대 한 골 관 절 종 양 학 회 지
J. of Korean Bone & Joint Tumor Soc.
Volume 15, Number 1, June, 2009

슬관절 주위에 발생한 연부조직 골육종 - 증례 보고 -

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연부조직에 발생되는 골육종은 매우 드문 종양으로, 세계적으로 소수의 예가 보고되었으며 한국에서는 2례가 보고되었을 뿐이다. 문헌상 세계에서 최고령의 증례인 91 세 남자에서 외상, 방사선 조사, 화골성 근염, 피부 근염 등과 관련없이 슬관절 주위에 발생한 연부조직 골육종을 경험하였다. 절제술만으로 치료하였으며, 환자는 수술 후 1년 추시 상 생존해 있고 국소 재발이나 전이의 징후가 없으며 슬관절의 기능도 양호한 상태이다.

색인 단어: 골육종, 연부조직, 슬관절

Extraskeletal osteosarcomas are uncommon malignancies that account for about 1.2% of all soft tissue sarcomas[2,3,5,9]. It is an aggressive high grade tumor that affects adults, usually in the sixth decades of life. The prognosis is poor because of multiple recurrences and metastases. In Korea, there were only 2 cases, the first one was occurred at the gluteal region after irradiation for the treatment of cervical carcinoma and the second one was reported in the calf muscle, in which seemed apparently to have been developed from a myositis ossificans.

We introduced an extraskeletal osteosarcoma developing at the oldest age and having no predisposing factors.

Case Report

A ninety-one-year-old man with a two years history of a mass around the right knee joint was presented to our hospital. He had a medication history for the control of parkinsonism. However, he had no history of trauma, irradiation, myositis ossificans, and heterotopic ossification of dermatomyositis.

Physical examinations demonstrated a small-melon sized, firm, immobile mass at the anterolateral aspect of the right knee

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* 본 논문의 요지는 2008년도 근골격계 종양 증례 토론에서 발표되었음.

joint. There were no pain, tenderness, and heating sense, but a 2×2 cm sized necrotizing area in the center of the mass (Fig. 1).

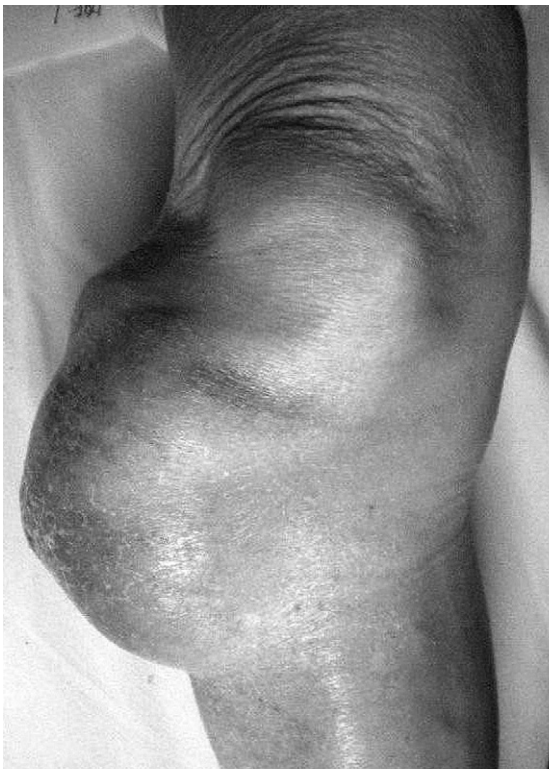

Fig. 1. Preoperative gross photograph shows a small-melon sized mass with overlying skin necrosis.

The laboratory investigations revealed no specific abnormalities, plain radiographs demonstrated a soft tissue mass at the anterolateral aspect of the right knee joint, and there was no evidence of bony involvement.

Preoperative magnetic resonance images showed a huge, well-circumscribed, heterogenous mixed echogenic mass which was septated and entirely encapsulated by well defined capsule in the lateral subcutaneous layer of the right knee (Fig. 2).

Preoperative whole body technetium-99m bone scans showed no specific abnormalities (Fig. 3).

The surgery was performed with a marginal excision of the tumor mass and a partial excision of overlying skin. The tumor mass was entirely encapsulated by relatively well defined capsule, measuring 18 cm x 10.5 cm × 8 cm. The resected specimen was soft to friable and variegated with hemorrhage and necrosis (Fig. 4A).

Microscopically, it showed malignant spindle cells with neoplastic osteoid formation. The tumor cells had obviously malignant

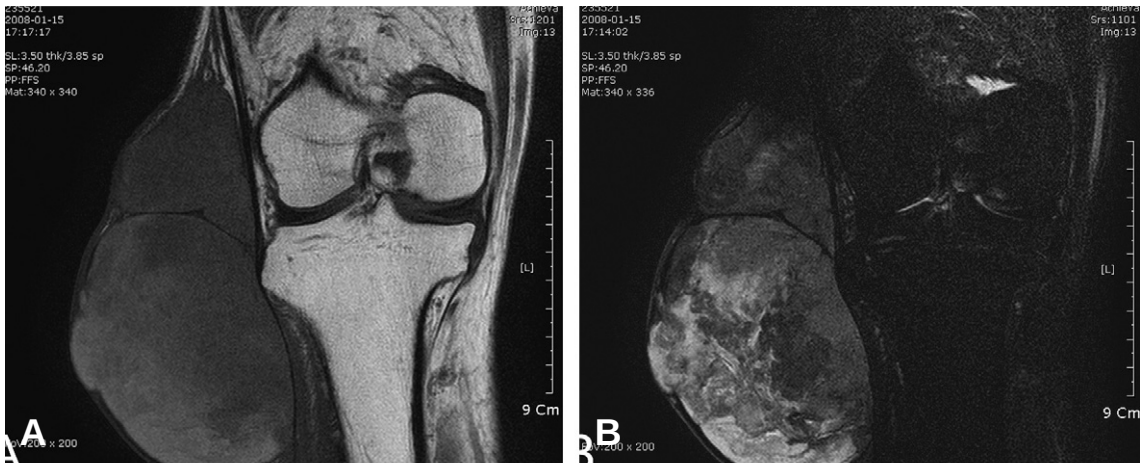

Fig. 2. (A) T1 SE coronal MR image reveals a huge mass with heterogenous high, intermediate, and low signal intensities. **(B)** T2 SPIR coronal MR image reveals a huge mass with heterogenous high and intermediate signal intensities and septation in the lateral subcutaneous layer of the right knee joint.

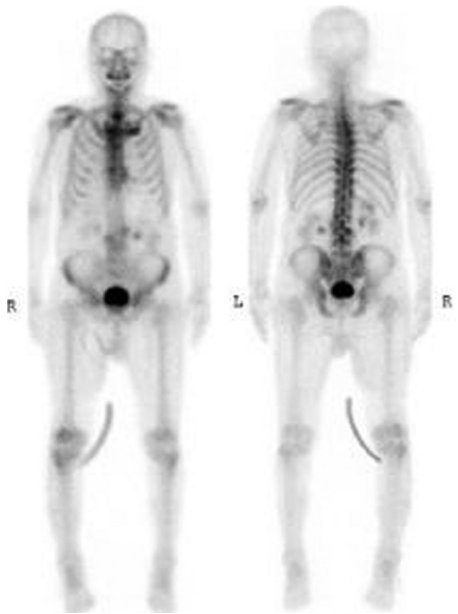

Fig. 3. Whole body Technetium-99m bone scans show no abnormal uptakes.

cytologic features, which showed significant pleomorphism and osteoid deposited in a fine, lacelike pattern (Fig. 4B-D).

After the surgery, the patient and his family refused all medical investigations and treatments including chemotherapy and radiotherapy. However, he was alive and there were no sign of local recurrence or distant metastasis and functional loss during 1-year follow-up (Fig. 5).

Discussion

The diagnosis of primary extraskeletal osteosarcoma rests on three criteria : First, the presence of a uniform morphological pattern of sarcomatous tissue that excludes the possibility of mixed malignant mesenchymal

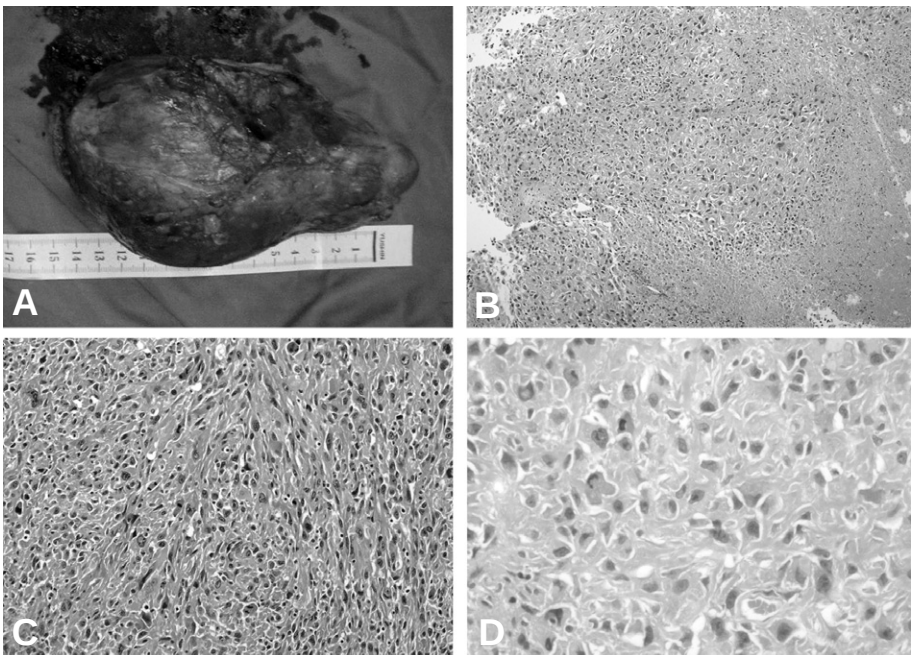

Fig. 4. (A) Postopeartive gross photograph shows a well defined mass with encapsulation. **(B)** The tumor cells produce tumor osteoid and marked coagulation necrosis (Hematoxylin and Eosin stain, x 100). **(C)** The tumor cells have obviously malignant cytologic features, which show significant pleomorphism and striking increases in the amount and granularity of chromatin (Hematoxylin and Eosin stain, ×200). **(D)** Osteoid formed by the malignant cells is narrow and laid down in a lace-like pattern (Hematoxylin and Eosin stain, ×400).

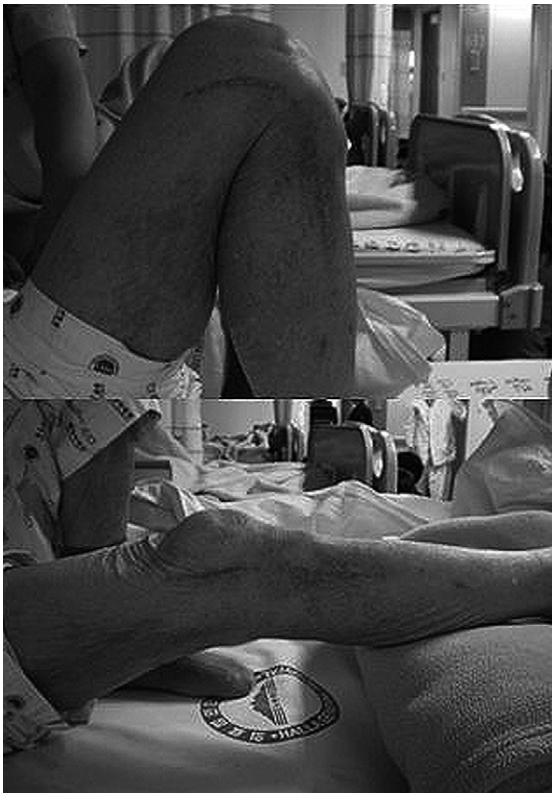

Fig. 5. Postoperative 1-year follow-up gross photograph shows no limitation of the motion of the knee joint.

tumor; second, the production by sarcomatous tissue of malignant osteoid or bone or both; and third, the ready exclusion of osseous origin[1].

Extraskeletal osteosarcoma is an uncommon tumor, of which only a small number of cases and studies have been reported[1-10]. It has been reported to constitute 1.2% of all the soft tissue sarcomas[2,3,5,9] and 4.6%[5,9] of all osteosarcomas.

Although osteosarcomas of bone occur chiefly during the first two decades of life, exteraskeletal osteosarcomas are rarely encountered in patients under 40 years of age[5]. Median age at diagnosis in several series is the sixth decades of life[2,3,5,6,7,9,10]. But, the age of our case was ninety-one that was the oldest one in the review of literatures.

Most series have found that the incidence of males and females is approximately equal for this disease[10].

The lower extremity including the buttock area is the most common site of origin, constituting almost 69%, followed by upper extremity, trunk region including retroperitoneum, and head and neck area[2,9].

Generally, there are no specific signs or symptoms. The tumor presents as a progressive enlarging soft tissue mass, which is painful in about one third of the patients[5,9]. The duration of symptoms varies from a few weeks to several months, with a mean of 6.5 months[5].

Several cases of extraskeletal osteosarcoma occurred after previous radiation therapy[1,2,3,6,9,10], trauma[1,2,3,7,10] including intramuscular injection[2,3,5,10] and fracture[5], myositis ossificans[2,5,8,10], and heterotopic ossification of dermatomyositis[4,5]. Our case was considered to a primary tumor without predisposing factors.

Simple excision is often followed by local recurrences and later, pulmonary metastases[9]. Recurrence after resection is a feature of extraskeletal osteosarcomas and usually occurs in more than half of the patients[7]. It is for this reason that a number of authors have recommended wide excision or radical resection as the initial operation[7,9,10]. Most medical centers recommend aggressive treatments with preoperative radiotherapy or adjuvant multichemotherapy[7].

Most local recurrences and distant metastases occur within 3 years postoperatively[7]. The lung are the most common site of metastasis (〉80% of cases) and the resection of the metastasis can occasionally achieve a

cure[7]. The prognosis is grave with five-year-survival rates of 25%[9], 37%[7].

We report the oldest patient with an extraskeletal osteosarcoma did not have predisposing factors. Although good results were shown on 1-year follow-up, a long term follow-up will be mandatory to verifying the final results.

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Abstract

Extraskeletal Osteosarcoma Around the Knee Joint - A Case Report -

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An extraskeletal osteosarcoma is a rare malignancy. A small number of cases and studies have been reported in the world and only two cases have been reported in Korea. We experienced an extraskeletal osteosarcoma around the knee joint of 91-year-old male who was the oldest case in the literatures. It was developed without history of trauma, irradiation, myositis ossificans, and heterotopic ossification of dermatomyositis. This patient was treated with excision alone, however he was alive and there were no sign of local recurrence or distant metastasis and functional loss during 1-year follow-up.

Key Words: Osteosarcoma, Extraskeletal, Knee

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