PNÖMONİYE NEDEN OLAN NADİR BİR OLGU: AİLESEL KOSTA KÖKENLİ OSTEOKONDROMA

A Rare Case of Pneumonia: Familial Costal Osteochondroma

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ÖZET

Giriş: Osteokondrom kemiğin iyi huylu tümörüdür ve soliter olanlarda %1, multiple herediter olanlarda %10 maligniteye yol açabilir. Kinikte daha çok kemikte deformite, sinir ve dokulara bası şeklinde prezente olur.
Olgu sunumu: Çocuk kliniğinde izlenen 4 yaşındaki hastanın bir yaşından beri tekrarlayan pnömoni öyküsü olduğu ve 2 yaşında ailesel multiple osteokondrom tanısı aldığı öğrenildi. Öksürük ve ateş yakınması ile başvuran hastanın akciğer grafisinde pnömonik infiltrasyonu mevcut idi. Fakat kostal osteokondrom ile süperpoze olması nedeniyle hastaya toraks tomografisi çekildi ve kosta kökenli osteokondromların hastanın akciğer parankimine bası yaptığı ve sekonder atelektaziye yol açtığı görüldü.
Sonuç: Kostaların osteokondrom için nadir bir yerleşim yeri olması ve osteokondromun akciğer parankim basısı ile pnömoniye yol açması sebebiyle bu olgu sunulmuştur.

Anahtar kelimeler: *Çocuk; Osteokondrom; Pnömoni*

ABSTRACT

Introduction: Osteochondroma is a benign tumor of the bone, but malign transformation may occur in 1% of solitary cases and 10% of hereditary multiple osteochondromas. The most frequent clinical presentation is due to bone deformities or nerve and tissue compressions.
Case presentation: We report a 4 year old patient followed up at pediatric clinics, with a history of recurrent pneumonia after first year and at the age of 2 he was diagnosed with hereditary multiple osteochondroma. He presented with cough and fever and his chest X-Ray revealed a pneumonic infiltration. As it was superimposed with his costal osteochondroma, a Thoracic CT (computed tomography) was performed which revealed secondary atelectasia due to the costal osteochondroma compressing his pulmonary parenchyma.
Conclusion: Given that ribs are rare localizations for osteochondromas and this osteochondroma has led to pneumonia due to pulmonary parenchymal compression, this case was considered to be worth presenting.

Key words: *Child; Osteochondroma; Peumonia*

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INTRODUCTION

Osteochondroma is frequently presented as a benign tumor of the bone. Its prevalence is 1/50.000 and is more frequent in males (1). It shows an autosomal dominant inheritance pattern. Familial forms, due to mutations in the EXT1 and EXT2 tumor suppressor genes, have been reported (2). It is clinically presented with bone deformities, tissue and nerve compressions. It can be solitary as well as multiple. Malign transformation is reported in 1% of solitary osteochondromas, whereas 10% of multiple osteochondromas (3). This report presents a case of hereditary multiple osteochondroma with secondary pneumonia due to costal osteochondroma compressing pulmonary parenchyma.

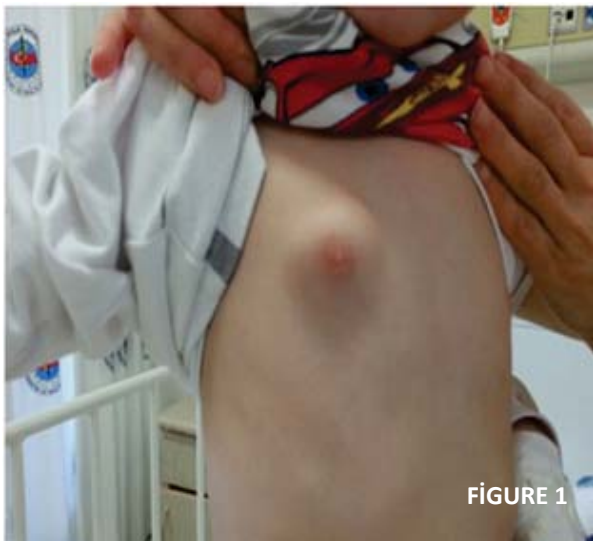
FİGURE 1

CASE REPORT

A 4 year old male referred to our Hospital with a cough and fever lasting for 3 days. In his physical examination his weight was 14kg (3-10p), height 99cm (10-25p), heart rate 98/min, respiratory rate 30/min, he had a wheezing and chest auscultation revealed bilateral rhonchus and coarse crackles. The patient had several palpable mass lesions; one beyond the right breast above the rib measuring about 5x5 cm (Figure 1), one at the lower edge of the left scapula measuring about 3x4cm (Figure 2) and one on the distal radius of right arm measuring about 1x.0.5cm (Figure 3). Examination of other systems was normal. He had a history of recurrent pneumonia after his first year of life. According to his family history his father, sister, grandmother, father's uncle and aunt were diagnosed with osteochondroma. He was 2 years old, when a mass growing on his right costa was noticed. His blood test results are normal but only milk and egg specific Immunoglobulin's were positive. There was an increased density bilateral at the level of fourth costa, on his chest x-ray (Figure 4). With the suspicion of pneumonia and osteochondroma superposition we performed a thoracic computed tomography (CT), which revealed bone lesions concordant with osteochondromas.


FİGURE 2

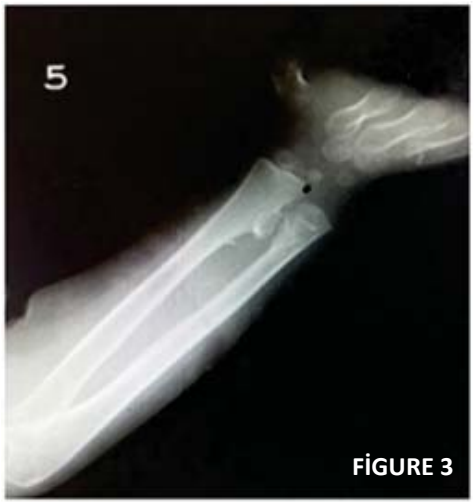

FİGURE 3

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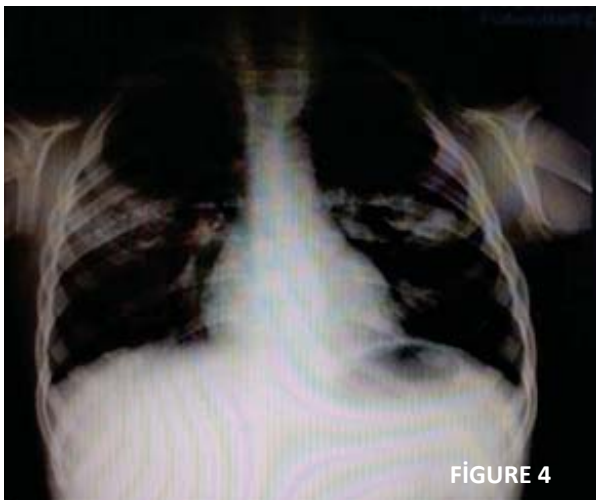

FİGURE 4

These were localized bilaterally on 3. costa anterior arcs measuring about 26x30mm on the right, 17x29 mm on the left and on the left 4. Costa anterior arc measuring about 11x12mm, which extended to the extra pleural soft tissue on the right side and to the pulmonary parenchyma on the left side (Figure 5). We started antibiotherapy for his bronchopneumonia and recurrent bronchiolitis. He recovered and was discharged on the first week of this treatment. The patient didn't need surgical resection and was incorporated in a follow-up every six months for his osteochondromas at the orthopedics polyclinic and for malign transformation at the pediatric oncology polyclinic.

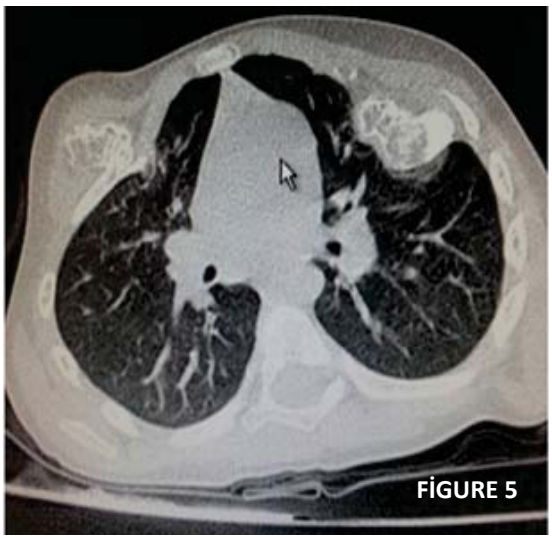

FİGURE 5

DISCUSSION

Osteochondromas are pseudo tumors derived from developmental malformations of the growth plate. These lesions are usually absent at birth, and become clinically and radiologically manifest between ages 2-10 (4). Ostechondromas can be found in almost every bone except the scull. It is mostly derived from the distal femur, proximal and distal humerus. In general 2 % of all osteochondromas are localized at the ribs and are usually slowly growing tumors (5). The first costal osteochondroma case was reported by Twersky in 1975 (6).

Osteochondromas are usually asymptomatic. Rare costal osteochondromas may present with a clinic of pulmonary parenchyma compression. Chest X-ray is usually insufficient in determining the localization. Magnetic Resonance Imaging (MRI) and CT give more reliable information about localization, morphology and the relation between tissues nearby (7). They may grow extra thoracic as well as intra thoracic. This can lead to an overgrown mass on the chest wall. It is reported that; intra thoracic growing osteochondromas may cause pneumothorax, heamothorax and atelectasis whereas some posterior localized osteochondromas may cause spinal compression (8). Malignancy is usually very rare in children but if the lesions are derived from pelvis and proximal femur there is an increased risk. The thickness of the cartilage is more than 2 cm in cases of malign transformation and can be distinguished with CT or MRI (7).

Autosomal dominant inherited osteochondroma is a genetically heterogen disease. Familial forms have been reported due to mutations in EXT1 and EXT2 tumor suppressor genes. The genetic spectrum of the disease may differ between populations and mutation analysis studies in families are offering genetic counseling and prenatal diagnosis. Even though several mutations are defined, the clinical heterogeneity in the same family as well as between different families still needs to be elucidated. It is reported that the EXT1 mutation carriers have a more aggressive disease course, have more exostoses and kondrosarcomas due to degeneration in exostoses are more frequent (3).

Because of economic restrictions, genetic investigations, which could be helpful in diagnosis and prognostic prediction, were not performed.

Costal osteochondroma is a rare cause of hemothorax in children. Duc et al. (9), reported a spontaneous hemothorax case, secondary to a costal osteochondroma in a 15 year old female. The spontaneous rupture of dilated pleural veins which are pressed between exostoses and pleura is thought to be the potential cause of heamothorax.

According to a case report by Cottalorda et al (10). The increased density on the chest x-ray of an 11 year old male, presenting with fever and headache, has been explicated as pneumonia. As the infiltration persisted after 2 months of antibiotic therapy, a thoracic CT was performed and revealed a solitary internal costal osteochondroma, which was later surgically resected.
A ten year old male, reported by Demirhan et al., presented with a mass lesion on his right 3.rib and the soft tissue nearby. The lesion was resected and the pathological report pointed out osteochondroma. No more pathology was detected in the follow up (11).

In conclusion; ribs are rare localizations for osteochondromas and clinically they are painful and have a higher risk of malign degeneration. In case of chest pain and palpable lesions accompanying recurrent pulmonary infections, costal osteochondroma may be an underlying cause and it should be kept in mind that malign transformation can be seen in these tumors. These patients should be put into a close follow up, and surgical treatment should be performed if necessary.

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