

# Diaphyseal osteosarcoma or synchronous osteosarcoma

Widyo Wahyu Pratomo, Rahadyan Magetsari

## ABSTRACT

**Introduction:** Osteosarcoma usually originates in the metaphyseal region of long bone, but may also arise in the diaphyseal of long bone. Diaphyseal osteosarcoma is a rare cases account for <10% of all osteosarcoma. In some cases two or more osteosarcoma lesions are seen in the same patient, at the same time (Synchronous osteosarcoma). It's unusual condition of osteosarcoma, they are found at a frequency of about 1–3% osteosarcoma cases. **Case Report:** A 15-year-old male patient, with history pain and swelling in the left knee since one year ago. The pain and swelling gradually progressive with weight loss and anorexia. Physical examination revealed big swelling on the left knee region, solid, fixed, with tenderness and deformity. X-ray shows define expansive lytic lesion replacing the entire proximal tibia and a fracture in the middle third of the left femur with lytic lesion around the fracture site. Nodular pulmonary metastatic was detected. Fine needle aspiration biopsy revealed sign of malignancy suspicious osteosarcoma. **Conclusion:** We reported a rare case of synchronous osteosarcoma. The diagnosis was confirmed with biopsy finding on the both lesion, in the left proximal tibia and in the middle third of the left femur.

**Keywords:** Diaphyseal, Malignancy, Osteosarcoma, Synchronous

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## INTRODUCTION

Osteosarcoma (OS) is a primary malignant bone tumor with a worldwide incidence of 3.4 per million people per year [1]. Osteosarcoma usually originates in the metaphyseal region of long bone, but may also arise in the diaphyseal of long bone. Among all osteosarcoma type, diaphyseal osteosarcoma is a rare case which only happen in less than 10% of all patients with osteosarcoma. In some cases, two or more osteosarcoma lesions are seen in the same patient, at the same time (Synchronous osteosarcoma). It's unusual condition of osteosarcoma, they are found at a frequency of about 1–3% osteosarcoma cases [2].

Nowaday, five-year survival rates for OS has increased compared to the last century. This is contributed by the introduction of adjuvant chemotherapy which increased the survival rate to 50%. Before chemotherapy, amputation was the routine treatment for high-grade OS. By 1990, the management of high-grade OS was shifted from amputation to chemotherapy and limb salvaging and with that combination therapy the current survival rate has increased to >65% [1].

## CASE REPORT

A 15-year-old male patient, with history pain and swelling in the left knee since one year ago. The pain and swelling gradually progressed with weight loss and anorexia. Physical examination revealed big swelling on

the left knee region, solid, fixed, with tenderness and deformity (Figure 1). X-ray shows define expansive lytic lesion replacing the entire proximal tibia (Figure 2) and a fracture in the middle third of the left femur with lytic lesion around the fracture site (Figure 3). Fine needle aspiration biopsy on the mass revealed sign of malignancy suspicious osteosarcoma (Figure 4). Fine needle aspiration biopsy was done at both sites, left proximal tibia and middle third of the left femur. Core biopsy was not done in this patient because of the limitation of the equipment in the hospital where this patient was being treated. Chest X-ray was taken to detect the metastatic to the lung. Nodular pulmonary metastatic was detected.

The diagnosis of synchronous Osteosarcoma was confirmed. Both tumor sites was found to be growing at the same time, it means that there are 2 primary tumor site in this patient. This is consistence to that of diagnosis of synchronous osteosarcoma rather than diaphyseal osteosarcoma. The patient was diagnosed with synchronous osteosarcoma stage IV-A. The patient underwent left hip disarticulation (Figure 5) and then followed by serial of chemotherapy regime with doxorubicine and cisplatine. However, the patient passed away one month after the surgery.

## DISCUSSION

Osteosarcoma (OS) is a primary malignant bone tumor with a worldwide incidence of 3.4 per million people per year. Nowadays, five-year survival rates for OS has increased compared to the last century. This is contributed by the introduction of adjuvant chemotherapy which increased the survival rate to 50%. Before chemotherapy, amputation was the routine treatment for high-grade OS. By 1990, the management of high-grade OS was shifted from amputation to chemotherapy and limb salvaging and with that combination therapy the current survival rate has increased to >65% [1].

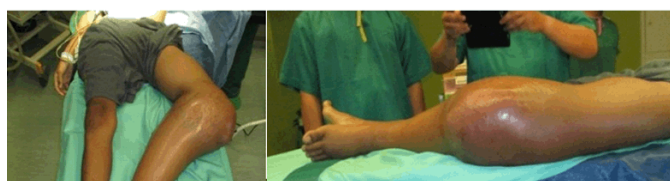


Figure 1: Mass on left knee region.

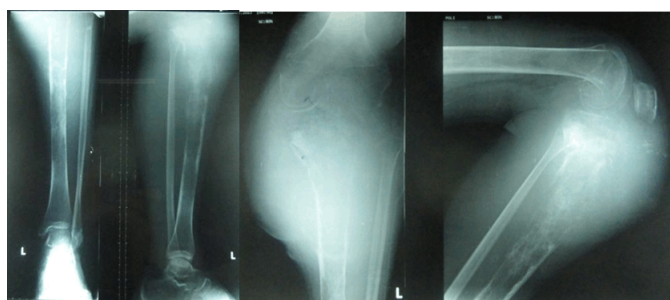


Figure 2: Osteolytic lesion in the left proximal tibia.



Figure 3: Expansive lytic lesion and a fracture in the middle third of the left femur.

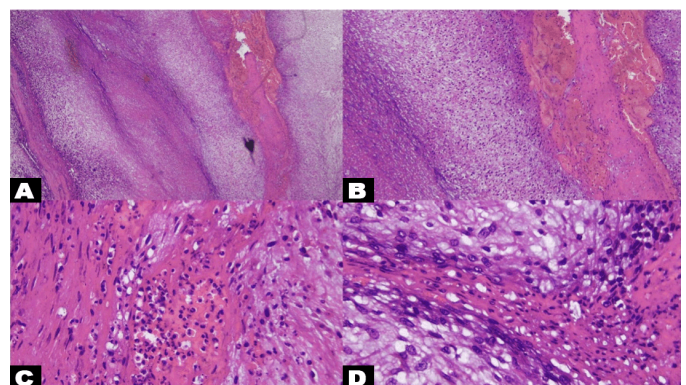


Figure 4(A-D): The biopsy showed sign of malignancy suspicious osteosarcoma.



Figure 5: Post disarticulation of the left hip.

Osteosarcomas are most often occurring in adolescents and such neoplasms are slightly more common in males than in females. Metastatic spread is predominantly haematogenous, and the lungs are by far the most common site of metastatic disease. After the introduction of chemotherapy for osteosarcoma (OS), extrapulmonary metastases are seen more often, and in earlier autopsy studies the frequency of such extrapulmonary OS manifestations was higher than observed clinically [2].

The World Health Organization's histologic classification of bone tumors divides OS into central and surface tumors, with a number of subtypes under each group [1].

Conventional OS is the most common type of OS, almost 80% of the patients with OS are conventional OS. It usually affect individuals in the first and second decades of their life. They involved the metaphysis region of the bones, preferredly occurring around the knees. The distal femur is the most common site for this type of tumor. It is very rare for Conventional Osteosarcoma to be present in the ankles and the wrist joints. Conventional OS can be subdivided into osteoblastic, chondroblastic, and fibroblastic groups depending on the predominant features of the cells, but from all those type there are no significant these differences in clinical outcome. OS is typically high grade and originates in the intramedullary cavity. The radiograph examination can show an appearance of osteolytic, osteoblastic, or both. OS can also arise in the diaphysis of long bones as well as the axial skeleton, but this is less likely to happen compared to the metaphyseal OS [1].

Telangiectatic osteosarcoma (TOS) is one of the rare subtypes of the osteosarcoma that accounts for 4% of OS. The rarity of this subtype make it poorly understood. In radiographic examination, TOS occurs in metaphyseal region, with description of geographic patterns of bone destruction, wide zone of transition and moth-eaten or permeative destruction. In histologic examination,

dilated blood-filled cavities and high-grade sarcomatous cells on the septae and peripheral rim can be seen [1].

Small-cell osteosarcoma (SOS) is one of the rarest variant of OS which constitutes only 1–2% of all OS. The histological features of SOS are similar to Ewing's sarcoma as both are made up of small round cells. But it can be differentiated from Ewing's sarcoma with the production of osteoid by tumor cells in the OS, which is not a characteristic feature of Ewing's sarcoma. In radiograph examination, a destructive process with lytic areas and sclerosis can be seen [1].

Low-grade osteosarcoma (LOS) is as rare as small-cell osteosarcoma. However, this type of osteosarcoma generally affects persons in the third or fourth decade of life. The diagnosis of LOS is occasionally difficult. LOS can be misdiagnosed as benign lesion like fibrous dysplasia or desmoplastic fibroma. There is a risk of transformation to conventional OS if treated with curettage alone, so it is suggested that LOS should be treated with wide excision. Although it is hard to recognized, this variant of osteosarcoma has significantly better prognosis compared to other variants [1].

Parosteal osteosarcoma (PAOS) is a subtype of osteosarcoma that arises from the outer layer of the periosteum. PAOS is a low-grade osteosarcoma that accounts for 4–6% of all OS. PAOS commonly affects the posterior aspect of the distal femur but may also occur in other sites including the proximal humerus and proximal tibia. Radiographic examination will show a densely ossified and lobulated mass on cortex of the bone, with the medullary cavities spared. In histologic examination, PAOS demonstrate a similar description to those of periosteal new bone reaction [1].

Periosteal osteosarcoma (PIOS) is one of the surface osteosarcoma form. This variant has a matrix component that is mainly cartilaginous and less common than parosteal. PIOS usually arises between the cortex and the cambium layer of the periosteum, and this condition will make the periosteal reaction visible on radiographs. On histopathologic examination, an intermediate-grade tumor is seen, containing a cartilaginous matrix with areas of calcification [1].

High grade surface osteosarcoma (HGOS) is a type of high grade osteosarcoma that develops on the surface of the bone from the outer cortex. This is a very rare neoplasm and constitutes <1% of all osteosarcomas. The lack of medullary involvement distinguishes this tumor from a conventional intramedullary osteosarcoma. Local growth is accelerated in HGSOS more than in parosteal osteosarcoma. On radiographic examination, this variant of OS demonstrates a surface lesion with partial mineralization, and the tumor may extend to surrounding soft tissues [1].

### Diaphyseal Osteosacoma

Most osteosarcoma occurs in the metaphysis of long bone, and diaphyseal osteosarcoma accounts for less than 10% of all osteosarcomas. The typical radiographic

appearances of diaphyseal osteosarcoma are thickening or destruction of the diaphyseal cortex with neoplastic bone and periosteal reaction extending into a broad-based soft tissue mass [3].

The presenting radiological appearances of diaphyseal osteosarcomas had four distinct patterns [4].

### 1. Group I (Typical Osteosarcoma)

The tumours in this group showed mixed osteosclerosis and osteolysis, with elevation of the periosteum forming Codman's triangles. The cortex was breached with surrounding well-defined soft-tissue swelling [4].

### 2. Group II (Atypical Sclerotic)

The radiological features were those of a dense sclerotic lesion expanding the bone with little or no separate periosteal reaction [4].

### 3. Group III

These presented with pathological fractures through purely osteolytic, ill-defined lesions. There was no bone expansion, evidence of periosteal new bone, sclerotic reaction or soft tissue extension of the tumor [4].

### 4. Group IV (Diaphyseal Osteosarcoma with remote intraosseus sclerotic lesion)

All the primary lesions showed both bone production and bone destruction. Multiple, dense, well-defined sclerotic lesions were also seen [4].

## Synchronous Osteosarcoma

Most osteosarcomas (OSs) arise as a solitary lesion. Osteosarcoma that is occur at two or more sites in a patient without pulmonary metastases are called Multifocal Osteosarcoma (MOS). This type of osteosarcoma can be synchronous or metachronous. This variant has been recognized since 1936, and constitute for 1–2% of all OS. Multifocal osteosarcoma is a rare condition with a poor prognosis despite treated with multi-agent chemotherapy, mainly high-dose methotrexate and cisplatin–doxorubicin [5].

Synchronous osteosarcoma is when two or more skeletal osteosarcoma lesions are seen in the same patient, at the same time (<6 months). It is different with metachronous osteosarcoma which has two or more skeletal osteosarcoma lesions in the same patient but in the separated time (> 6 months) [5]. MOS has poorer prognosis compared to those who only has one skeletal osteosarcoma but it does not mean that it can't be treated [2].

The comprehensive management of synchronous osteosarcoma has not been investigated in a large series. Bacci G *et al* who study 16 patients with synchronous osteosarcoma that was given neoadjuvant chemotherapy and surgery showed that the prognosis of the patients

still poor despite of the chemotherapy and surgery. In that study, from 16 patients that was given the therapy, 12 patients experienced relapse and died [6].

In this case, at first, the patient was diagnosed with synchronous osteosarcoma and diaphyseal osteosarcoma as a differential diagnosis. The mass at the femur region and proximal tibia region was known to be growing at the same time. The mass was firstly felt by patient at 1 year prior to admission to the hospital. At the hospital, from physical examination, laboratory finding, X-ray and biopsy, it was confirmed that the diagnosis is synchronous osteosarcoma.

## CONCLUSION

The case report highlights a rare case of diaphyseal osteosarcoma or synchronous osteosarcoma. After a comprehensive examination it was confirmed that the diagnosis is synchronous osteosarcoma.

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## Author Contributions

Widyo Wahyu Pratomo – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Rahadyan Magetsari – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

**Guarantor of Submission**

The corresponding author is the guarantor of submission.

**Source of Support**

None.

**Consent Statement**

Written informed consent was obtained from the patient for publication of this case report.

**Conflict of Interest**

Authors declare no conflict of interest.

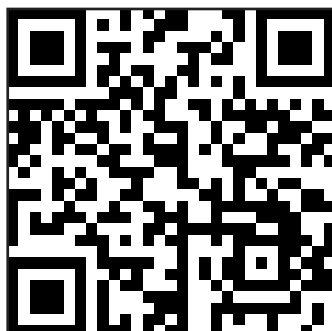
**Data Availability**

All relevant data are within the paper and its Supporting Information files.

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