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Case report

Primary rhabdomyosarcoma of the iliac bone in an adult: A case mimicking fibrosarcoma

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Abstract. Primary rhabdomyosarcoma of bone is exceedingly rare. We present a case of rhabdomyosarcoma of the iliac bone in a 32-year-old male. Histologically, the tumour consisted mainly of a uniform proliferation of elongated spindle cells arranged in a herring bone pattern, simulating fibrosarcoma. Focally there was a conventional embryonal pattern with scattered rhabdomyoblasts possessing an eosinophilic cytoplasm. Immunohistochemical studies disclosed expression of muscle markers such as desmin and muscle-specific actin, in both the embryonal and spindle-cell areas and myoglobin only in the embryonal areas. Such histological features are unusual for classical embryonal rhabdomyosarcoma. The anatomical site and age of the patient are also atypical.

Key words: Rhabdomyosarcoma – Bone tumour – Immunohistochemistry

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Introduction

Rhabdomyosarcoma arises principally in the soft tissues of the head and neck, the urogenital tract, and the retroperitoneum (Enzinger and Weiss 1988). Its occurrence in adults is rare (Lloyd et al. 1983; Seidal et al. 1989). There have been few reports of its occurrence as a primary neoplasm of bone (Pasquel et al. 1976; Hsueh et al. 1986).

Histologically, the majority of the cells in the tumour reported here were composed of spindle cells arranged in either a fascicles or a herring bone pattern, associated with embryonal areas. No unequivocal reports of rhabdomyosarcoma showing such unusual histological features in the bone are known to us. We report herein a case of an elderly patient with a rhabdomyosarcoma in the iliac bone and discuss its unusual histological features.

Materials and methods

Histological sections from paraffin-embedded tumour tissue, fixed in formalin, were stained with haematoxylin and eosin, Masson's trichrome, periodic acid-Schiff both before and after digestion with diastase, phosphotungstic acid-haematoxylin, and silver impregna-

Table 1. Primary antibodies used for immunohistochemistry

Antigen	Supplier	Dilution	Clonality
Vimentin	Enzo, New York, N.Y., USA	1:30	M
Desmin	Dakopatts, Glostrup, Denmark	1:100	M
Desmin	Immunotech, Marseille, France	Undiluted	M
HHF-35[a]	Enzo	1:16000	M
Myoglobin	Dakopatts	1:300	P
Myosin[c]	Sigma, St. Louis, Mo., USA	1:200	M
Alpha-SMA[b]	Sigma	1:5000	M
S-100[c]	Dakopatts	1:400	P

M, Monoclonal; P, polyclonal
[a] Muscle-specific actin
[b] Alpha-smooth muscle actin
[c] Pretreated with trypsin

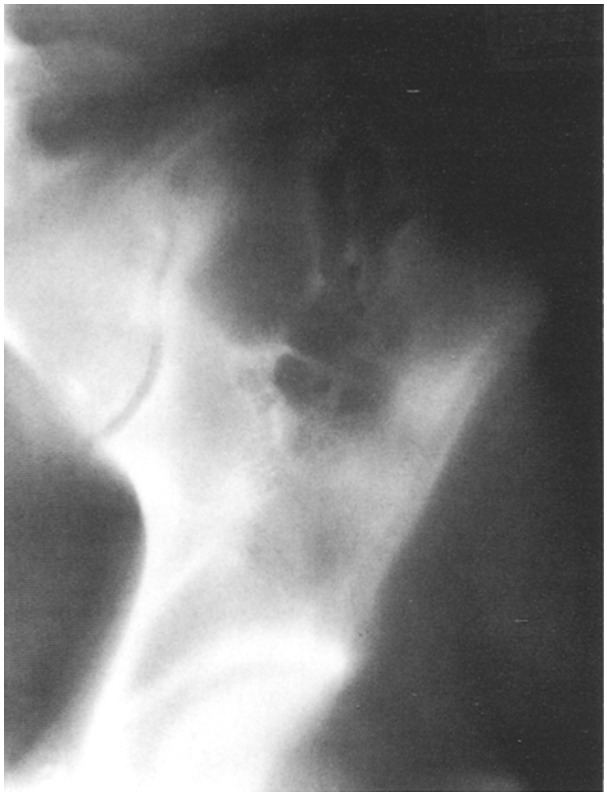

Fig. 1. Plain tomography reveals a multilocular osteolytic lesion without marginal sclerosis from the left iliac wing to the acetabulum

tion for reticulin. Additional sections were stained using the peroxidase-avidin-biotin method (reagents from Vector, Burlingame, Calif., USA) with diaminobenzidine as chromogen and the primary antibodies listed in Table 1. Tissue from normal skeletal muscle and smooth muscle from a vessel wall were used as a positive

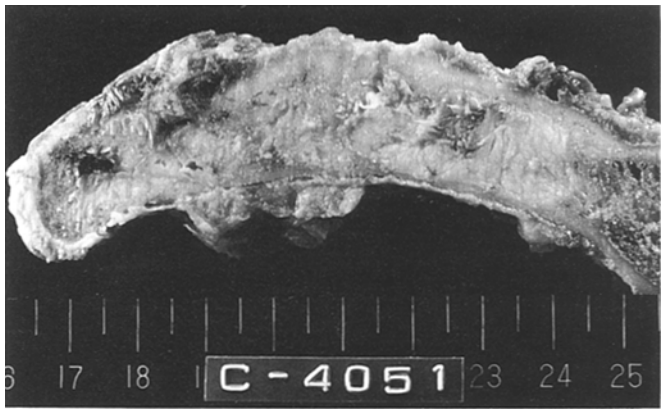

Fig. 2. Grossly, a whitish-yellow mass fills the medullary cavity on its cut surface

control for muscle markers and substitution of the primary antibody by phosphate-buffered saline was employed as a negative control.

Case report

A 32-year-old man presented with a 2-week history of left gluteal pain and coxalgia. Palpation of the left gluteal region showed marked tenderness. Laboratory studies showed no remarkable changes. A roentgenographic examination revealed a multilocular osteolytic lesion in the left iliac bone (Fig. 1). The margins of the lesion were irregular, with no evidence of any reactive bone formation. Computed tomography showed an intramedullary osteolytic lesion with cortical destruction and a tiny extending soft tissue mass. Angiography revealed slightly increased neovascularity. A left hemipelvectomy was performed. Postoperatively, the patient received one course of adjuvant chemotherapy (endoxan, vincristine, doxorubicin and dacarbazine). For 4 years following surgery, the patient has remained tumour-free with no signs of either local recurrence or metastasis.

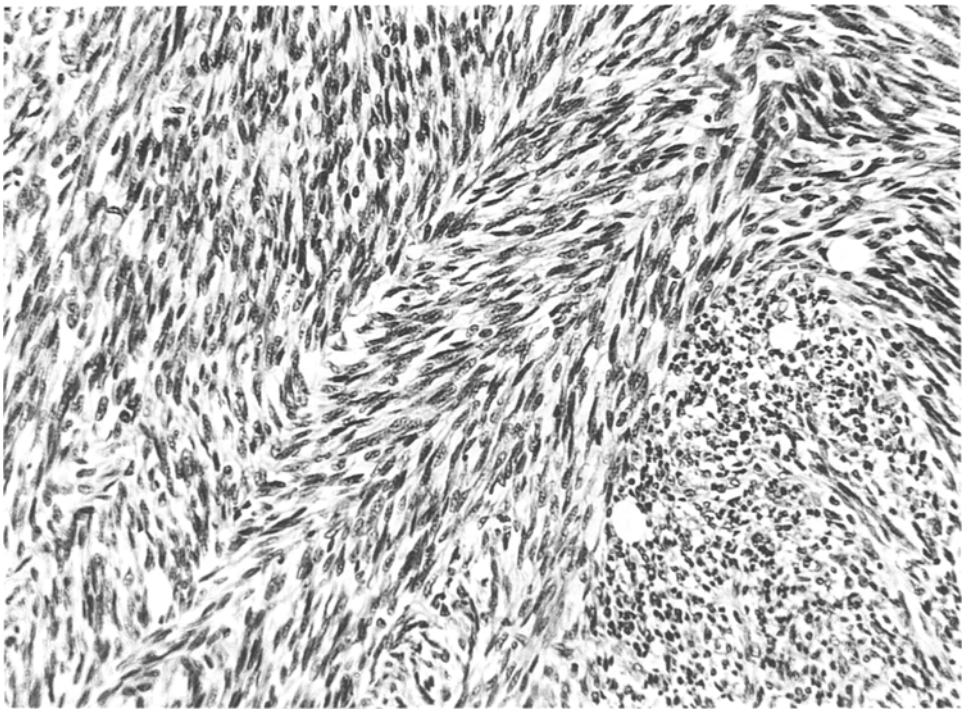

Fig. 3. The majority of the tumour is composed of long fascicles of spindle cells arranged in a herring bone pattern simulating fibrosarcoma. H & E, ×260

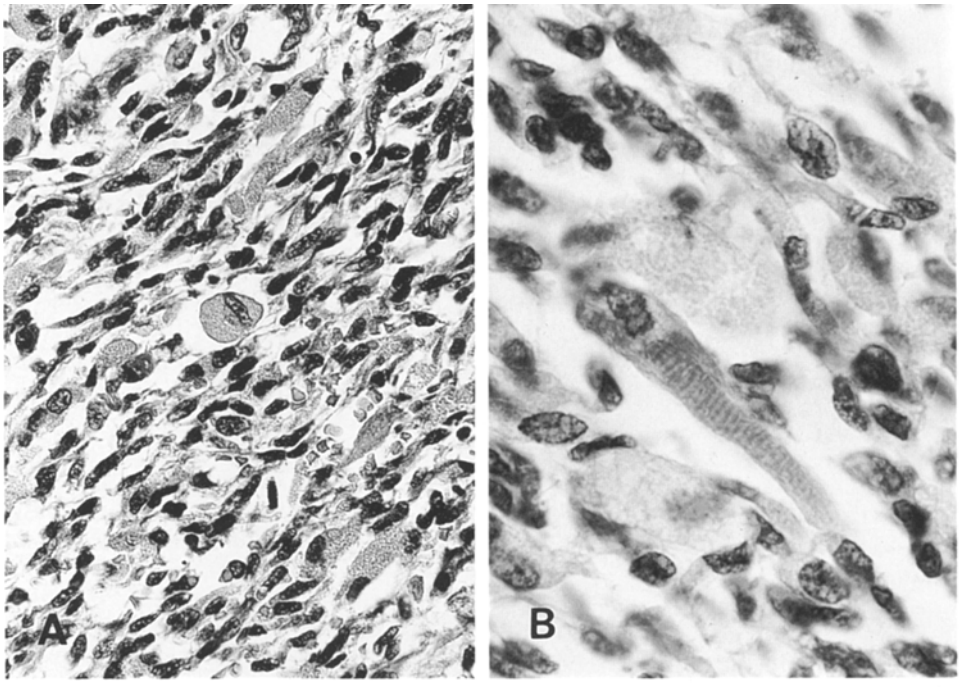

Fig. 4. A Embryonal areas made up of a mixture of spindle, stellate, and round cells with an eosinophilic cytoplasm. H & E, ×460. **B** Some of the ribbon-shaped cells in embryonal areas show cytoplasmic cross-striations. H & E, ×1000

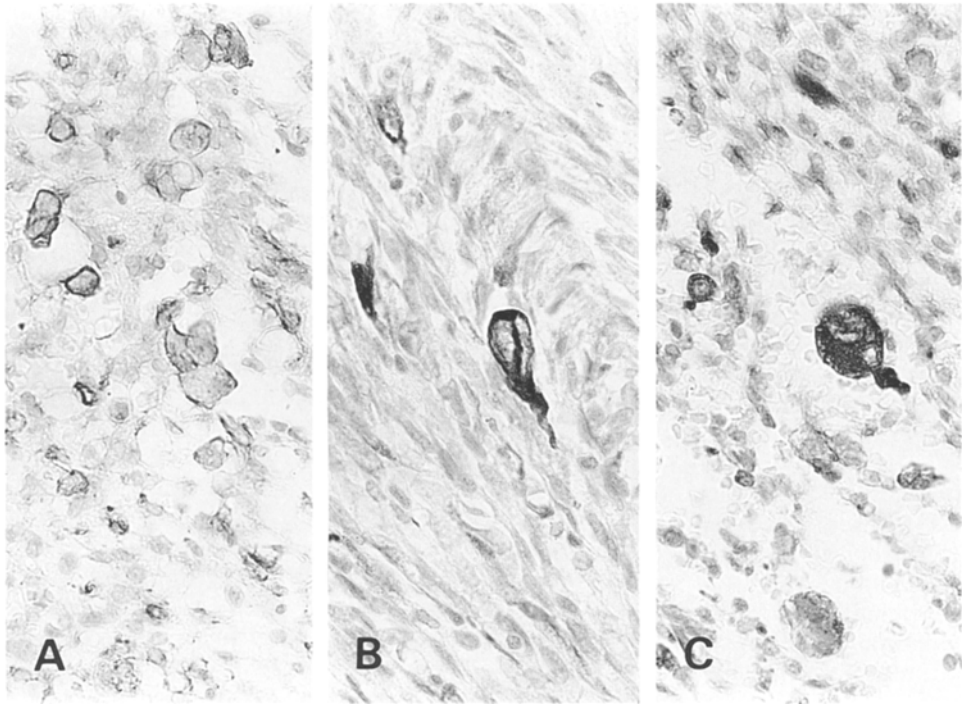

Fig. 5. A Some of the large round cells are positive for muscle-specific actin (HHF-35). **B** The strap-shaped and short fusiform cells are strongly immunoreactivity with desmin. **C** Some of the strap-shaped cells and small cells possess positive reaction products for myoglobin. ABC method, ×500

Pathology

On the cut surface of the left iliac bone, a whitish-yellow mass occupied the medullary cavity (Fig. 2). There was focal destruction of the cortical bone with limited invasion of the skeletal muscle and soft tissues on the external and internal aspect of the iliac bone.

Microscopically, the tumour was composed mainly of elongated fusiform cells. These cells were organized into long fascicles mimicking a fibrosarcoma (Fig. 3). The tumour cells had eosinophilic and fibrillar cytoplasm with distinct cellular borders. Classical embryonal rhabdomyosarcoma-like areas made up of a mixture of spindle, stellate, and round cells were also occasionally randomly observed (Fig. 4A). Scattered round cells or elongated cells with conspicuously eosinophilic cytoplasm were easily found. Some of the elongated cells showed obvious cross-striations (Fig. 4B). Although reactive bone formation was partially seen, no malignant osteoid was found.

Vimentin was strongly expressed in all tumour cells regardless of the cytological features. Desmin and muscle-specific actin (HHF-35) reactivities were recognized in both the spindle-cell areas and embryonal areas. However, such expression was more marked in the embryonal than in spindle-cell areas (Fig. 5A, B). Myoglobin expression was only observed in the embryonal areas and was more

Table 2. Primary rhabdomyosarcoma of bone: summary of the clinical findings as reported in the literature

Case	Authors	Age/sex	Symptom	Duration	Site	Prognosis
1	Pasquel et al. 1976	13/female	Pain	3 months	Femoral shaft	Died after 8 months with lung metastasis
2	Hsueh et al. 1986	11/male	Painful mass	2 months	Distal femur	Unknown
3	Oda et al. 1992[a]	32/male	Pain	2 weeks	Ilium	Alive and well after 4 years

[a] Current case

often localized in the embryonal strap cells and cytoplasm-rich rhabdomyoblasts than in the oval or round myoblasts (Fig. 5C). No tumour cells had any positive reaction products for myosin, S-100 protein or alpha-smooth muscle actin.

Discussion

The previously reported cases of rhabdomyosarcoma of the bone consisted mostly of primary soft tissue tumours with secondary involvement of the bone (Calinog et al. 1971; El-Gothamy et al. 1973; Naufal 1973; Huvos 1991) or were considered to be metastatic lesions, even if in some of these areas the primary site had not been found (Case records of Massachusetts General Hospital 1972; Henderson et al. 1976; McLean and Murray 1984). For instance, Calinog et al. (1971) reported two cases of rhabdomyosarcoma of the sternum with a large soft tissue tumour mass and El-Gothamy et al. (1973) reported a case in the temporal bone, and believed it to be secondary involvement by a primary soft tissue tumour. Another case was recorded at Massachusetts General Hospital of embryonal rhabdomyosarcoma with multiple boney involvement (1972), while McLean and Murray (1984) reported a case of metastatic alveolar rhabdomyosarcoma in the femur. In our case, radiography and computed tomography revealed evidence of a primary bone tumour rather than any secondary involvement by a soft tissue primary. In addition, the resected specimen revealed a large intramedullary tumour mass with a smaller soft tissue tumour extension. We therefore considered this case to be an example of primary rhabdomyosarcoma of bone.

A misdiagnosis of both fibrosarcoma and malignant fibrous histiocytoma of bone could made with this case. However, these tumours never show cytoplasmic cross-striations or immunoreactivity for myoglobin.

Malignant mesenchymoma was originally described by Stout (1948) as a malignant neoplasm with two or more heterogeneous, differentiated mesenchymal elements other than fibrosarcomatous elements. In our case, there was no other mesenchymal component such as osteosarcoma, chondrosarcoma or liposarcoma.

The differential diagnosis included fibroblastic osteosarcoma and dedifferentiated chondrosarcoma, whose fibrosarcomatous component may be devoid of any specific intercellular substances such as osteoid or chondroid matrix over large areas.

Furthermore, dedifferentiated chondrosarcoma may show divergent differentiation toward skeletal muscle (Astorino and Tesluk 1985; Têtu et al. 1986; Niezabitowski et al. 1987). Foci of tumour osteoid and/or cartilage were not seen in our case.

Microscopically, a malignant Triton tumour could be another possible differential diagnosis. However, this patient displayed no stigmata of von Recklinghausens' disease and immunohistochemically, no tumour cells showed positive reaction for S-100 protein.

Rare cases of primary leiomyosarcoma of the bone have been reported previously (Sanerkin 1979; Angervall et al. 1980; von Hochstter et al. 1984). In our case, smooth muscle differentiation (blunt-ended nuclei and paranuclear vacuoles) was not recognized on light microscopy and immunohistochemical studies did not demonstrate a positive reaction for alpha-smooth muscle actin: specific markers for smooth muscle tumours (Fletcher 1992).

To our knowledge, there have only been two reported instances in which the prerequisites for a diagnosis of primary rhabdomyosarcoma of the bone have been satisfied: one in the femoral shaft of a 13-year-old girl (Pasquel et al. 1976), and the other in the distal femur of an 11-year-old boy (Hsueh et al. 1986; Table 2). Histologically, both cases revealed classical embryonal rhabdomyosarcoma. The current patient was older and the tumour was mainly made up of elongated fusiform cells arranged in fascicles associated with embryonal areas, which would be considered unusual for classical embryonal rhabdomyosarcoma.

The histological features of the current case closely resemble those of the recently described spindle cell rhabdomyosarcoma (Cavazzana et al. 1992), a rare variant of the embryonal form. Cavazzana et al. (1992) documented 21 cases of this rare variant, collected from 471 cases of rhabdomyosarcoma. They discussed a favourable prognosis of this variant when comparing it with classical forms of embryonal rhabdomyosarcoma. In fact, our patient is alive and well 4 years after undergoing both surgery and one course of adjuvant chemotherapy. Although the histological findings of our case are similar to spindle cell rhabdomyosarcoma, the immunohistochemical features differed somewhat.

In the series of Cavazzana et al. (1992), all cases examined had immunoreactivity for myoglobin not only in the embryonal areas, but also in the spindle-cell areas. Our case did not show immunoreactivity for myoglobin in the spindle-cell areas, and we can still not explain these immunohistochemical differences between our case and the series of Cavazzana et al. (1992) completely.

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