specimen (Fig. 5, 6) showed extensive proliferation of spindle-shaped cells occasionally with a storiform pattern compatible with a malignant fibrous histiocytoma (MFH). Further radiotherapy (50 Gy) was given to the site of the operation.

The patient died 8 months after the first operation. Multiple metastases were found at autopsy involving the lungs, pleurae, serous pericardium, liver, left adrenal gland, right ureter, vertebral column, sternum, ribs, para-aortic and the left supraclavicular lymph nodes. All the metastatic lesions consisted of a MFH-like anaplastic sarcoma without a cartilaginous component.

Discussion

Secondary sarcomas arising from areas of bone infarct or Paget's disease are well recognised [4,

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Fig. 1. Radiographs showing a pathological fracture through a lesion in the distal femur. The appearance was compatible with a calcifying enchondroma with multiple flocculent areas of calcification

Fig. 2. CT showing intense intraosseous calcification and swelling of the soft tissue surrounding the femur. There is no cortical destruction or extraosseous tumour

Fig. 3. Photomicrograph showing necrotic or degenerate cartilage tissue with foci of irregular calcification. The appearances are compatible with a calcifying enchondroma (HE, $\times 40$)

Fig. 4. Photomicrograph showing anaplastic tumour clearly abutting the cartilage without atypical cells (HE, $\times 40$)

Fig. 5. Photomicrograph showing interlaced bundles of spindle-shaped anaplastic cells in a storiform pattern (HE, $\times 20$)

Fig. 6. Photomicrograph showing proliferation of anaplastic cells with marked nuclear pleomorphism and abundant foamy cytoplasm consistent with a malignant fibrous histiocytoma (HE, $\times 40$)

9], but they rarely arise from a solitary enchondroma and only 4 osteosarcomas, 4 fibrosarcomas and 2 MFHs have been reported [5, 6, 8]. According to Sanerkin and Woods secondary sarcoma do not appear to arise directly from neoplastic cartilage, but from dense fibrous tissue surrounding necrotic areas of enchondroma by a process analogous to that responsible for the development of secondary sarcoma in bone infarcts and chronic osteomyelitis [6]. Smith et al have stated that these tumours which arise in relation to an enchondroma represent a collision tumour, but they also raised the possibility that the lesion could be a dedifferentiated chondrosarcoma derived from a benign neoplasm [8]. Mirra and Marcove described them as dedifferentiated chondrosarcoma developing in an enchondroma [3].

The features of secondary sarcoma arising from enchondroma are: (1) the radiographs show a heavily calcified lesion in one of the major long bones; (2) patients are usually over 50 years of age; (3) the prognosis is very poor; (4) histologically a highly malignant sarcoma is seen near to cartilage.

Dedifferentiated chondrosarcoma was first described by Dahlin and Beabout in 1971 who defined them as highly anaplastic sarcoma juxtaposed to a low grade or borderline chondrosarcoma [1], with a radiographic appearance similar to that of an ordinary chondrosarcoma associated with areas of osteolysis [1, 2].

Schajowicz described highly calcified benign cartilaginous lesions in the metaphysis of a long bone which behaved differently from ordinary enchondroma, and he assumed that they were not

genuine neoplasms but cartilaginous hamartoma [7]. The most common sites were the proximal humerus and distal femur and radiographs showed dense ring-like, or flocculent, radio-opacities.

The radiographic features, except for the pathological fracture, supported the diagnosis of a benign cartilaginous lesion with intense calcification in our case, and histology failed to demonstrate any chondrosarcomatous foci.

Acknowledgements. The authors wish to thank Mr S. Suzuki for his technical assistance.

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