Primary epiphyseal Ewing sarcoma: a case report

Murat BÜLBÜL, Harzem ÖZGER, Bilge BİLGİÇ, Levent ERALP

Department of Orthopedics and Traumatology, Faculty of Medicine, Istanbul Medipol University, Istanbul, Turkey;
Department of Orthopedics and Traumatology, Istanbul Faculty of Medicine, Istanbul University, Istanbul, Turkey;
Department of Pathology, Istanbul Faculty of Medicine, Istanbul University, Istanbul, Turkey

Ewing sarcoma is a frequently seen malignant bone tumor of the childhood. The tumor involves the diaphyseal and metaphyseal bone and cases located in the epiphysis are unusual. We present a case of Ewing sarcoma limited to the epiphysis in an immature skeleton. We would like to emphasize the importance of the biopsy without contaminating the joint space which will reduce the morbidity during the curative surgery.

Key words: Bone tumors; epiphysis; Ewing sarcoma.

Ewing sarcoma is one of the most frequently seen primary malignant tumor occurring between the first and third decade. Clinical, radiological and histological findings are well-defined. The extent of this tumor is generally localized in the diaphysis and metaphysis of the long bones. Epiphyseal involvement occurs generally with invasion of a primary metaphyseal or diaphyseal tumor. Primary epiphyseal involvement is quite rare. Epiphyseal Ewing sarcoma may be mistaken as a benign, aggressive tumor of the epiphysis, such as chondroblastoma, osteoblastoma, giant cell tumor or eosinophilic granuloma, presumably leading to a suboptimal treatment.

We report a case of epiphyseal Ewing sarcoma. We think that the diagnostic work-up and therapeutic approach of the case would be of interest to the surgeons dealing with tumor surgery.

Case report

A 16-year-old male patient presented with right knee pain of more than one year duration. He had initially been treated in another hospital with non-steroidal anti-inflammatory medication and a previous biopsy had not revealed sufficient material for histopathological diagnosis. During physical examination, the patient had tenderness on the lateral part of the upper leg, mainly on the lateral proximal tibia. The range of motion of the knee was limited between 30 and 90 degrees. Direct radiographs of the right proximal tibia showed a predominantly sclerotic heterogeneous appearance of the epiphysis (Fig. 1).

A lytic-sclerotic type pathological bone structure change in the right tibia proximal epiphysis with thinning of the cortex in the anterior contour and generalized small cortical defects were seen in the CT examination (Fig. 2). In the T1- and T2-weighted MRI sequences, a heterogeneous area of pathological intensity in the right tibia proximal epiphysis and scattered open areas of a permeative type in the bone cortex contours was observed. Extensive cortical destruction-expansion areas were seen in the anterior from infrap-
atellar fatty tissue protruding anteriorly together with the soft tissue component. Small areas of cortical expansion were located outside the area of extensive cortical destruction in the anterior. The second soft tissue expansion was in the posterior contour towards the popliteus muscle group.
The soft tissue mass in the infrapatellar fatty tissue was located in front of the anterior cruciate ligament tibial attachment and extended between the cruciate ligaments. The infrapatellar tendon tibial attachment was surrounded by the mass. Intravenous contrast dye showed heterogeneous involvement of the lesion. It showed immediately in front of the posterior cruciate ligament attachment on the epiphysis line, and two separate areas extending from the epiphysis line to the metaphysis lateral to the anterior cruciate ligament tibial attachment. Apart from these two separate extended areas, a metaphyseal-diaphyseal wide area of edema appeared reactive (Fig. 3). Increased activity in the proximal end of the right tibia in the late term was observed through three-phase bone scintigraphy (Fig. 4). No metastatic lesions were determined through CT of the thorax. Hypermetabolic involvement in the proximal right tibia was observed on Positron emission tomography (Fig. 5). In the histopathological examination of the sample obtained by incisional biopsy, tumoral infiltration of small round cells was observed (Fig. 6). A differential diagnosis of small cell osteosarcoma was discounted due to the lack of osteoid and positive MIC-2. The possibility of lymphoma was removed as lymphoid markers were negative and a diagnosis of Ewing sarcoma was made.

The patient underwent neoadjuvant chemotherapy four times preoperatively. CT revealed the continuation of the cortex on the extensive lytic area in the anterior. The lesion continued to show a lytic-sclerotic heterogeneous appearance and it was noticeable that lytic sections in the sclerotic areas were more pronounced. No comment can be made related to the extension to the metaphysis. MRI examination following chemotherapy determined an evident regressive change in the lesion and the soft tissue mass of the lesion; and areas of cortical opening were seen to have completely disappeared. Surgical treatment was applied after the neoadjuvant chemotherapy. An extensive tumor resection was performed and coverage of

Fig. 4. Three-phase scintigraphy images of the right end of proximal tibia showing increased activity in the late-term.

Fig. 5. Positron emission tomography images showing hypermetabolic involvement in the proximal right tibia. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]
the tumor resection prosthesis was achieved with medi-al gastrocnemius flap and skin graft. The specimen taken from resection was determined to be 95% necrotic. Chemotherapy was continued after surgery.

Discussion

Ewing sarcoma is one of the most frequently seen malignant bone tumors. Although it most commonly appears in the metaphysis and diaphysis of the long bones, primary involvement in the epiphysis can occur rarely. To our knowledge, only one case of Ewing sarcoma of primary epiphysial origin presenting together with a case of osteosarcoma was reported by Muscolo et al.[5]

The presented case is one of Ewing sarcomas with primary epiphysial involvement not spread from the metaphysis. The metaphysis changes in the MR findings are associated with edema and there is no question of spread from the metaphysis to the epiphysis as following neoadjuvant chemotherapy, this edema was seen to disappear. The symptoms extended throughout the knee joint and proximal tibia. On direct radiographs and CT, mixed lytic and sclerotic areas were seen. Generally, in this situation, occurrences such as chondroblastoma, giant cell tumor, epiphysial enchondroma, eosinophilic granuloma, neuroblastoma metastasis, and atypical osteomyelitis are seen.

Following surgical procedure and pathological confirmation of diagnosis of a malignant case of Ewing sarcoma, an extensive resection and reconstruction can be performed by sacrificing only one joint surface. However, when contamination of the joint tumoral cells is present, a more radical procedure such as closed joint resection and gastrocnemius pedicle flap, as performed in this case, may be applied, causing high morbidity. To avoid such situations, a needle (Tru-cut, Jamshidi) biopsy should be performed at the center the treatment is to be completed. Performing the biopsy and treatment by the same team and at the same center reduces the potential mortality and morbidity.

In conclusion, although rare, primary epiphysial involvement of malignant bone tumors may occur. Most epiphysial lesions are benign lesions. However, mistaking malignant epiphysial bone tumors such as Ewing sarcoma as a benign tumor may delay the diagnosis and curative surgical procedures.

Conflicts of Interest: No conflicts declared.

References