The Korean Journal of Pathology
2006; 40: 314-7

Osteosarcoma of the Skull Resembling Desmoplastic Fibroma
- A Case Report -

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Received : August 10, 2005
Accepted : July 20, 2006

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* This case was discussed in the 30th Annual Autumn Meeting of the Korean Society of Pathologists.

Primary osteosarcoma of the skull is a rare finding. We report here on a pathologically proven case of osteosarcoma that presented as a painless mass in the frontal bone of a 7-year-old boy. This unusual form of osteosarcoma had features of desmoplastic fibroma in a large portion of the tumor. We also include a review of the medical literature related to osteosarcoma.

Key Words : Osteosarcoma; Skull

Osteosarcoma is a malignant neoplasm of the bone that produces osteoid or a bony matrix, and it usually affects the metaphysis of long bones. Osteosarcoma rarely affects the skull; such cases constitute fewer than two percent of all osteosarcomas.[1,2] Most of the previous such articles have reported on only a single case. There are only three large series reporting on osteosarcomas of the skull, except in the jaw bones.[1-3] Because of the rarity of this tumor and the diagnostic difficulties, we report here on a case of osteosarcoma of the skull.

CASE REPORT

A seven-year-old boy presented to our institution because of a painless frontal mass that had slowly enlarged in the 3 years prior to admission. On admission, he showed no neurological abnormalities. There was no history of trauma, a family history of cancer, any preexisting bony lesion or a history of irradiation to the head. On physical examination, there was a protruding, non-movable firm mass in the frontal bone without tenderness. The blood chemistry parameters were within normal limits, including alkaline phosphatase. A plain radiograph of the skull showed a large, solitary expansile osteolytic lesion with a reticulated pattern at the diploic space of the frontal bone. The outer table of the frontal bone showed remodeling with inner decreased radiopacity of the osteolytic lesion (Fig. 1A). Magnetic resonance imaging of the skull revealed an expansile lesion with iso-signal intensity on the T1- and T2-weighted images at the outer table of the frontal bone. The diploic space and inner table showed low signal intensity with a thickened contour (Fig. 1B). After Gd-DTPA enhancement, the mass at the outer frontal bone was well enhanced and it had an inner nonenhancing portion. The thickened diploic space showed spotted and honey-comb enhancement. The radiological diagnosis was hemangioma of the frontal bone. The patient subsequently underwent simple excision of the mass without a preoperative biopsy. The specimens were fragments of bony tissue approximately $5 \times 4 \times 3.5$ cm in size. Microscopically, the tumor consisted mostly of small spindle cells sep-

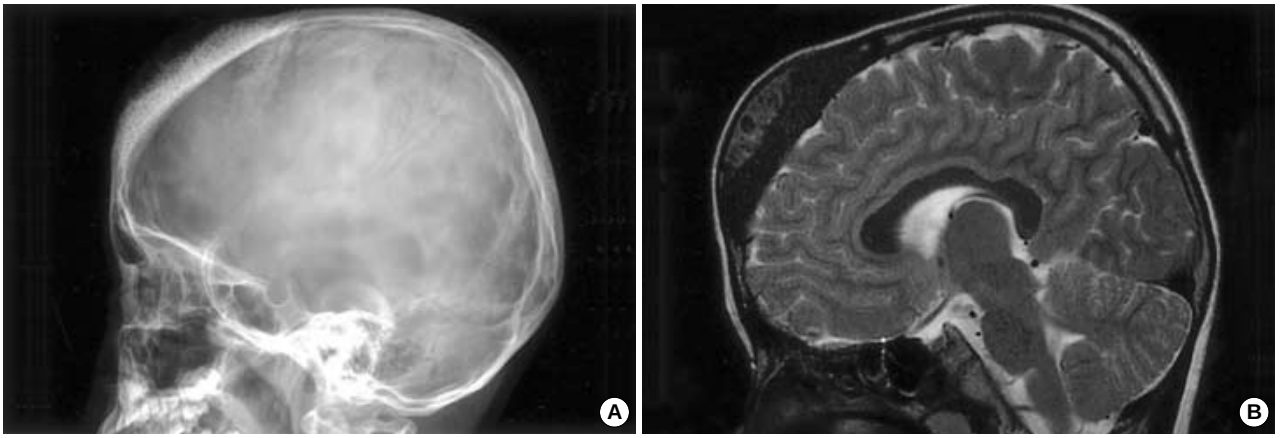

Fig. 1. (A) Skull lateral view of plain radiograph shows increased radiopacity and honey combed osteolytic lesion of the frontal bone. (B) T2-wighted sagittal image shows an expansile lesion, with iso-signal intensity at the outer table of frontal bone. Diploic space and inner table shows low signal intensity with thickened contour.

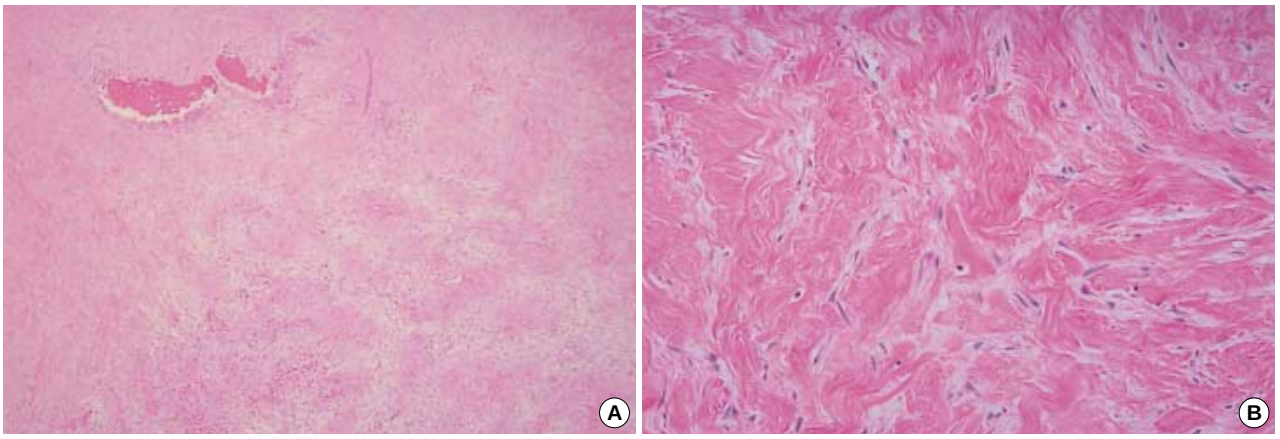

Fig. 2. (A) Microscopically, the tumor is composed of spindle cells separated by abundant collagen fibers. Entrapped remnants of bone with evidence of osteoclastic resorption are noted. (B) The spindle cells have a plump and elongated nucleus. Mitotic figures are absent. Collagen fibers are wavy and densely distributed and form broad bands.

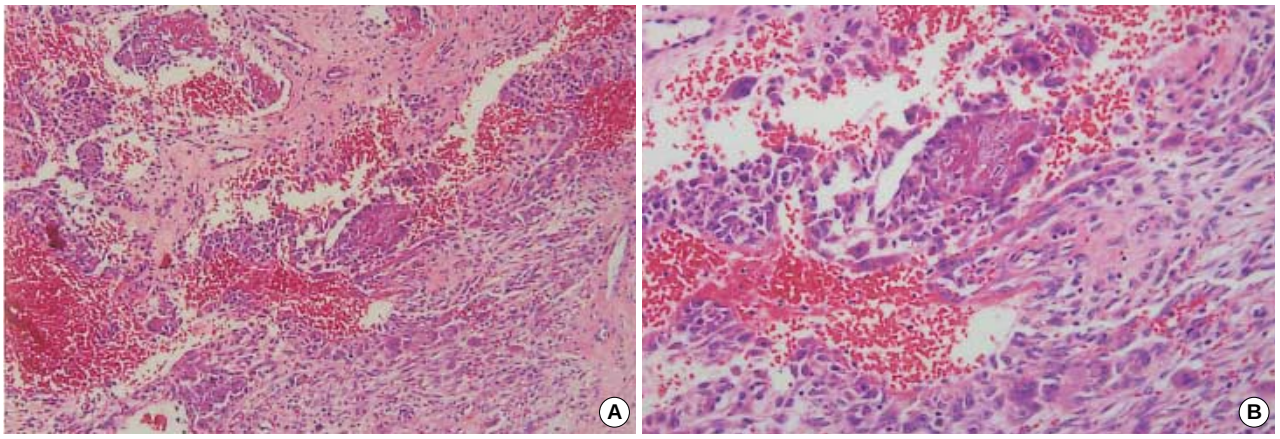

Fig. 3. (A) Area of osteosarcoma shows neoplastic osteoblasts, and osteoid production. (B) Neoplastic osteoblasts have nuclear pleomorphism, prominent nucleoli and mitotic figures.

arated by abundant collagen fibers. Entrapped remnants of bone with evidence of osteoclastic resorption were noted (Fig. 2A). The spindle cells had a plump and elongated nucleus. Mitotic figures were absent. Collagen fibers were wavy and densely distributed and they formed broad bands (Fig. 2B). These areas resembled desmoplastic fibroma of the bone. Areas simulating the appearance of low grade intraosseous osteosarcoma were also present in continuity with the desmoplastic fibroma-like lesion. These areas showed spindle cell proliferation embedded in a fibrous stroma and foci of osteoid production. The osteoid seemed to arise directly from spindle cells. However, there were foci of high grade osteosarcoma that consisted of anaplastic osteoblasts and unequivocal osteoid production (Fig. 3A). The osteoblasts had nuclear pleomorphism, prominent nucleoli and mitotic figures (Fig. 3B). The pathological diagnosis was high grade osteoblastic osteosarcoma. The patient received chemotherapy. At the 12-month follow-up, the patient was well, and he did not experience a recurrence.

DISCUSSION

Osteosarcoma rarely affects the skull, preferring instead the appendicular skeleton and especially the distal end of the femur. Since the first case of primary osteosarcoma of the skull was reported by Garland in 1945, there have been only 3 large series reporting on osteosarcoma of the skull, except in the jaw bones.[1-3] There are many corresponding studies that reported the rarity of *de novo* osteosarcoma of the skull. For instance, Nora *et al*. reported that 21 of 1,000 osteosarcoma cases had tumor in the skull, and only 14 out of 21 cases (1.4%) were *de novo* tumor; Huvos *et al*. reported that only 10 out of 1,200 osteosarcoma cases (0.8%) over a 60 year period were *de novo* osteosarcoma of the skull; and Salvati *et al*. reported 12 out of 19 skull osteosarcoma cases as *de novo* for a 36 year period.[1-3] Shinoda *et al*. in 1993 provided a review of 99 cases of primary osteosarcomas of the skull that were reported in the literature from 1945 to 1992.[4] In these reports, osteosarcomas of the skull usually occurred in the third and fourth decades, unlike those of the extremities. The gender predominance was variable. The primary sites of the tumor were more frequently in the calvaria than in the skull base. Of the 97 cases that mentioned the primary sites of the tumor in the literature, 75 cases were located at the cranial vault (26 in the temporal bones, 26 in the parietal bones, 13 in the frontal bones and 10 in the occipital bones) and 22 cases were located at the skull base, including the orbit.[2,4] The clinical symptoms and signs were site dependent, but a painless or tender lump in the calvaria was the most frequent symptom. The other presented symptoms were neurologic symptoms, including cranial nerve deficits. The duration of symptoms varied considerably from 15 days to 5 years. The mean duration of symptoms was less than a year. The predominant histologic type was ostoblastic, and the other types were the fibroblastic, telangiectactic or mixed types. Local recurrence of the tumor was a major cause of death, whereas distant metastasis was infrequent.[4,5] The prognosis depended mainly on the degree of intracranial involvement at the time of the diagnosis rather than the mode of therapy.[4] However, the introduction of chemotherapy has been found to improve the survival rate of patients with osteosarcoma of the head and neck.[3,5]

This case we report on showed foci of obvious high grade osteosarcoma with areas resembling desmoplastic fibroma and this simulated the appearance of low grade intraosseous osteosarcoma. Desmoplastic fibromas may infiltrate the surrounding trabeculae of bone at their periphery, and this resembles features of low grade osteosarcoma. Some of low grade osteosarcoma that have scanty osteoid and a fibroblastic stroma may resemble desmoplastic fibroma. However desmoplastic fibromas lack osteoid or bony production, and low grade osteosarcomas usually contain minimal cytological atypia.[6,7]

Transformation of desmoplastic fibroma to sarcoma is extremely rare even after a long course of disease. In the literature, there are two reports of a high grade osteosarcoma developing in a desmoplastic fibroma.[8,9] These two cases had been treated 11 to 16 years earlier for a desmoplastic fibroma at the same areas. Because there was a long disease-free interval and lung metastasis was not seen, the authors considered these lesions as desmoplastic fibroma that transformed to osteosarcoma. Unlike these cases, our case showed areas of high grade osteosarcoma with areas resembling desmoplastic fibroma, and the final diagnosis was primary high grade osteosarcoma arising in the frontal bone.

This case was presented at the 30th Annual Pathology Meeting in 2005. At that time, the presenter of the report referred to this case as low grade osteosarcoma. Low grade osteosarcoma is more common than high grade osteosarcoma; however, low grade osteosarcoma of the skull is rarer than high grade osteosarcoma.[6,7] As indicated in Fig. 3, we noticed obvious high grade lesion in the resected specimen. After we diagnosed it as high grade osteoblastic osteosarcoma, we obtained a second opinion from Dr. Unni KK (Mayo clinic, Rochester, MN) and he also considered this case as high grade osteoblastic osteosarcoma.

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