CASE REPORT

Dedifferentiated chondrosarcoma with telangiectatic osteosarcoma-like features

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A 35-year-old Japanese man was admitted to the National Cancer Center, Tokyo, Japan, in December 2000, with a 2-month history of pain around the left thigh. Radiographs showed a poorly demarcated osteolytic lesion with focal mineralisation and endosteal scalloping in the left proximal femur. Biopsy showed a proliferation of highly anaplastic cells without any cartilaginous component. A wide excision of the left proximal femur with a replacement by endoprosthesis was carried out in February 2001 after treatment with methotrexate and 20 Gy radiation therapy. Pathological examination of the surgical specimen showed a focus of low-grade chondrosarcoma and the coexistence of telangiectatic osteosarcoma-like features. The patient was diagnosed with dedifferentiated chondrosarcoma with telangiectatic osteosarcoma-like features. Lung metastasis appeared in July 2001 despite an adjuvant chemotherapy including methotrexate, cis-platinum and doxorubicin. The latest follow-up study in June 2004 showed multiple lung metastases. Establishing a definitive diagnosis of dedifferentiated chondrosarcoma may be difficult with limited small biopsy specimens. Dedifferentiated chondrosarcoma should be included in the differential diagnosis of osteolytic tumours with focal calcification and endosteal scalloping even if an extrasosseous tumour component is not identified.

The histological features of dedifferentiated chondrosarcoma may be those of conventional osteosarcoma, malignant fibrous histiocytoma, fibrosarcoma or anaplastic spindle-cell sarcoma.^{1,2} Telangiectatic osteosarcoma is a relatively rare subtype of osteosarcoma, and there is only one case of its presence in a dedifferentiated chondrosarcoma in the literature in English.³ We present a case of dedifferentiated chondrosarcoma in which the dedifferentiated component of the tumour showed a close histological resemblance to a telangiectatic osteosarcoma.

CASE REPORT

A 35-year-old Japanese man was admitted to the National Cancer Center, Tokyo, Japan, in December 2000, with a 2-month history of pain around the left thigh. Radiographs showed a poorly demarcated osteolytic lesion with a focal calcification and cortical thickening in the left proximal femur (fig 1). A subsequent computed tomography scan showed the osteolytic lesion in the femur, with focal calcification and endosteal scalloping (fig 2). On magnetic resonance images, the lesion measuring 7×4×3 cm showed isosignal intensity on T1-weighted images and high signal intensity on T2-weighted images (fig 3); no extrasosseous soft-tissue mass was observed. Biopsy results showed a proliferation of atypical cells without any cartilaginous component. We treated the patient with methotrexate (8 g/m²) and

20 Gy radiation therapy. As these were not effective, a wide excision of the left proximal femur was carried out in February 2001, with a replacement by endoprosthesis (fig 4).

Figure 1 Radiographs showing a poorly demarcated osteolytic lesion, with focal mineralisation and cortical thickening in the left proximal femur.

Figure 2 Computed tomography scan showing the osteolytic lesion in the femur, with focal mineralisation and endosteal scalloping.

Figure 3 Magnetic resonance images of the osseous lesion showing iso-signal intensity on T1-weighted images and high signal intensity on T2-weighted images. No extraosseous soft-tissue mass can be identified.

The surgical specimen showed multiple blood-filled cysts (fig 5A). Intervening septa contained malignant spindle and plump cells. Small scattered foci of osteoid were seen in the septa (fig 5B). In addition, the focal area showed a well-differentiated cartilaginous tumour with grade 1 features, and a sharp margin was observed between the multiple blood-filled cystic structures and the low-grade chondroid part (fig 5C). No tumour necrosis associated with systemic chemotherapy was observed. The overall appearance was that of a dedifferentiated chondrosarcoma with telangiectatic osteosarcoma-like features. Lung metastasis appeared in July 2001 despite adjuvant chemotherapy including methotrexate, *cis*-platinum and doxorubicin. The latest follow-up study in June 2004 showed multiple lung metastases.

DISCUSSION

Up to 11% of low-grade chondrosarcomas contain an additional component of high-grade mesenchymal sarcoma.

Figure 4 Radiograph after the surgery. A wide excision of the left proximal femur was carried out, with a replacement by endoprosthesis.

In 1971, the term dedifferentiated chondrosarcoma was introduced by Dahlin and Beabout¹ to describe this variant. The importance of recognising this dedifferentiated component is related to its locally aggressive behaviour, frequency of recurrence and, ultimately, its poor prognosis. The term dedifferentiation has been questioned by some authors who prefer the term chondrosarcoma, with additional mesenchymal component.⁴ The histological features of dedifferentiated chondrosarcoma may be those of conventional osteosarcoma, malignant fibrous histiocytoma, fibrosarcoma or anaplastic spindle-cell sarcoma.¹⁻² Several cases with rare

Figure 5 (A) Surgical specimen showing multiple blood-filled cysts (original magnification $\times 40$, haematoxylin and eosin). (B) Intervening septa containing malignant spindle and plump cells. Small scattered foci of osteoid can be seen in the septa ($\times 400$). (C) The sharp border between the multiple blood-filled cystic structures and the low-grade chondroid part ($\times 100$).

components such as giant-cell tumour⁵ or rhabdomyosarcoma⁶ have been reported in the literature.

Dedifferentiated chondrosarcoma with a component mimicking a telangiectatic osteosarcoma is extremely uncommon and only one case has been reported in the literature. In 1999, Radhi and Loewy³ reported a dedifferentiated chondrosarcoma with a component mimicking a telangiectatic osteosarcoma in the body of the left scapula of a 44-year-old woman. Although the site affected in our patient and that in the case reported by Radhi and Loewy was different, the ages of these two patients were less than usual, suggesting that young people may be uniquely prone to this unusual variant of dedifferentiated chondrosarcoma. In general, dedifferentiated chondrosarcoma usually occurs in patients aged >50 years.^{1,4}

On radiographs, characteristic features in our patient were cortical thickening, endosteal scalloping and focal calcification, suggesting chondrosarcoma. Similarly, in the case reported by Radhi and Loewy,³ endosteal scalloping and small calcification were noted, especially in the computed tomography scan. In the largest study, calcification was found in around 50% of the lesions and an extraosseous mass was observed in roughly 55% of tumours.² Although the biopsy result of our patient showed only highly malignant cells, excluding a diagnosis of conventional low-grade chondrosarcoma, we should consider the possibility of dedifferentiated chondrosarcoma in patients with focal calcification and endosteal scalloping. In addition, the lack of an extraosseous tumour component does not exclude the diagnosis of dedifferentiated chondrosarcoma.

An essential histological feature of dedifferentiated chondrosarcoma is an abrupt interface between the low-grade cartilaginous and high-grade mesenchymal components. As the microscopic findings focally showed a low-grade chondrosarcoma, the diagnosis of dedifferentiated chondrosarcoma with telangiectatic osteosarcoma-like features was made. As pathological examination of this tumour showed foci of haemorrhagic cysts without endothelial cell lining, differential diagnosis included an aneurysmal bone cyst. However, the septa intervening in the diffuse blood-filled spaces contained highly malignant spindle cells with focal osteoid formation, excluding the possibility of an aneurysmal bone cyst.

The prognosis of dedifferentiated chondrosarcoma is poor. The 5-year survival rate ranges from 10.5% to 18%.^{2,7} The prognosis of telangiectatic osteosarcoma is similar to that of conventional osteosarcoma, although their prognosis was

extremely poor in the primary report in 1976.^{4,8} Good responses to systemic chemotherapy have been reported recently in patients with telangiectatic osteosarcoma. The response to systemic chemotherapy in our patient was poor. In general, dedifferentiated chondrosarcomas have not shown much response to chemotherapy. Some authors reported relatively good clinical results in patients treated adequately with surgery and systemic chemotherapy, although the preoperative chemotherapy effect was poor in 80% of patients.⁷ The dedifferentiated component resembling telangiectatic osteosarcoma may be resistant to chemotherapy.

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Informed consent was obtained for publication of the patient's details in this report.

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