A Rare Case of Periosteal Osteosarcoma with Pathological Fracture at Metaphysis of Distal Femur

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Abstract: Osteosarcoma is one of the common malignancies affecting young adults. The most common sites are the femur (42%, with 75% of tumours in the distal femur). A pathological fracture in an osteosarcoma is rare with an incidence, either at diagnosis or during the pre-operative treatment of 5-10%. A 20-year-old male complained of swelling in lower end of right femur since one year. The swelling was insidious in onset and progressive in nature. Patient complained of pain at the site of swelling with significant increase in the size of swelling for past one month. There was no history of trauma and evidence of Café-au-lait spots or other signs of neurofibromatosis. Radiograph of right femur revealed wide zone of transition, cortical destruction, perpendicular and aggressive periosteal reaction and soft tissue involvement at the distal end of right femur. Pathological fracture was noted at metaphysis of distal femur. Reconstructed CT image of right femur showed evidence of chondroid and osteoid matrix in the posterior aspect of distal femur. No intramedullary extension was noted. Later wide excision was performed and histopathology examination revealed findings of consistent with periosteal osteosarcoma. Here we present a rare case of periosteal osteosarcoma with pathological fracture at metaphysis of distal femur.

Key words: Periosteal Osteosarcoma, Pathological fracture, Periosteal reaction, Chondroid matrix, Cortical destruction, Osteoid matrix.

INTRODUCTION

Osteosarcoma is one of the common malignancies affecting young adults. The most common sites are the femur (42%, with 75% of tumours in the distal femur), the tibia (19%, with 80% of tumours in the proximal tibia), and the humerus (10%, with 90% of tumours in the proximal humerus). Other likely locations are the skull or jaw (8%) and the pelvis (8%) [1]. Rachel et al in their research of epidemiology of bone cancer in 0-39 years old in Northern England, discovered incidence rates (per million persons per year) for osteosarcoma were of 2.5 per million per year in age group of 0-14, 4.5 at age 15 - 29 years and 1.0 at age 30 - 39 years [2]. A pathological fracture in an osteosarcoma is rare with an incidence, either at diagnosis or during the pre-operative treatment of 5-10% [3].

Here we present a rare case of periosteal osteosarcoma with pathological fracture at metaphysis of distal femur.

EXPERIMENTAL SECTION

A 20-year-old male complained of swelling in lower end of right femur since one year. The swelling was insidious in onset. There was no history of trauma. Initially the swelling was painless, smaller in size and gradually progressive in nature. Patient complained of pain at the site of swelling with significant increase in the size of swelling for past one month. Patient had difficulty to perform routine activities since one month.

On examination, there was warmth and tenderness over the site of swelling. On palpation the swelling was firm in consistency, non-fluctuant and measured ~ 10 x 7 cm. There were no Café-au-lait spots or other signs of neurofibromatosis. There was no
Osteosarcoma is one of the common malignancies affecting young adults. The most common sites are the femur (42%, with 75% of tumours in the distal femur), the tibia (19%, with 80% of tumours in the proximal tibia), and the humerus (10%, with 90% of tumours in the proximal humerus). Other likely locations are the skull or jaw (8%) and the pelvis (8%) [1]. Rachel et al., in their research of epidemiology of bone cancer in 0-39 years old in Northern England, discovered incidence rates (per million persons per year) for osteosarcoma were of 2.5 per million per year in age group of 0-14, 4.5 at age 15 - 29 years and 1.0 at age 30 - 39 years [2]. A pathological fracture is rare entity in an osteosarcoma which is found at diagnosis or during the pre-operative treatment of 5-10% cases [3].

Periosteal osteosarcoma is a rare primary malignant bone tumour and is a subtype of osteosarcoma. It is a surface lesion without medullary involvement. It was initially recognized by Ewing in 1939. Lichtenstein in 1959 delineated periosteal osteosarcoma as a periosteal counterpart of central or inter-medullary osteosarcoma [4]. Periosteal osteosarcoma is an intermediate-grade chondroblastic osteosarcoma emerging on the surface of bone. Periosteal osteosarcoma, as an intermediate tumour, has less aggressive biologic behaviour with a lower tendency to metastasize compared to conventional osteosarcoma or high-grade surface osteosarcoma [5-11]. Conventional osteosarcoma is a primary, intramedullary, high-grade malignant tumour in which neoplastic cells produce osteoid, even if only in small amounts. Osteosarcoma is the most common nonhemopoietic, primary malignant tumours of bone with an estimated incidence of 4 to 5 per million population. High-grade surface osteosarcoma is a high-grade, bone-forming malignancy that arises from the surface of bone. High-grade surface osteosarcoma is a rare entity and comprises <1% of all osteosarcomas.

Conventional osteosarcoma and high-grade surface osteosarcoma share the same prognosis and the same treatment. The use of surgery is curative in only <15% of patients, whereas the addition of systemic chemotherapy has resulted in survival rates of 60% to 80% for those patients who do not have clinically evident metastases at presentation.

Another osteosarcoma arising on the bone surface is parosteal osteosarcoma, which is a low-grade osteosarcoma that accounts for approximately 4% of all osteosarcomas. Its prognosis is excellent, with a 91% overall survival rate at 5 years with surgical treatment alone, and it does not require the use of chemotherapy [5, 9-11].

Wide excision of the tumour is the mainstay of therapy for periosteal osteosarcoma, although controversy remains about the role of chemotherapy [8, 12-15]. Ill-defined lobules of atypical cartilage are found histologically in periosteal osteosarcoma.

Histopathological appearance of periosteal osteosarcoma shows cartilaginous areas separated by sarcomatous bands of undifferentiated spindle cells with primitive appearance. Within these bands, osteoid and immature bone produced by neoplastic cells are seen. Perpendicular streaks of reactive bone with osteoblastic rimming traverse the neoplasm [16].

Periosteal osteosarcoma is an intermediate-grade chondroblastic osteosarcoma arising on the surface of bone. Perios-teal osteosarcoma, as an intermediate tumor, has less aggressive biologic behavior with a lower propensity to metastasize compared with conventional osteosarcoma or high-grade surface osteosarcoma. Perios-teal osteosarcoma is an intermediate-grade chondroblastic osteosarcoma arising on the surface of bone. Perios-teal osteosarcoma, as an intermediate tumor, has less aggressive biologic behavior with a lower propensity to metastasize compared with conventional osteosarcoma or high-grade surface osteosarcoma.
Later wide excision was performed and histopathology examination revealed findings of consistent with periosteal osteosarcoma.

**Fig. 1:** Radiographs of right knee (AP and lateral view) Orange arrow represents pathological fracture at distal metaphysis of right femur

**Fig. 2:** Radiograph of right femur (Lateral view) Orange arrow represents pathological fracture at distal metaphysis of right femur

**Fig. 3:** Reconstructed CT image of right knee Exophytic and lobulated tumour with multiple hypodense foci representing chondroid matrix and multiple hyperdense foci representing osteoid matrix in the posterior aspect of distal femur

**Fig. 4:** Shows specimen of right distal femur after wide excision surgery

**CONCLUSION**

Here we present a rare case of periosteal osteosarcoma with pathological fracture at metaphysis of distal femur.
REFERENCES


