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CASE REPORT

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Sarcomatoid chordoma: chordoma with a massive malignant spindle-cell component

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Abstract We report a case of chordoma containing a spindle cell sarcomatoid component with a gradual transition from conventional chordoma. Immunohistochemically, many tumor cells in both conventional chordoma and sarcomatoid components were positive for cytokeratins (AE1/AE3, CAM5.2) and epithelial membrane antigen as well as vimentin. This report provides a rare example of sarcomatoid chordoma. Familiarity with this type of bone tumor should help to avoid confusion with dedifferentiated chordoma and other spindle cell sarcomas or carcinomas.

Key words Chordoma · Sarcomatoid chordoma · Transitional feature · Sacrum · CT · MRI

Introduction

Chordoma is a slow-growing, low-grade malignant bone tumor that usually arises in the axial skeleton, with propensity for frequent local recurrence but relatively rare true distant metastasis [1]. Highly malignant sarcomatoid areas are rarely observed in cases of chordoma with or without radiation therapy. In the cases without irradiation, two histologically distinct groups of tumors are included; one is characterized by a transitional feature between conventional chordoma and a sarcomatoid component, referred to as chordoma with a sarcomatous transformation [2] or sarcomatoid chordoma, and the other group without a transition between two components is described as "dedifferentiated chordoma" [3]. We report an example of sarcomatoid chordoma, and the distinction between the two groups of tumor is discussed.

Case report

A 74-year-old Japanese woman had noticed low back pain for approximately 3 months prior to admission. A radiograph of the pelvis showed extensive destruction of the sacrum (Fig. 1A). The lateral radiograph of the sacrum demonstrated an expansive lytic lesion with cortical disruption. Computed tomography (CT) disclosed a sacral tumor measuring approximately 8×6 cm with inhomo-

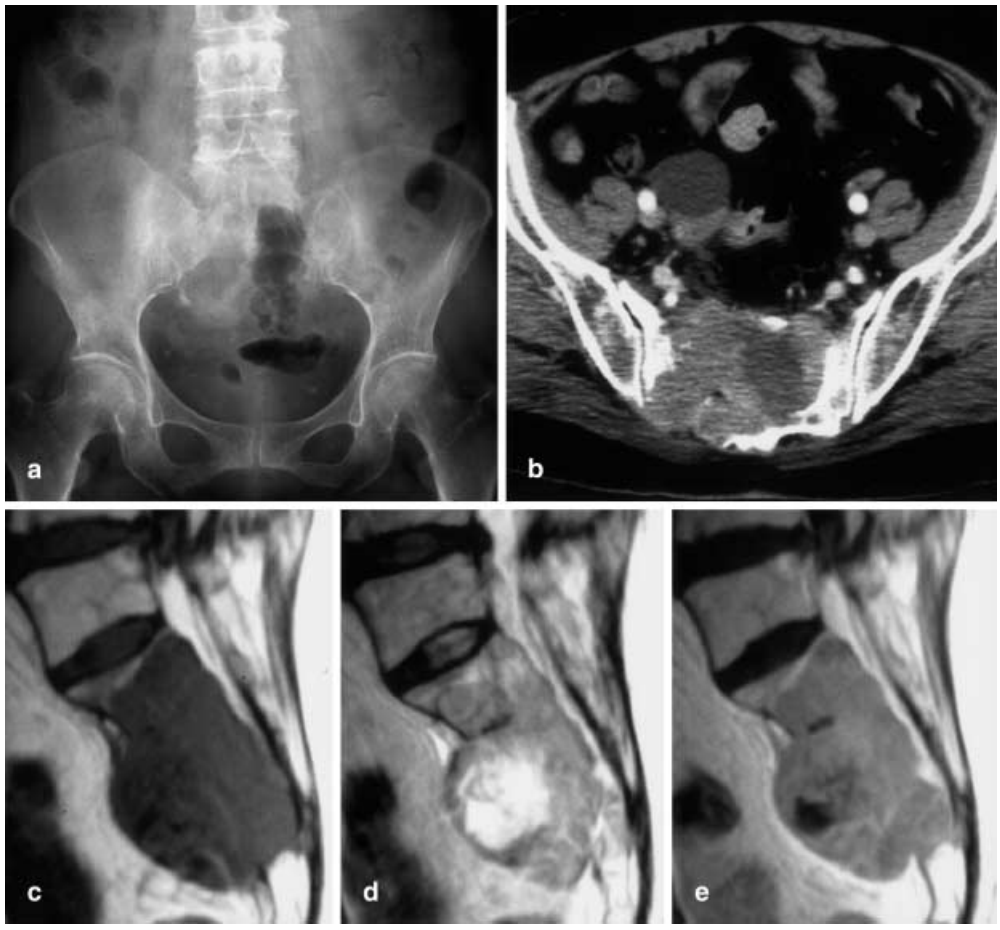

Fig. 1 **a** Radiograph of the pelvis showing extensive destruction of the sacrum. **b** CT scan showing a sacral tumor with inhomogeneous attenuation, extending into the presacral region. **c** T1-weighted sagittal MR image showing an extensive tumor with low intensity in the first to third sacral vertebrae. **d** T2-weighted MR image showing low intensity in the main part of the tumor with a central high-intensity area, without two sharply demarcated distinct components in the tumor. **e** Postcontrast T1-weighted MR image showing markedly inhomogeneous enhancement following contrast

geneous attenuation, extending into the presacral region and involving the sacro-iliac joint (Fig. 1b). The T1-weighted sagittal magnetic resonance (MR) image demonstrated an extensive tumor with low intensity in the first to third sacral vertebrae (Fig. 1c). On the T2-weighted image, the intensity of the main part of the tumor was lower than fat, and the central area exhibited very high intensity with an irregular border, probably indicating necrosis (Fig. 1d). The gadolinium-DTPA enhanced T1-weighted image displayed markedly inhomogeneous enhancement (Fig. 1e). However, there was no evidence of two sharply demarcated distinct components in the tumor on any CT or MR images. Physical and radiological examinations did not disclose malignant tumors at any other sites. A diagnosis of chordoma was clinically made and open biopsy was performed.

Microscopically, alternating cellular and loose-looking components were seen at a low-power view (Fig. 2a). The loose-looking areas were composed of cords or nests of epithelioid tumor cells in a myxoid intercellular matrix. The tumor cells had round or oval nuclei and eosinophilic cytoplasm focally with vacuolar changes and a small number of physaliferous cells (Fig. 2b). This feature was compatible with conventional chordoma. The cellular portions consisted of a proliferation of spindle cells having atypical spindle or oval nuclei arranged in short fascicles (Fig. 2c). Mitotic figures were frequently encountered in the cellular portions. Gradual transition was recognized between the two components (Fig. 2c). Immunohistochemically, many tumor cells in both components were positive for cytokeratins (AE1/AE3, CAM5.2) and epithelial membrane antigen (EMA) as well as vimentin and HBME-1 (Fig. 2d). Except for neuron-specific enolase, the tumor cells were negative for neurogenic (S-100, chromogranin A, glial fibrillary acid protein, neurofilament, synaptophysin) and myogenic (alpha-smooth muscle actin, HHF35, desmin) markers. The tumor was diagnosed as sarcomatoid chordoma, or descriptively as chordoma with massive spindle cell areas.

The patient was treated only by radiation therapy. Seventeen months after the open biopsy, the tumor had increased in size but there was no evidence of distant metastasis.

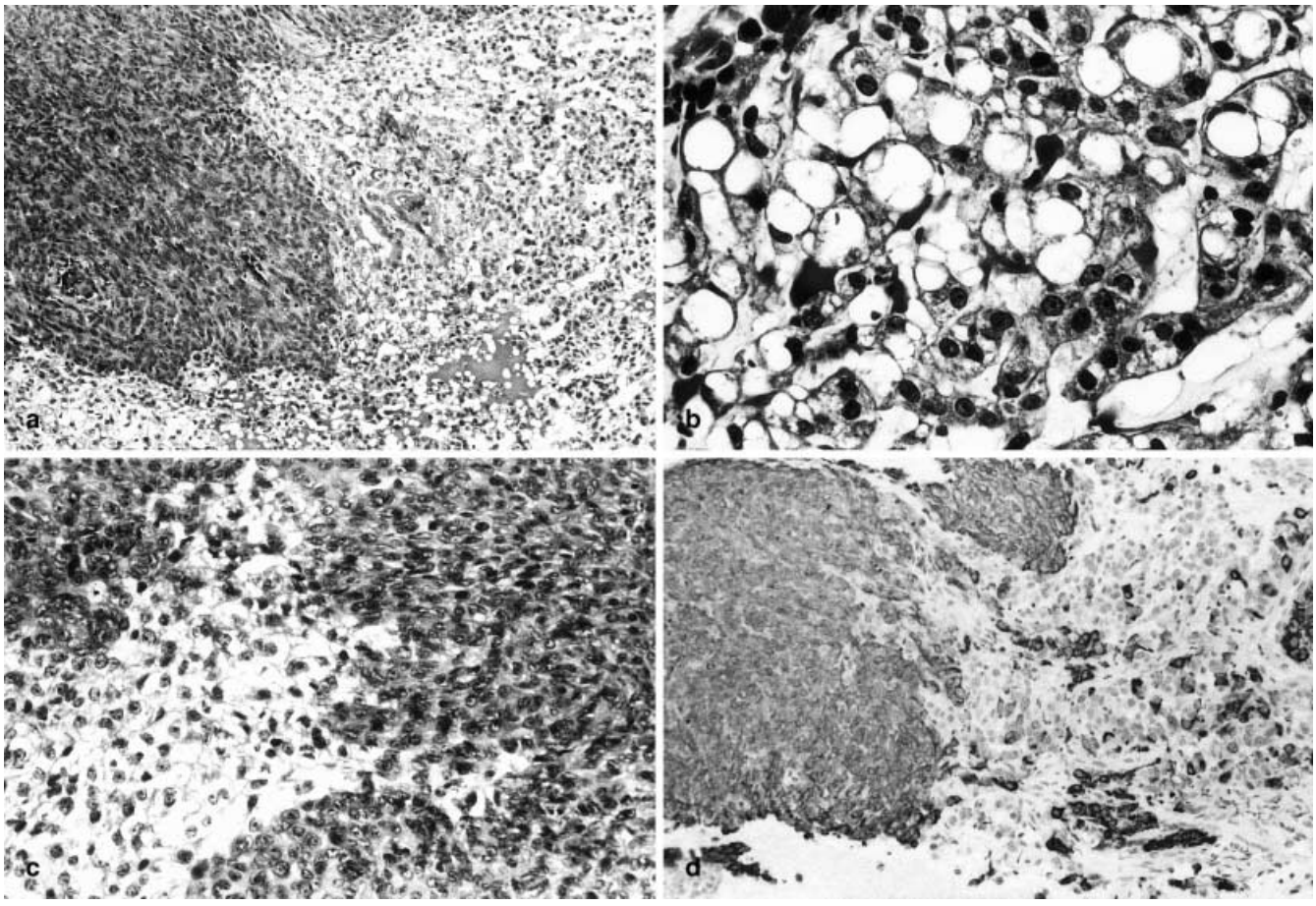

Fig. 2 **a** The tumor consisting of cellular and loose-looking components. **b** Physaliferous cells with oval nuclei and intracytoplasmic vacuolar changes. **c** Cellular portions consisting of highly atypical spindle cells arranged in short fascicles with gradual transition between cellular and loose-looking components **d** Cytokeratin (CAM5.2)-positive tumor cells in both cellular and loose-looking components

Discussion

Chordoma coexisting with a highly malignant sarcomatous component has been reported by several authors [2–14] (Table 1). In most of the reported cases, a sarcomatous component is composed of pleomorphic sarcoma resembling malignant fibrous histiocytoma [6, 8–10] or so-called spindle cell sarcoma similar to that of fibrosarcoma [5], and rarely is osteosarcoma reported [3, 14]. In some cases, a sarcomatous component was observed in a recurrent or initial tumor after radiation therapy [6, 8–11, 14], but has developed de novo in other cases [2–4, 13]. Most of the cases with a de novo sarcomatous component had poor prognosis, suggesting that these tumors were biologically much more aggressive than conventional chordoma. Different terminology has been used for such tumors, including chordoma with sarcomatous features (spindle cell metaplasia) [4], dedifferentiated chordoma [3], chordoma with a sarcomatous transformation [2, 13], or chordoma with a malignant spindle cell component [12, 14].

Dedifferentiated chordoma, first designated by Meis et al., is characterized by sharp demarcation of conventional chordoma and a high-grade sarcomatous component with minimal or no transitional features between the two components and by negative cytokeratin-immunoreaction in the sarcomatous component [3, 15]. In contrast, Miettinen et al. reported one case of chordoma containing a spindle cell sarcomatous component with a transitional feature from conventional chordoma, in which both components were immunoreactive to cytokeratin [2]. Further, Hruban et al. described four cases of chordomas with malignant spindle cell components, two of which (cases 2 and 3) showed a transitional feature between conventional chordoma and malignant spindle cell components positively immunoreactive to cytokeratin [12]. The current case is considered to be in the same category as the cases reported by Miettinen et al. referred to by the descriptive term “chordoma with a massive spindle-cell sarcomatous transformation” [2].

Dedifferentiation is a well-known phenomenon in some types of bone and soft tissue sarcomas, including

Table 1 Reported cases of chordoma with sarcomatous component in 1970–2000

Author	Age 1/2[a]	Sex	Radiation[b]	Location	Histology[c]	Transition[d]	Outcome[e]
Knechtges [4]	62/62	M	No	Sacrum	?	?	2 months
Kishikawa et al. [5]	53/53	M	Yes	Sacrum	DC	No	3 years
Mekek et al. [6]	52/67	M	Yes	Sacrum	DC	No	4 years
Volpe and Mazabraud [7]	9/9	M	No	Sphenoid	?	?	1 months
	36/36	M	?	C3–C4	?	?	16 months
Miettinen et al. [8]	49/56	F	Yes	L1–L2	DC	No	6 months
Halpern et al. [9]	68/76	M	Yes	Sacrum	?	?	10 months
Belza et al. [10]	57/63	F	Yes	C1–C5	SC	Yes	18 months
Meis et al. [3]	66/66	M	No	Sacrum	DC	No	6 months
	44/44	M	No	Sacrum	DC	No	5.5 months
	65/65	M	No	Sacrum	DC	No	76 months
Miettinen et al. [2]	26/26	F	No	Sacrum	SC	Yes	3 months
Hruban et al. [11,12]	71/71	M	Yes	L2–S1	DC	No	AWD 6 months
	62/62	M	No	Sacrum	DC	No	42 months
	70/72	M	Yes	Sacrum	SC	Yes	22 months
	57/57	M	Yes	Sacrum	SC	Yes	AWD 18 months
Tomlinson et al. [13]	55/57	M	Yes	Clivus	?	?	1 month
	56/56	M	No	Cranium	?	?	6 months
Fukuda et al. [14]	48/54	F	Yes	Sacrum	SC	Yes	3 years
	63/72?	M	Yes	Sacrum	SC	Yes	>2 years
Present case	74/74	F	No	Sacrum	SC	Yes	

[a] Age 1, age at presentation; age 2, age at development of a sarcomatous component
[b] Radiation therapy before development of a sarcomatous component
[c] DC, dedifferentiated chordoma; SC, sarcomatoid chordoma
[d] Transitional feature between a conventional chordoma area and a sarcomatous component
[e] Duration between recognition of a sarcomatous component and death; AWD, alive with disease

chondrosarcoma and liposarcoma. In such tumors, the term "dedifferentiation" has traditionally been used for a bimorphic neoplasm consisting of well or moderately differentiated low-grade sarcoma and high-grade malignant fibrous histiocytoma (MFH)-like or fibrosarcoma-like undifferentiated sarcoma, without a transitional feature between the two components [15]. Thus, according to the original definition, tumor cell differentiation identical to that in a low-grade sarcomatous area should not be observed in the dedifferentiated portions.

Sarcomatoid chordoma can be distinguished from dedifferentiated chordoma by the presence of a transitional feature between conventional chordoma and sarcomatoid components, and positive cytokeratin immunoreactivity in both components. A number of hypotheses have been proposed for the occurrence of the high-grade sarcomatoid component in chordoma, including spontaneous progression of a single tumor, collision of two distinct tumors, and radiation-induced malignant transformation [11, 14]. The current case may provide a model of spontaneous progression from conventional chordoma to high-grade sarcomatoid tumor.

The HBME-1 antibody is sometimes useful for distinguishing mesothelioma from adenocarcinoma on immunohistochemical trial, although the precise nature of its antigen is uncertain. Recently O'Hara et al. reported that positive immunoreaction to HBME-1 was observed in most chordomas [16]. When typical physaliferous cells, a characteristic feature of chordoma, are not frequently recognized in the specimen, as in the current case, HBME-1 may be useful to distinguish chordoma with or without a sarcomatoid component from epithelial tumors such as carcinoma metastatic to the sacral vertebrae.

In summary, we describe a case of sarcomatoid chordoma containing high-grade spindle cell component arising de novo in conventional chordoma. Recognition of a transitional feature between the two components and immunoreactivity to cytokeratin in both portions are crucial to distinguish such a tumor from dedifferentiated chordoma. Familiarity with this type of bone tumor should help to avoid confusion with dedifferentiated chordoma and other spindle cell sarcomas or carcinomas.

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