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Well-differentiated Osteosarcoma of the Rib

Well-differentiated osteosarcoma is extremely rare, there is no sex predominance and the mean age is in the third decade. The tumor has a strong predilection for the long bones of the extremities, especially the metaphysis but may also occur in the diaphysis. Radiologically, the lesion shows no distinctive features, often simulating fibrous dyplasia or desmoplastic fibroma. We report a case of well-differentiated osteosarcoma involving the rib of a 45-year-old female. This is a peculiar case of well differentiated osteosarcoma involving an unusual site and older patient. We also discuss histological differential diagnosis as well as clinical features of this rare disease.

Key Words : *Osteosarcoma; Rib*

Yong-Koo Park, Shin Eun Choi,* Kyung Nam Ryu†

Departments of Pathology, College of Medicine, Kyung Hee University and Kangnam General Hospital*; Department of Diagnostic Radiology,† College of Medicine, Kyung Hee University, Seoul, Korea

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Address for correspondence
Yong-Koo Park, M.D.
Department of Pathology, Kyung Hee University Hospital, 1 Hoeki-dong, Dongdaemoon-gu, Seoul 130-702, Korea
Tel : +82.2-958-8742, Fax : +82.2-957-0489
E-mail : damia@chollian.net

INTRODUCTION

Well differentiated osteosarcoma of the bone is rare, accounting for approximately 1.2% of all osteosarcomas (1) and was first described by Unni et al. in 1977 (2). Presenting symptoms and signs are nonspecific and typically consist of pain and swelling. Often symptoms have been present for more than one year, a rare occurrence for conventional, high grade osteosarcoma (3, 4). There is no obvious sex predilection, although one study suggested a female predominance (3). Patients are distinctly older, on average, than those with conventional osteosarcoma. It often peaks out in the third decade of life, but affected individuals are often in their fourth, fifth, sixth, or even seventh decades (4). There is a predilection for the metaphyseal regions of the long bones of the lower extremity (4). In this report, we had a well-differentiated osteosarcoma involving the rib of a 45-year-old female. We report this unique location, radiological and histologic findings for the well-differentiated osteosarcoma.

CASE HISTORY

This 45-year-old female had suffered from a palpable mass on her left chest wall for several months. The antero-posterior (Fig. 1A) and oblique (Fig. 1B) chest x-ray showed a bony mass, measuring about 6×3 cm in size in the left seventh rib. The lesion revealed a sharply defined lobulated margin. Marginal irregularity was noted at the lower border of the lesion, however, there was no definite discontinuity of the periosteal reaction. Segment of the rib lesion including normal looking bone were resected. Grossly, the rib showed bulging cortical bone and marrow space filled with gray white sclerotic bony tissue. Histologically, there were broad trabeculae of heavily mineralized bone surrounded by spindle cell proliferation (Fig. 2). The tumor was composed of newly formed irregular bony trabeculae and hypocellular spindle cells in fibrous stroma. The spindle cells showed slightly hyperchromatic spindle shaped nuclei and indistinct cytoplasmic borders (Fig. 3). Often the spindle cells revealed in an interlacing pattern and infiltration with permeation of the bone marrow. There were abundant cartilage formations (Fig. 4).

DISCUSSION

Most osteosarcomas are highly malignant and hence, cytologically obvious. However, a small number of osteosarcomas are extremely well-differentiated and may be mistaken for a benign condition, such as fibrous dysplasia (2). Low grade well-differentiated osteosarcoma is rare; there are only 20 cases of the 1,649 osteosarcomas in the Mayo Clinic series. This tumor, although malignant,

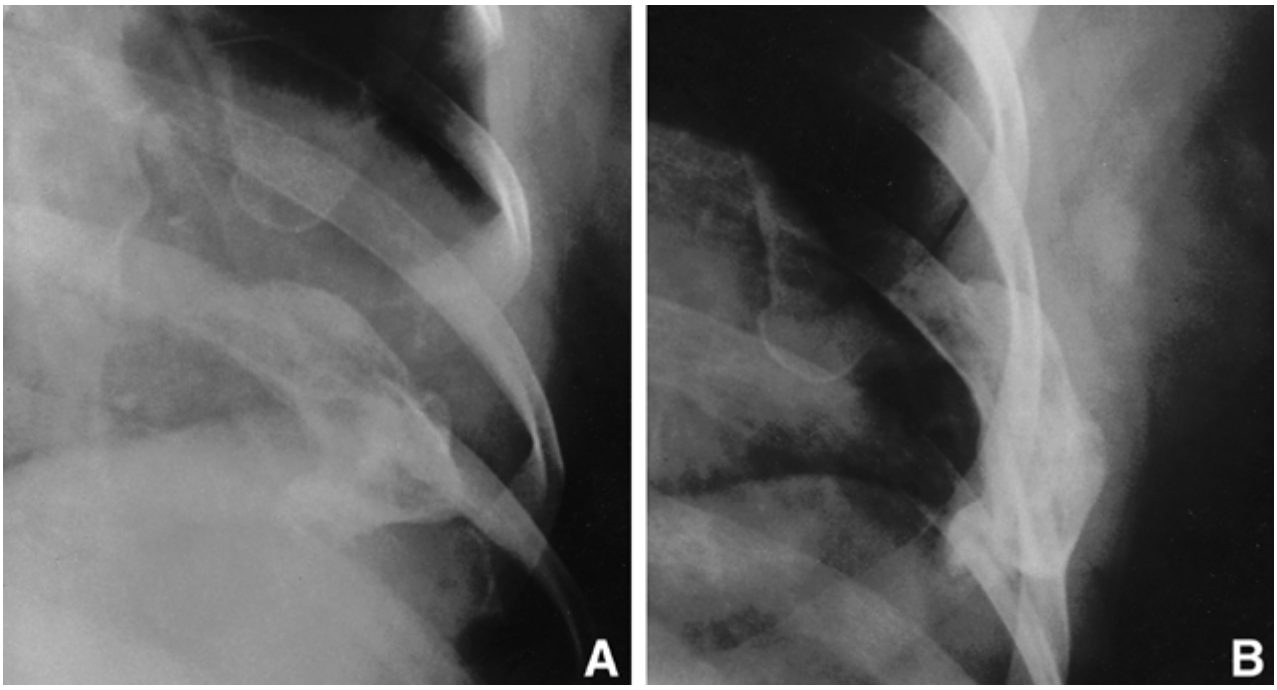

Fig. 1. The antero-posterior (A) and oblique (B) radiographs show a sharply defined lobulated bony mass measuring 6×3 cm in size. Marginal irregularity is noted at the lower borer of the lesion.

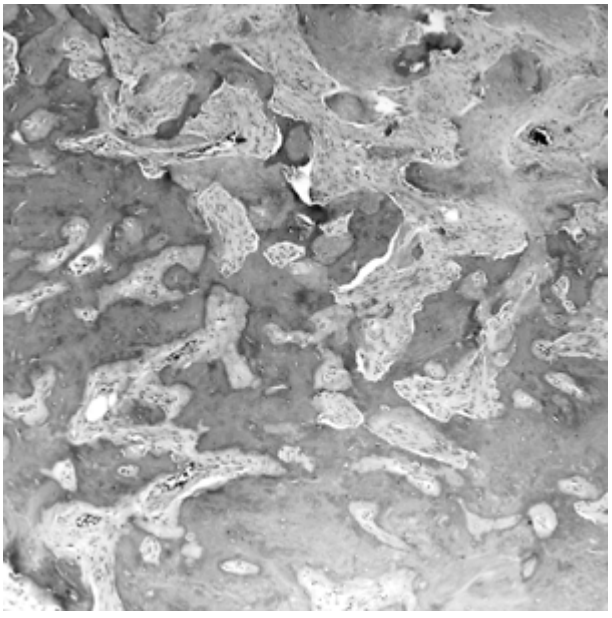

Fig. 2. There are abundant new bone formations with spindle cells between the newly formed trabecular bone (H&E, ×40).

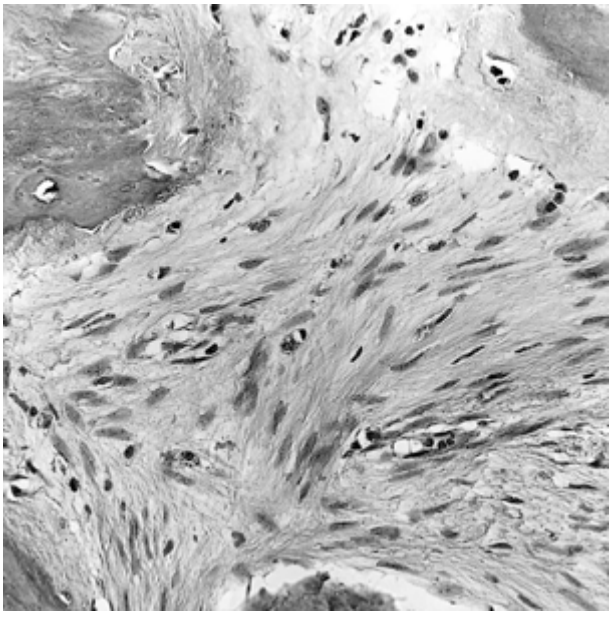

Fig. 3. The spindle cells between the bony trabeculae show minimal cytologic atypia (H&E, ×200).

is indolent and must be distinguished from conventional osteosarcomas because of its much better prognosis, and from parosteal or periosteal osteosarcomas because of its location. Patients tend to be older than those with conventional osteosarcoma. According to Kurt et al. (4), peak incidence is in the third decade of life. In this case, the patient was 45-year-old. There is no sex predilection. The distal femur and upper tibia are the most common sites of involvement (1). However, two cases have been reported in the occipital bone and mandible (4, 5), and intracranial sphenoid bone (6). Usually the roentgenographic appearance is not diagnostic. In this case, radiologi-

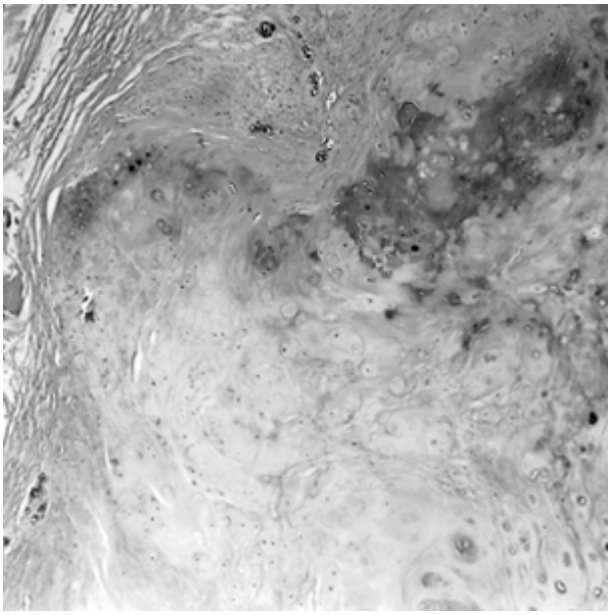

Fig. 4. There are abundant cartilage formations (H&E, ×40).

cally, fibrous dysplasia is listed for the differential diagnosis.

Histologically, most of these tumors bear a striking resemblance to parosteal osteosarcoma and differ just as strikingly from conventional high grade osteosarcoma. Well-differentiated osteosarcoma is essentially a spindle cell tumor with irregular bone production, low cellularity, and few mitotic figures. This well-differentiated osteosarcoma may be confused with fibrous dysplasia, nonossifying fibroma, and osteosarcoma, as well as other entities. It is frequently diagnosed erroneously as fibrous dysplasia; it is not easy to make the distinction on histologic findings alone. Radiologically, fibrous dysplasia has a typical "ground glass" appearance with a sharply demarcated margin, often associated with a rim of reactive sclerosis. Trabeculations are commonly seen radiographically in well-differentiated osteosarcoma and are less common in fibrous dysplasia. In fibrous dysplasia, histology is often simulated by the well-differentiated osteosaroma, although the new bone observed in well-differentiated osteosarcoma is more disorderly than fibrous dysplasia (7). The striking difference is that the fibroblastic nuclei in well-differentiated osteosarcoma are more hyperchromatic than those in fibrous dysplasia. The stromal cells are arranged generally parallel to the trabeculae in fibrous dysplasia (6). Another diagnosis of a benign neoplasm originally rendered histologically was metaphyseal fibrous defect (nonossifying fibroma). In this, the cells are arranged loosely and in a cartwheel fashion, and they lack cellular atypia (7).

In summary, a case of well-differentiated osteosarcoma of the rib was reported. The pathological differentiation of this rare entity from fibrous dysplasia, the radiological appearance and the differential diagnosis were discussed.

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