Osteosarcoma in the Proximal Humerus of an Adult Male: A Case Report and Review of the Literature

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Summary

Osteosarcoma is the most common primary bone malignancy. The incidence rate of the disease peaks during adolescence when bone growth is the most rapid. Primary osteosarcoma is typically seen in patients aged 10-20 years old, with 75% of cases under the age of 20 years. The incidence of osteosarcoma is 1.5 times higher in males than females. Osteosarcoma commonly occurs in the metaphyseal region of the long bones with the most commonly affected sites being the knee and humerus at rates of 60% and 10%, respectively. This paper, presents the clinical and radiological findings of a young adult male patient with osteosarcoma which was detected in the proximal humerus after presentation with shoulder pain. The diagnostic value of direct radiography is emphasized.

Key Words: Osteosarcoma, adult, direct radiography

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Introduction

Osteosarcoma is a bone tumor of malignant character formed from mesenchymal cells of immature bone and osteoid [1]. The age distribution of osteosarcoma is typically shown to be bimodal, with the first peak between 10-20 years and the second in adults in the 5th and 6th decades of life [2]. The incidence of primary osteosarcoma peaks in adolescence when bone growth is rapid and they are often located in the metaphysis of the long bones. The most commonly involved areas are the femur at a rate of 42% (75% in the distal), the tibia at 19% (80% in the proximal) and the humerus at 10% (75% in the proximal). Secondary osteosarcoma peak in elderly adult patients and predisposing factors can be determined as a history of previous exposure to radiation, Paget's disease, chronic osteomyelitis or fibrosis dysplasia. Secondary osteosarcoma involve the axial bones such as the pelvis and spine in particular.

Direct radiography and magnetic resonance imaging (MRI) are of great importance in the radiological diagnosis of the disease. A definitive diagnosis is made as a result of biopsy from the lesion area.

The Case

A 34-year old male patient presented at our polyclinic with complaints of pain in the right shoulder and arm which had been ongoing for 2-3 months and restricted movement of the shoulder joint. In the physical examination, there was limited and painful range of movement in the right shoulder joint and the right shoulder and proximal humerus were found to be painful on palpation. No redness or increased temperature was determined. There were no systemic symptoms such as fever, sweating, loss of appetite or loss of weight.

In the laboratory examination, the values of haemogram, erythrocyte sedimentation rate, CRP, lactate dehydrogenase (LDH) and alkaline phosphatase (ALP) were within normal limits.

On direct radiograph, localized in the humerus head and proximal metaphyseal-diaphyseal section, an osteoblastic lesion 11x4.5 cm in size was observed containing dense sclerotic changes, which was seen to extend beyond the cortex. The preoperative direct radiograph of the patient is shown in Figure 1.

On magnetic resonance imaging, an extensive mass lesion 11.6x4.2 cm in size was defined in the humerus head posterolateral section, which had passed through a wide zone with roots from the medullar area, extending beyond the bone to the muscle structures by cortex destruction in the proximal diaphyseal area. Within the lesion, scattered areas without signal, consistent with calcification were determined as heterogeneous intense contrast from examination with contrast enhancement. Preoperative MR image of the patient is shown in Figure 2.

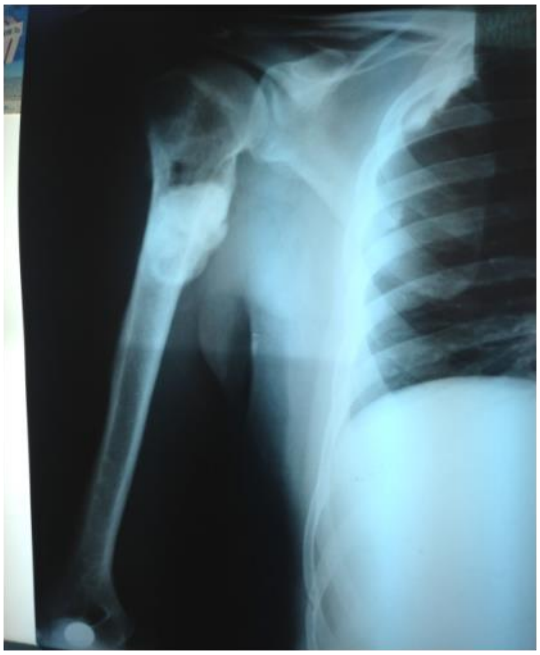

Figure 1. AP radiograph of the right humerus

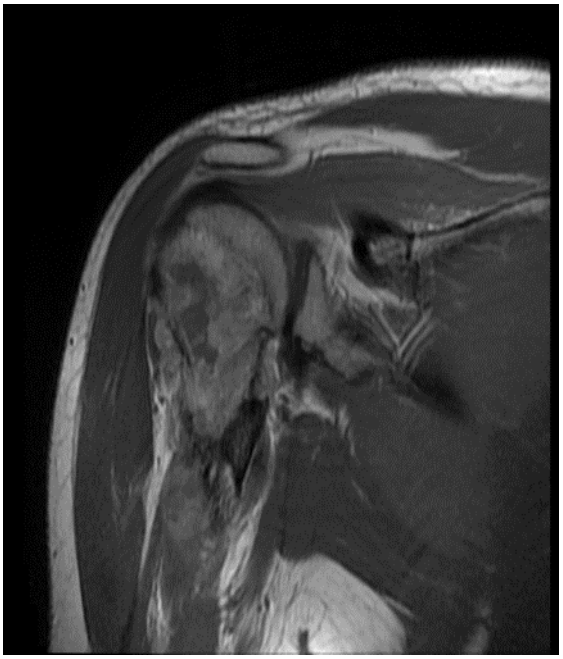

Figure 2. MRI of sagittal slice with contrast

After biopsy made in the lesion area, neo-adjuvant chemotherapy was started with a diagnosis of osteosarcoma. A positive response to chemotherapy was observed on the 2-month follow-up MRI, so tumor resection was performed and reconstruction surgery with a tumor prosthesis was applied. Adjuvant chemotherapy was started following surgery. At 9 months postoperatively, the patient presented at the Chest Clinic with complaints of cough and respiratory difficulties. On bronchoscopy, an endobronchial lesion was determined, completely blocking the left main bronchus, and the mass was removed with foreign body forceps. The biopsy result was reported as malignant mesenchymal tumor (osteosarcoma

metastasis). At the 9th postoperative month the patient was still undergoing treatment in the Oncology Clinic.

Discussion

With the exceptions of multiple myeloma and metastatic bone tumors, osteosarcoma is the most frequently seen bone tumor [3]. It is often located in the metaphysis of the long bones. In the case presented here, the mass was localized in the proximal metaphyseal-diaphyseal area of the right humerus.

Patients generally present with symptoms of 1–3 months duration. There is a mean 15-week delay period between the onset of symptoms and a definitive diagnosis of osteosarcoma [4]. In the majority of patients with high-grade osteosarcoma, there is a 2-3 week period of intermittent pain following an initial minor trauma. Then the pain becomes more severe and a sensitive soft tissue mass develops approximately 2 months after the onset of pain. At this stage, osteosarcoma can be easily diagnosed both clinically and radiologically [5]. Occasionally, following intensive physical exercise or a minor trauma, the tumor is noticed with a pathological fracture secondary to bone weakening in the lesion area. When a pathological fracture develops, sudden and severe pain occurs. Pathological fractures are more often determined in pediatric patients.

In 80% of patients, when diagnosis is made, the tumor is localized and non-metastatic in character. Occasionally, effusion in the joint, increased temperature and hyperemia in the skin over the tumor and thinning in the distal of the lesion due to muscle atrophy secondary to non-use, may be determined. Systemic symptoms such as loss of appetite, weight loss and fever are not common. The current case presented with complaints of pain and restricted movement which had been ongoing for 2-3 months.

Although there is no particular laboratory finding for osteosarcoma, increased levels of erythrocyte sedimentation rate, ALP, LDH and ferritin may be determined. No abnormalities were determined in the laboratory findings of the current case.

Direct radiography findings play a significant role in diagnosis. Typical appearances of osteosarcoma on direct radiography include mixed lytic and sclerotic areas, cortical

destruction, periosteal new bone formation and soft tissue mass. Most cases of osteosarcoma at a relatively advanced stage are diagnosed easily both clinically and radiologically.

Although early radiographic changes cannot be very well differentiated in the early stages of the disease when patients have complaints of pain, changes in the intensity of bone marrow together with areas of trabecular destruction on direct radiography, are seen as the main finding of early stage osteosarcoma. Later, cortical destruction, soft tissue mass outside the bone and scattered areas of increased intensity in the metaphyseal region can be observed. Additional to these findings, the presence of an evident non-specific periosteal reaction strongly increases the probability of a malignant bone tumor [5]. On the right humerus direct radiograph of the current case, an osteoblastic lesion was observed which contained intense sclerotic changes and was localized in the humerus head and proximal metaphyeso-diaphyseal section extending beyond the cortex.

Computerized tomography (CT) plays more of a role in biopsy and staging of osteosarcoma than direct evaluation of the tumor. CT is superior to MRI in the visualization of cortical changes and new bone formation in the affected bone.

In the evaluation of the relationship of tumor soft tissue with skip lesions, MRI is superior to CT. It is important in the evaluation of neighboring muscle groups, subcutaneous fat tissue, and joint and neurovascular structures [6]. In addition, in the evaluation of the response to neo-adjuvant chemotherapy prior to surgical resection, it is used in the planning of correct staging and limb-sparing resection. Biopsy is the gold standard in the diagnosis of osteosarcoma [7].

Radiological differential diagnosis of metastatic bone tumor include giant cell tumor of the bone and aneurismal bone cyst. Metastatic bone tumors are often determined in patients over 60 years of age and particularly in the spine and pelvis [8]. In adult patients aged over 40 years, multiple lytic lesions on radiography almost always suggest multiple myeloma [9].

The spread of osteosarcoma is basically hematogenous. The lungs are the area of first involvement in 90% of cases found to have metastasis [10]. The prognosis of metastatic osteosarcoma is poor, with a reported rate of 29% for 5-year survival [11]. Prognostic factors have been reported to be age, gender, involvement of the trunk, presence of metastatic lesion

on diagnosis, chemotherapeutic effects, tumor size, serum alkaline phosphatase level, recurrence and p-glycoprotein level. Of these factors, the presence of necrosis in the histological analysis of 90% of excised tumors and the evaluation of the response to chemotherapy are the most meaningful methods of determining prognosis [12]. In a study by He et al, CD133 expression determined in tissue with osteosarcoma was found to be related to a high risk of lung metastasis and short survival [13]. Young adult patients have a worse prognosis than pediatric or elderly adult patients [14].A retrospective examination by Haddox et al of 111 patients with high grade osteosarcoma, compared young adults aged 18-40 years with pediatric patients aged below 18 years, and a higher rate of survival was determined in the pediatric group. It was also determined from data that 61% of the young adult patients had received less cumulative chemotherapy than the pediatric group [15].

The current treatment protocol valid for osteosarcoma patients is the application of limb-sparing surgery and chemotherapy together. In patients who have a positive response to neo-adjuvant (preoperative) chemotherapy, surgical treatment can be applied at 9-12 weeks. Usually after excision of a bone with a tumor, the placement of a bio-compatible metal prosthesis can be made and even though it is rare, following mechanical cleaning of the tumorous tissue from the bone after excision of a bone with a tumor, the bone may be replaced with autoclave sterilization or pasteurization or extracorporeal phototherapy [16]. With the addition of chemotherapy to surgical treatment, the 5-year survival rate in osteosarcoma has increased from 20% to 70% [17]. As osteosarcoma is a radio-resistant tumor, there is no place for radiotherapy in the treatment.

In the current case, as lung metastasis was determined 11 months after diagnosis despite surgical treatment and chemotherapy, the prognosis was poor.

In this paper, the clinical, direct radiography and MRI findings are presented of a male patient diagnosed with osteosarcoma in the proximal humerus. The aim of this paper was to draw attention to adult patients presenting with knee and shoulder pain taking into consideration the frequently involved areas of osteosarcoma.

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