



Osteosarcoma of the rib: A rare presentation

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Cite this article as: Büyükkapu Bay S, Kebudi R, İribaş A, et al. Osteosarcoma of the rib: a rare presentation. Turk Pediatri Ars 2018; 53: 57-60.

Abstract

In children and adolescents with chest pain and dyspnea, pneumonia, pleural effusion, and empyema are the frequent causes in the differential diagnosis. Malignant tumors of the chest wall are rare and most originate from the ribs. In children, the most frequent malignant tumor of the rib is Ewing's sarcoma. Osteosarcomas of the rib are very rare. Osteosarcoma has a predilection for rapidly growing long bones including the femur, tibia and humerus in adolescents. In this paper, we present an adolescent girl who presented with chest pain and dyspnea with osteosarcoma that originated from the rib and extended to the right hemithorax.

Keywords: Adolescent, osteosarcoma, rib

Introduction

In children and adolescents who present with chest pain and respiratory distress, infectious, metabolic, and psychologic causes should also be considered in addition to causes related with the lung, heart, gastrointestinal system, and musculoskeletal system, though the frequencies vary by age. The most common causes related with the lungs include pneumonia, pleural fluid, pneumothorax, and empyema. Primary malignant tumors of the thoracic wall occur rarely (1). Malignant bone tumors localized in the thoracic wall originate most commonly from the ribs. In pediatric patients, the most common malignant rib tumor is Ewing sarcoma. Primary osteosarcoma of the ribs occurs very rarely (2, 3).

In this article, a case of osteosarcoma originating from the rib and occupying one hemithorax is examined.

Case

A fourteen-year-old girl presented to the Chest Diseases outpatient clinic with intermittent pain in the right side of her back, which started two months ago and had increased and dyspnea developed. Lung X-ray revealed a radiopacity occupying the right hemithorax, and thoracic computed tomography (CT) revealed consolidation occupying the whole right lung and pleural fluid. A thoracic tube was placed with a prediagnosis of empyema and antibiotic treatment was initiated. After the pleural fluid was drained off, a mass was found on the follow-up thoracic CT and an incisional biopsy was performed. Pathologic examination revealed osteosarcoma (chondroblastic) and the patient was referred to our division.

Her physical examination was as follows: paleness and malaise were present and the respiratory rate was increased. Tubular soufflé was heard in the upper part of

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Received: 06.08.2016

Accepted: 17.03.2017

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DOI: 10.5152/TurkPediatriArs.2018.4689

the right lung. Respiratory sounds could not be heard in the middle and lower areas. Dullness was found on percussion. The complete blood count and biochemical values were within the normal limits except for an erythrocyte sedimentation rate of 63 mm/h and a mild increase in lactate dehydrogenase (530 U/L). Radiopacity was found in the whole right hemithorax in addition to a reduction in aeration in the upper part of the right lung and the heart was shifted to the left hemithorax (Figure 1). On thoracic CT, a mass with a size of 12x13 cm, which caused bone destruction, was found anterior to the fourth right rib. Its right thoracic half extended

to the anteroposterior-diaphragmatic face (Figure 2). The patient was diagnosed as having osteosarcoma that occupied the whole right hemithorax by deflating the right lung and compressed the right side of the heart. Positron emission tomography (PET)-CT revealed no distant metastasis. Chemotherapy with an osteosarcoma treatment protocol was initiated in our center (4). Pain in the right back part of the thoracic wall and dyspnea regressed and her symptoms improved after the second course of treatment. The patient's respiratory sounds were found to be normal starting from the right upper part. On direct X-rays, it was found that the mass

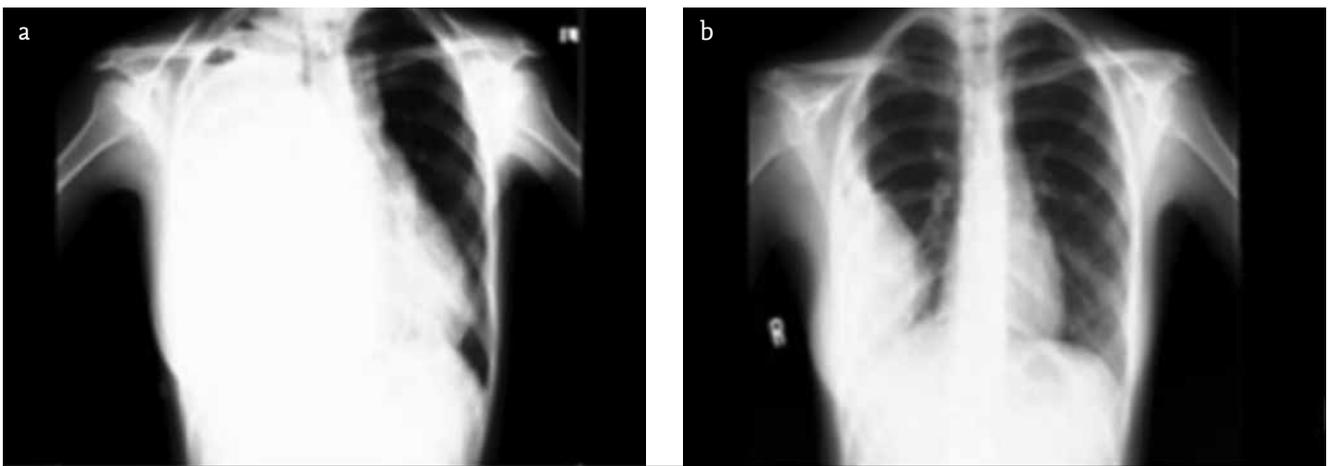


Figure 1. a, b. Appearance of the mass occupying the right hemithorax on postero-anterior lung X-ray before (A) and after (B) six courses of chemotherapy (before surgery)

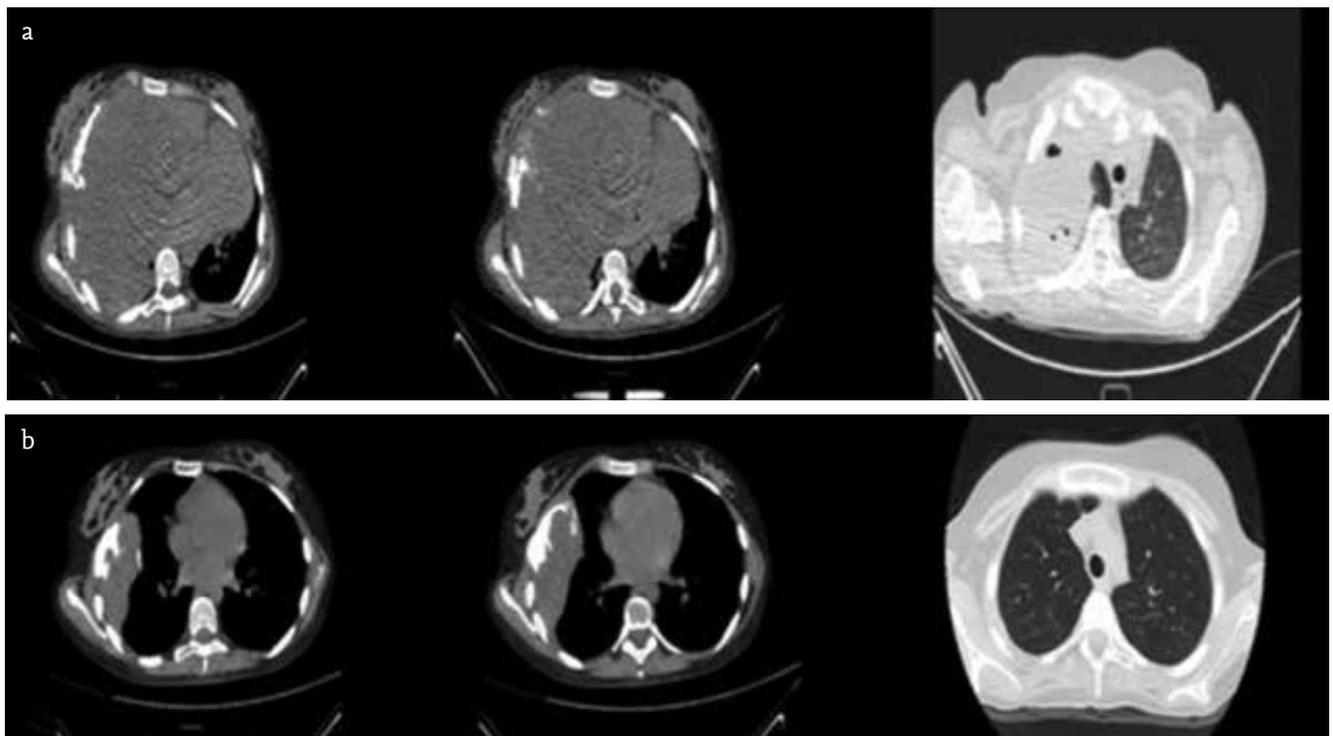


Figure 2. a, b. Thoracic computed tomography axial section: Appearance of the mass, which caused sclerosis in the anterior part of the 4th rib and collapse in the lung before (A) and after (B) six courses of chemotherapy (before surgery)

was reduced starting from the upper medial region of the lung and the parenchyma could be visualized. After the sixth course of treatment, about 60% reduction occurred in the mass (Figure 1). The mass did not change after the seventh course of treatment. The case was discussed in the tumor committee and radiotherapy was recommended with the objective of achieving an intact operation border, though it was not the usual approach. Subsequently, the mass and the anterior faces of the 4th and 5th ribs were excised and corrective surgery for the thoracic wall was not needed. On pathologic examination, the surgical border was found to be intact and 95% necrosis was observed. After surgery, four courses of chemotherapy were given for the objective of consolidation and she has been followed up in a disease-free state for eight months. Verbal informed consent was obtained from the parents of the patient for participation in this study.

Discussion

Osteosarcoma (OS) is the most common bone tumor of childhood. It frequently develops in the metaphyses of long bones in adolescents who are in the growth spurt period. It is most commonly localized in the lower end of the femur, the upper end of the tibia, and in the upper end of the humerus. Flat bone, vertebral, and rib involvement occurs rarely. Rib involvement occurs with a rate of 1-3% in osteosarcoma (5). In our center, primary rib involvement has been observed in one (0.5%) of 190 patients with osteosarcoma in 25 years. Osteosarcoma was found very rarely in various studies conducted with pediatric patients with primary malignant rib lesions (3/17; 2/8) (2, 6).

Primary rib tumors may originate from bone, cartilage, bone marrow, and vascular or nerve structures. The majority of rib lesions in children are malignant. Benign rib lesions originating from bone or cartilage include fibrous dysplasia, enchondroma, aneurismal bone cyst, and eosinophilic granuloma. Malignant rib lesions most commonly include Ewing sarcoma and rarely osteosarcoma, chondrosarcoma, lymphosarcoma and fibrosarcoma (2, 3).

Benign and malignant lesions originating from the rib may show similar clinical properties. The most common symptoms reported in the literature include pain, swelling, dyspnea, and pleural fluid (2, 6). In patients who have pleural fluid as the initial clinical sign, infection is considered primarily in the differential diagno-

sis and oncologic diagnosis may be delayed if careful assessment is not made. In our patient, the initial clinical sign was pain and pleural fluid was found. Thoracic wall tumors generally grow towards the thoracic cavity and may reach a large size when the diagnosis is made. Therefore, imaging studies following history and physical examination are important in the diagnostic approach (2). Usually, direct lung X-rays are obtained primarily and large masses or bone lesions can be detected. Computed tomography provides important information related with the location and size of the mass, bone involvement, invasion to adjacent structures, and metastasis in the lung. Magnetic resonance imaging shows soft tissue components, fluid content, and spinal-epidural invasion of the lesion. Positron emission tomography may be helpful in the assessment of the presence of metastasis (7).

Diagnostic histopathologic examination is important. With this objective, depending on the individual patient and imaging findings, a direct incision may be performed for lesions that have a benign appearance, Trucut biopsy or incisional biopsy may be performed for lesions that have a malignant appearance, and excisional biopsy may be performed for cystic lesions if the lesion is small and has a risk of rupture into the pleura (2, 6, 8).

In benign lesions, the rib should be excised partially. In malignant lesions, en bloc resection is performed as a surgical intervention such that the borders are microscopically intact in order to reduce the local recurrence risk.

Correction of the thoracic wall may be needed depending on the case. If malignant masses without metastasis cannot be excised completely, the best treatment option is primary chemotherapy and subsequent surgical intervention. Radiotherapy may be applied depending on the diagnosis (especially Ewing sarcoma) and the patient (2, 6, 8).

In primary malignant tumors of the thoracic wall, tumor pathology, intact surgical border, complete excision of the mass, and absence of distant metastasis are important for survival (9).

Despite advances in surgical methods, skeletal complications related with rib resection including scoliosis and thoracic wall deformities may cause significant morbidity and these patients should also be monitored in this regard (6).

In children and adolescents presenting with chest pain or respiratory distress, malignant tumors should be included in the differential diagnosis in addition to lung, cardiac, gastrointestinal, and musculoskeletal system problems; metabolic and psychologic causes; trauma; and infection. Although the most common malignant thoracic wall tumors in childhood are Ewing sarcoma and rhabdomyosarcoma, osteosarcoma should also be considered among other causes including bone tumors, soft tissue masses, and hematologic tumors. Survival can be improved with appropriate diagnosis with a multi-disciplinary approach and patient-focused therapies.

Informed Consent: Verbal informed consent was obtained from patients' parents who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - S.B.B., R.K.; Design - S.B.B., R.K.; Supervision - S.B.B., R.K.; Funding - S.B.B., A.I., O.G.; Materials - S.B.B., R.K., A.I., O.G., F.A., F.G., A.C., E.D.; Data Collection and/or Processing - S.B.B., R.K., A.I., O.G., F.A., F.G., A.C., E.D.; Analysis and/or Interpretation - S.B.B., R.K., A.I., O.G., F.A., F.G., A.C., E.D.; Literature Review - S.B.B., A.I.; Writing - S.B.B.; Critical Review - R.K.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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