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Olgu Sunumu / Case Report

A Rare Cause of Diagnostic Delay: Primary Bone Lymphoma

Tanısal Gecikmenin Nadir Bir Nedeni: Primer Kemik Lenfoması

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ABSTRACT

Primary lymphomas of bone in childhood are not frequently diagnosed. Because of its rarity and little information about the issue, diagnostic delays can ocur in patients who present with complaints related with skeletal system. In this report, a 7-year-old female with bone lymphoma was presented, who initially diagnosed as osteomyelitis and could be diagnosed after 11-month-delay during an investigation of abdominal pain, probably originated from enlarged and conglomerated lymph nodes caused by tumoral infiltration. Clinicians must be aware of symptoms involving skeletal system, such as pain and swelling and malignant neoplasms should be considered in differential diagnosis.

Key words: Non-Hodgkin lymphoma, diagnostic delay, bone, childhood

ÖZET

Primer kemik lenfomaları çocukluk çağında nadir görülen tümörlerdendir. Az görülmesi ve konu hakkında bilgi birikiminin yeterli olmayışı nedeniyle iskelet sistemi ile ilgili şikayetlerle başvuran hastalarda tanısal gecikmeler olabilmektedir. Bu yazıda başlangıçta osteosarkom tanısı alan ve 11 aylık gecikme sonrasında, muhtemelen tümör infiltrasyonuna bağlı büyümüş ve konglomere olmuş lenf nodlarının neden olduğu, karın ağrısı nedeniyle araştırılırken kemik lenfoması tanısı alan 7 yaşında bir kız hasta sunulmuştur. Klinisyenlerin, ağrı ve şişlik gibi iskelet sistemiyle ilgili semptomları dikkate alması ve malign tümörlerin de ayırıcı tanıda düşünülmesi gerekmektedir.

Anahtar kelimeler: Non-Hodgkin lenfoma, tanısal gecikme, kemik, çocukluk çağı.

INTRODUCTION

Primary lymphomas of bone rarely ocur in childhood. Among other malignant diseases of the bone; osteosarcoma, Ewing's sarcoma, chondrosarcoma and malignant fibrous histiocytoma are encountered more frequently than lymphomas[1]. The differential diagnosis of bone tumors is generally difficult as these tumors can be confused with injuries in skeleton in clinical, radiologic and sometimes pathologic aspects. The vast majority of primary lymphomas of bone are non-Hodgkin lymphoma (NHL) and Hodgkin lymphoma accounts for a very small proportion[2]. Lower extremities are the most common sites of presentation for primary NHL of bone although it can arise from any part of the skeleton. Patients with primary NHL of bone commonly present with local bone pain, soft tissue mass or a pathological fracture[3]. Here, we present a patient with bone lymphoma localized in distal femur, diagnosed after 11-month-delay during investigation of abdominal pain.

CASE

A 7-year-old female presented with abdominal pain that have been present for 2 months. The patient had experienced abdominal pain localized around umblicus, lasting a few hours with acompanying nausea and vomiting one or two times in a day. In patient history it was learnt that, the patient had left knee pain 11 months ago and it had been attributed to minor trauma and she had been investigated because of additional swelling in the same localization 6 months ago and diagnosed as osteomyelitis in another center. She had been treated with parenteral antibiotics for 2 weeks and oral antibiotherapy for additional one week with a partial relief in complaints. Familiy history was unremarkable. Physical examination was normal except for minimal swelling in left knee and 2 cm increase in left knee diameter compared to the right. Laboratory findings revealed hemoglobin 11.8 g/dL; hematocrit 33.9%; white blood cell count 11.6 x 10^6 / µL; platelets 550 x 10^6 / µL and in serum biochemistry there was no abnormality. Abdominal ultrasonography and computerized tomography (CT) showed multiple conglomerated lymhadenopathies around celiac truncus, superior mesenteric artery and paraaortic region. Magnetic resonance imaging of the left knee showed a 3.5 × 2.1× 2.0 cm solid mass lesion in metaphysis of the femur. Biopsy from the femoral mass revealed malignant infiltration but no specific tumor was identified. Tru-cut biopsy from the abdominal lymph nodes showed diffuse large B cell lymphoma. Thoracal CT, bone scintigraphy and bone marrow examination showed no additional disease. The patient was treated according to the NHL-BFM Grup protocol. After courses of maintenance therapy consisting of vincristin, actinomycin-D and cyclophosphamide the treatment was stopped in complete remission. The patient is well without tumor recurrence for 18 months and followed in outpatient clinic.

DISCUSSION

Because primary lymphomas of bone in pediatric age group are rare, there is limited information in literature concerning the clinical presentation, diagnostic difficulties and proper treatment of these patients. In a study from Pediatric Oncology Group, the most frequent site of involvement were reported to be femur in 29% among patients with primary NHL of bone and histopathologic examination revealed most of the patients (80%) had large cell lymphoma[4]. Most of the patients with primary lymphoma of bone have disease stage of IE or IIE according to Saint Jude staging system[5]. Although the presented case was diagnosed after an 11-month-delay she was assigned to the stage IIE with femoral and abdominal involvement.

Studies in literature showed that patients with primary NHL of bone have an advantageous survival as in the case of our patient[6,7]. of children with early-stage NHL have a primary site in bone and have traditionally been treated with radiotherapy. As for many pediatric malignancies, NHL is a systemic disease and local therapy alone is not sufficient treatment, even for localized disease. The introduction of multiagent chemotherapy has resulted in a substantial improvement in survival, and more than 90% of children with early-stage NHL are now cured[4]. Risks of delaying in diagnosis of lymphoma can result in life threatining complications such as; advanced stage disease, difficulties in management of tumor, tumor lysis syndrome and electrolyte imbalances. Fortunately our patient was diagnosed after an 11-month-delay and she was in the stage IIE with femoral and abdominal involvement and cured after appropriate treatment.

Clinicians should keep in mind the wide range of skeletal neoplasms that may ocur in childhood especially in the presence of symptoms related with musculo-skeletal system[8-10]. Musculoskeletal

pain is one of the most common presenting symptoms at the pediatric outpatient clinics. Etiology ranges from benign conditions to serious ones requiring prompt attention. Bony complaints are well-described symptoms of malignant diseases if they present together with other systemic signs such as fever, weight loss and loss of appetite. In the absence of systemic signs, musculoskeletal presentations of malignancies may not be easily recognised and pose as a diagnostic challenge even to the most experienced clinicians. In a sutudy, the prevalence of bony complaints as the first presentation of childhood haematological malignancy was reported as 14.2% and the diagnosis of malignancy was challenging with a high rate of undiagnosed cases at 71.4% at first visit[11]. Our patient was initially diagnosed as osteomyelitis in another center with the complaints of knee pain and swelling and could be diagnosed as NHL after 11-month-delay during an investigation of abdominal pain, probably originated from enlarged and conglomerated lymph nodes caused by tumoral infiltration.

This case highlights the frequent delay in the diagnosis of primary bone lymphoma, and the fact that symptoms can be attributed to incidental trauma. The case also illustrates that symptoms involving skeletal system such as bone pain and swelling in joints in children and adolescents, should not be underestimated and warrant further investigation.

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