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High Grade Surface Osteosarcoma
- Case Report -

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High grade surface osteosarcoma is the most rare subtype of osteosarcoma arising on the surface of bone, accounting for less than 1% of the total number of osteosarcomas. Only a few case reports and studies have been reported in the world. In Korea, only one case out of 127 osteosarcomas has been described up to now, but there was no information about the patient, clinicopathologic features and treatment. We experienced a case of high grade surface osteosarcoma in the subtrochanteric area of a 66-year-old female and treated her with neoadjuvant chemotheraphy, wide resection and limb salvage operation with tumor prosthesis and adjuvant chemotheraphy. This tumor is identical to conventional high grade intramedullary osteosarcoma in histology, treatment and prognosis. So, this tumor should be differentiated from other surface osteosarcomas such as parosteal osteosarcoma and periosteal osteosarcoma.

Key words : High Grade Surface Osteosarcoma, Subtrochanteric area

Osteosarcomas arising on the surface of the bone have been classified into three subgroups: parosteal osteosarcoma, periosteal osteosarcoma and high grade surface osteosarcoma. High grade surface osteosarcoma is the most rare. According to Mayo Clinic records from 1926 to 1996, high grade surface osteosarcoma accounted for 8.9% of surface osteosarcomas and less than 1% of total osteosarcomas[6]. Only a few case reports and studies were reported in the world[1,2,4-9]. In Korea, one case of high grade surface osteosarcoma in 127 osteosarcomas has been described up to now, but there was no information about the patient, clinicopathologic features and treatment[3].

Because of its poorer prognosis, more attention is needed for its diagnosis and treatment.

CASE REPORT

A 66-year-old female with a 4 months history of right hip and thigh pain was pre-

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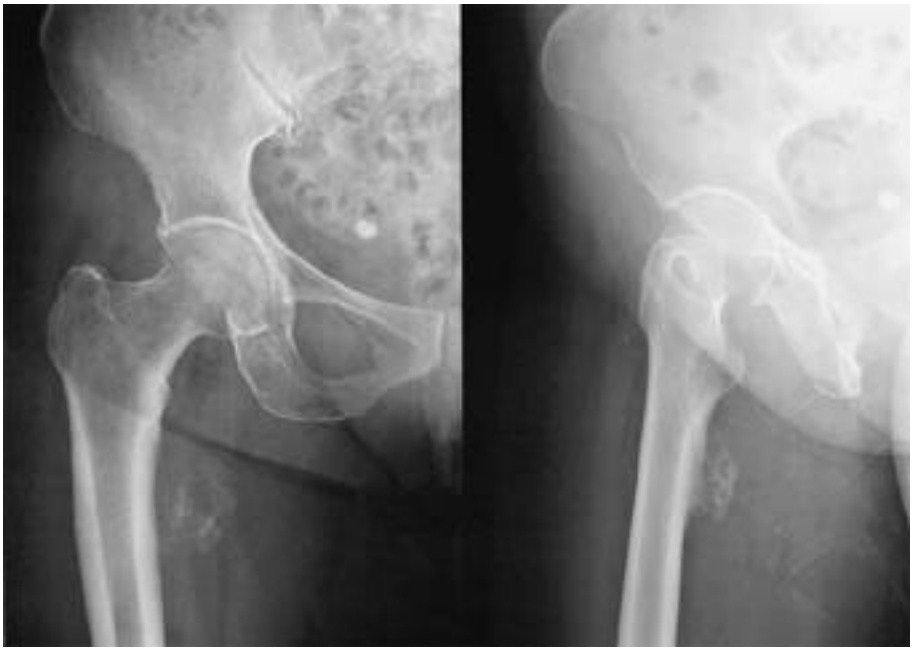

Fig. 1. Plain anterior-posterior and lateral radiographs reveal that a 3×4cm sized mass arise on the medial surface of the subtrochanteric area. The mass has an irregular and indistinct margin and exhibits moderate mineralization with a fluffy immature appearance. Cortical thickening is present but Codman's triangle or spiculated periosteal reaction is absent.

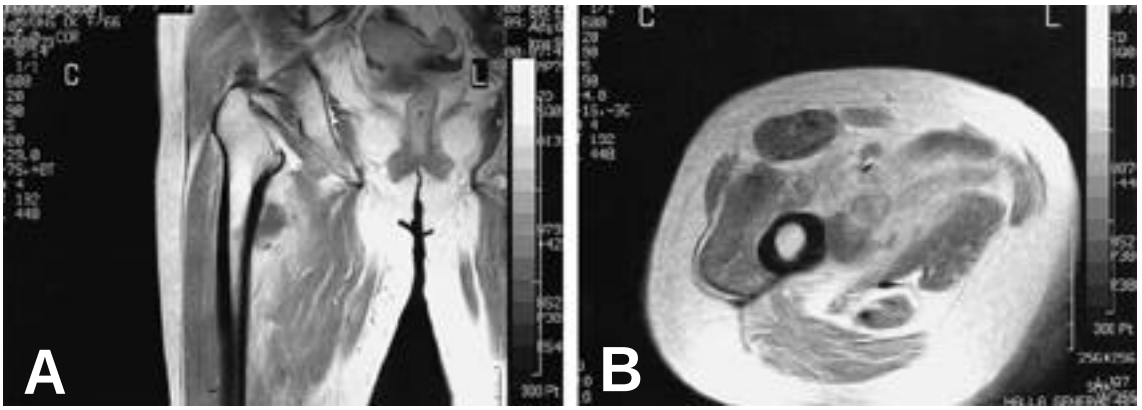

Fig. 2. Coronal(A) and axial(B) T1-weighted MR images with gadolinium enhancement reveal that the tumor shows inhomogeneous low signal intensity with peripheral enhancement and suspicious extension to the adductor muscle group with poorly defined margin. Thickening of the underlying cortex and alteration in the marrow signal intensity are also noted.

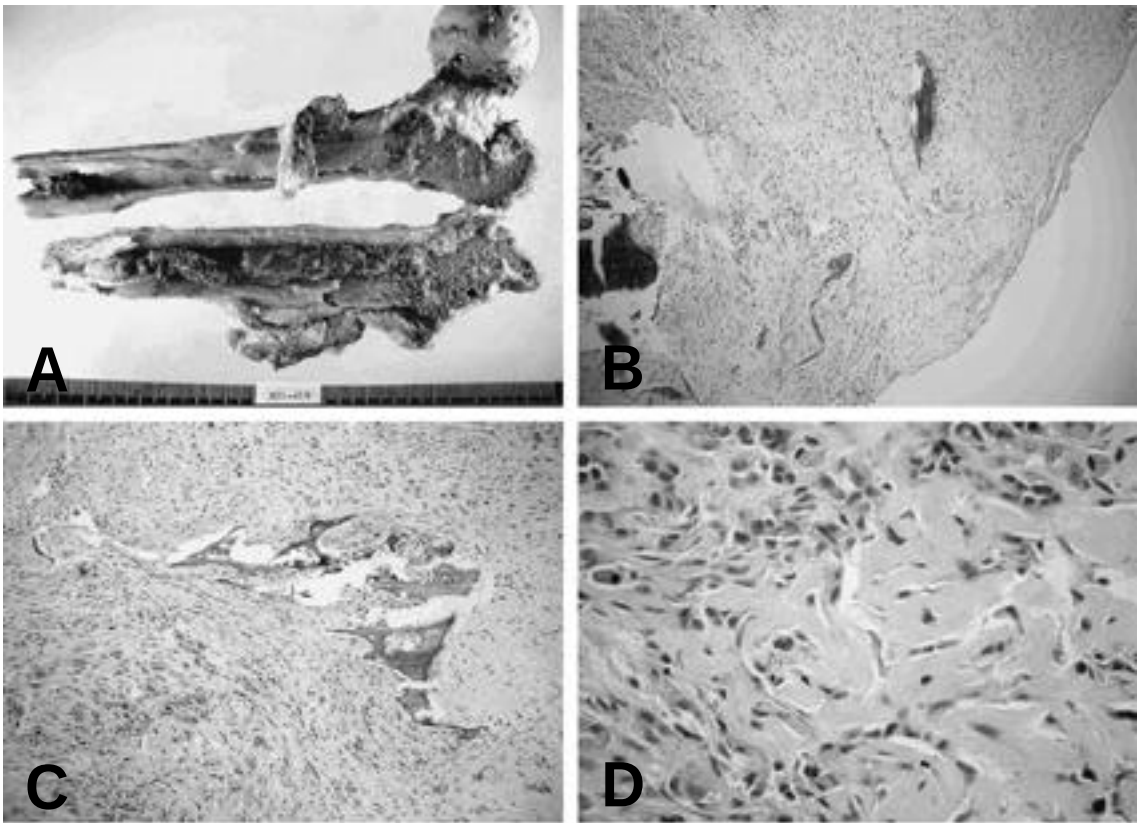

Fig. 3. (A)Gross specimen demonstrates that a tumor mass(6×3×3cm) is bulky and multilobulated and has a broad attachment to the underlying cortex of the subtrochanteric area without intramedullary extension.
(B)Histologically, most of the tumor shows active proliferation of osteoblasts associated with neoplastic bone formation(hematoxylin and eosin stain, ×100).
(C)The typical reticulated osteoid surrounded by a rim of osteoblasts is recognized by eosinophilic-staining quality, glassy appearance and irregular contours(×200).
(D)High power microscopic study shows a high grade(Broder grade 4) neoplasm characterized by cellular atypia, marked pleomorphism, abundant osteoid production identical to features of conventional osteosarcoma (×400).

sented to our hospital. Physical examination demonstrated swelling and a tender mass on palpation in the medial side of the thigh. Plain radiographs revealed that a 3×4cm sized mass arose on the medial surface of the subtrochanteric area. The mass had an irregular and indistinct margin and exhibited moderate mineralization with a fluffy immature appearance(Fig. 1). There was no evidence of distant metastasis on the chest CT scan and the whole body bone scan. On MRI, the tumor matrix appeared inhomogeneous low signal intensity with peripheral enhancement. It had a poorly defined margin and a suspicious extension to the adductor muscle group. Thickening of the underlying cortex and alteration in the marrow signal intensity were also noted(Fig. 2). The diagnosis of primary high grade surface osteosarcoma was made on an incisional biopsy of the lesion. Neoadjuvant chemotheraphy was performed by T-20 treatment reg-

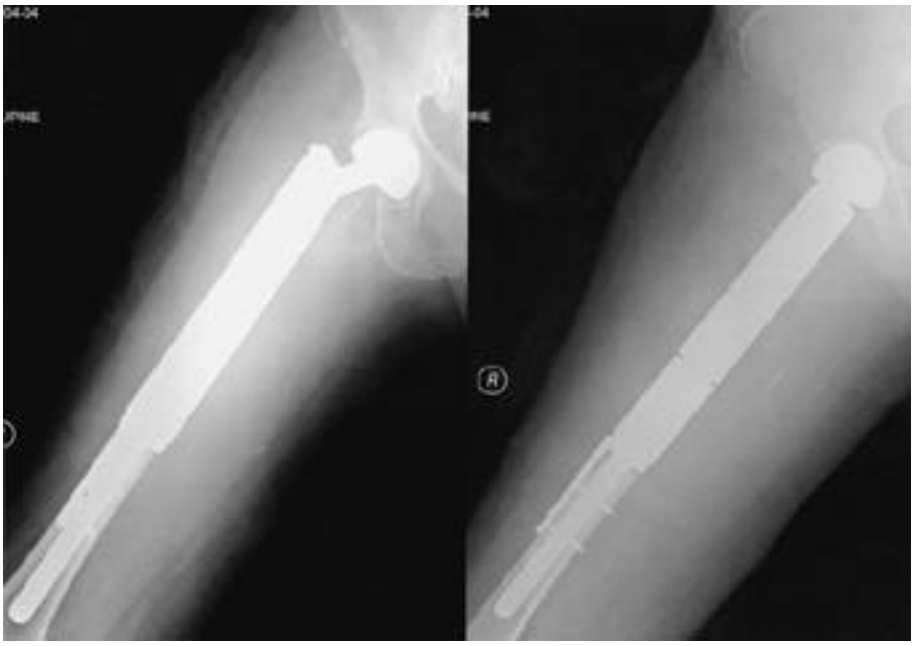

Fig. 4. Postoperative anterior-posterior and lateral radiography show limb-salvage operation with tumor prosthesis.

imen for osteosarcoma (high dose methotrexate(12 gm/m^2), ifosfamide/bleomycin, adriamycin, cisplatin/doxorubicin : Memorial Sloan Kettering Cancer Center, New York) during preoperative 6 months period. The surgery was composed of wide resection of the tumor and limb-salvage operation with tumor prosthesis(HMRS system, Howmedica, U.S.A.) (Fig. 4). Adjuvant chemotheraphy was performed with the same regimen. Because of poor general conditions including pseudomembranous colitis, pneumonia, urinary tract infection and fungal sepsis, the chemotheraphy was interrupted on the way. Unfortunately, the patient died due to pneumonia 11 months after the initial presentation. The resected specimen demonstrated that a tumor mass(6×3×3cm) was bulky and multilobulated and had a broad attachment to the underlying cortex without intramedullary extension(Fig. 3A). Microscopic study showed a high grade (Broder grade4) neoplasm characterized by cellular atypia, marked pleomorphism and abundant osteoid production identical to features of conventional osteosarcoma(Fig. 3B-3D).

DISCUSSION

Upon reviewing clinical characteristics of the high grade surface osteosarcoma, men are affected more frequently than women (male: female = 2: 1). This is in contrast to parosteal osteosarcoma, in which females outnumber males. The age at diagnosis ranged from 8 to 70 years(averaging 25 years) and 70% of the patients were in the second or third decade of life, which is similar to the age group of conventional osteosarcoma. The most frequent location of high grade surface osteosarcoma is mid-femur, in contrast to tibial proximal metaphysis in periosteal osteosarcoma and distal femoral metaphysis in parosteal osteosarcoma[6]. The

significance of the current case is that the patient was a 66-year-old female and the location was the subtrochanteric area.

Radiographically, the majority of lesions showed dense to moderate mineralization with a fluffy immature appearance. In contrast to parosteal osteosarcoma, lucent zones between the tumor and the underlying cortex were quite rare and radiating bony spicules perpendicular to the long axis of the bone, which are characteristic of periosteal osteosarcoma, were sparse[6,7].

Histologically, these tumors are indistinguishable from conventional high grade intramedullary osteosarcoma. When a high grade surface osteosarcoma consists predominantly of chondroblastic elements, differential diagnosis from the periosteal osteosarcoma can be a problem. However, in the high grade surface osteosarcoma, perpendicular arrangement of chondral lobules and malignant osteoid formation in the center of the lobules, which are characteristic features of periosteal osteosarcoma, are absent. To exclude dedifferentiated parosteal osteosarcoma, an extensive examination of the specimen is essential. If there is a coexistence of low-grade well-differentiated osteosarcoma and high grade sarcoma, then dedifferentiated parosteal osteosarcoma should be considered[4,7].

In the treatment of this tumor, preoperative chemotheraphy, adequate operative treatment and postoperative chemotheraphy are essential because its prognosis is similar to those of conventional intramedullary osteosarcomas. The treatment must be more aggressive than in the lower grade parosteal or periosteal osteosarcoma[6,7,9].

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국문초록

고등급 표면(High Grade Surface) 골육종
- 증례 보고 -

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고등급 표면(high grade surface) 골육종은 골의 표면에서 발생하는 골육종의 희귀한 아형으로, 발생빈도는 전체 골육종의 1%에 못 미친다고 한다. 몇 편의 증례 보고와 연구 논문에서 이 고등급 표면 골육종을 소개하였는데, 국내의 경우 127예의 골육종 중 1예가 고등급 표면 골육종이었음을 기술한 논문이 있으나, 환자 및 진단과 치료에 대한 정보가 포함되어 있지 않았다. 저자들은 66세 여자 환자의 대퇴골 전자하부에서 발생한 고등급 표면 골육종 1예를 진단하고, 수술 전 항암 화학 요법, 광범위 절제술과 사지 구제술 및 수술 후 항암 화학 요법을 시행하였다. 이 종양은 위치만 골 표면에서 발생할 뿐 조직학적으로 전형적인 골수강 내 골육종과 같으며, 다른 표면 골육종과는 달리 치료나 예후에 있어 전형적인 골수강 내 골육종과 같기 때문에, 진단과 치료에 있어 방골성 골육종이나 골막성 골육종 등의 표면 골육종과 구별하여야 할 것으로 생각한다.

색인 단어 : 고등급 표면 골육종, 전자하부

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