슬관절 주위에 발생한 연부조직 골육종
- 증례 보고 -

제주한라병원 정형외과. 병리과*
이봉진 ∙ 김태호 ∙ 하창원* ∙ 김성수

연부조직에 발생되는 골육종은 매우 드문 종양으로, 세계적으로 소수의 예가 보고되었으며 한국에서는 2례가 보고되었을 뿐이다. 문헌상 세계에서 최고령의 증례인 91 세 남자에서 외 상, 방사선 조사, 화골성 근염, 피부 근염 등과 관련없이 슬관절 주위에 발생한 연부조직 골육 종을 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. 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

※ 통신저자: 김 성 수
제주도 제주시 연동 1963-2
제주한라병원 정형외과
Tel: 064) 740-5030, Fax: 064) 743-3110, E-mail: sskos@unitel.co.kr
* 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).

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, heterogeneous 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 x 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
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
tumor: second, the production by sarcoma-
tous tissue of malignant osteoid or bone or
both; and third, the ready exclusion of
osseous origin
1).

Extraskeletal osteosarcoma is an uncom-
mon tumor, of which only a small number
of cases and studies have been reported1-10).
It has been reported to constitute 1.2% of all
the soft tissue sarcomas2,3,5,9) and 4.6%
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
age5). Median age at diagnosis in several
series is the sixth decades of life2,3,5,6,7,9,10).
But, the age of our case was ninety-one
that was the oldest one in the review of lit-
eratures.

Most series have found that the incidence
of males and females is approximately equal
for this disease10).

The lower extremity including the buttock
area is the most common site of origin, con-
stituting almost 69%, followed by upper
extremity, trunk region including retroperi-
toneum, and head and neck area2,9).

Generally, there are no specific signs or
symptoms. The tumor presents as a progres-
sive enlarging soft tissue mass, which is
painful in about one third of the patients5,9).
The duration of symptoms varies from a few
weeks to several months, with a mean of
6.5 months10).

Several cases of extraskeletal osteosarcoma
occurred after previous radiation ther-
apy1,2,3,6,9,10), trauma1,2,3,7,10), including intramuscu-
lar injection2,3,5,10) and fracture5), myositis
ossificans2,5,8,10), and heterotopic ossification of
dermatomyositis4,5). Our case was considered
to a primary tumor without predisposing
factors.

Simple excision is often followed by local
recurrences and later, pulmonary metas-
tases9). Recurrence after resection is a fea-
ture of extraskeletal osteosarcomas and usu-
ally occurs in more than half of the
patients7). It is for this reason that a num-
ber of authors have recommended wide exci-
sion or radical resection as the initial opera-
tion7,9,10). Most medical centers recommend
aggressive treatments with preoperative
radiotherapy or adjuvant multichemothera-
py7).

Most local recurrences and distant metas-
tases occur within 3 years postoperatively7).
The lung are the most common site of
metastasis (>80% of cases) and the resection
of the metastasis can occasionally achieve a

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

REFERENCES


Extraskeletal Osteosarcoma Around the Knee Joint
- A Case Report -

Bong-Jin Lee, M.D., Tae-Ho Kim, M.D., Chang-Won Ha, M.D.*, Sung-Soo Kim, M.D.

Department of Orthopaedic Surgery, Pathology* Cheju Halla General Hospital, Jeju, Korea

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

Address reprint requests to
Sung-Soo Kim, M.D.
Department of Orthopaedic Surgery, Cheju Halla General Hospital
1963-2 Yeon-dong, Jeju-si, Jeju-do, Korea
TEL: 82-64-740-5030, FAX: 82-64-743-3110, E-mail: sskos@unitel.co.kr