

Sarkoidozda Kemik İliği Tutulumu, Olgu Sunumu

Bone Marrow Involvement Of Sarcoidosis, Case Report

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Özet

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Sarkoidoz sistemik granümatöz bir hastalıktır ve en sık akciğerleri tutar, nadir olarak kemik iliği tutulumu ile seyredebilir. Kemik iliği tutulumu olan hastalarda hematolojik anormallikler saptanabilmektedir. 47 yaşında bel ağrısı, yürümede güçlük ve çift taraflı alt ekstremitelerde güçsüzlük şikayetleri olan bayan hastada bisitopeni ve L2-L3 vertebrada lezyon saptandı. Hasta opere edildi. Lezyonun patolojik incelemesi sonucu sarkoidoz tanısı kondu. Yapılan diğer incelemeler sonucunda Akciğerde sarkoidoz tutulumu saptandı. 74 yaşında halsizlik ve yorgunluk şikayetleri olan bayan hastada yapılan tetkiklerde pansitopeni saptanması üzerine kemik iliği biyopsisi yapıldı. Kemik iliğinin patolojik olarak incelemesi sonucunda sarkoidozla uyumlu non-kazeifiye granülom saptandı. Yapılan diğer incelemelerde başka bir tutulum saptanmadı. Burada nadir görülen kemik iliği sarkoidozu saptanan iki olguyu sunduk.

Anahtar Kelimeler: Sarkoidoz, Kemik iliği tutulumu, Granülom

Abstract

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Sarcoidosis is a systemic granulomatous disorder and most often involve in lung, rarely may present with bone marrow involvement. Hematologic anomalies can be determined in patients with bone marrow involvement, but it is not a rule. A 47-year-old woman patient having complaints from backache, difficulty in walking and weakness in bilateral lower extremities admitted to our hospital we detected bicytopenia and lesion in L2-L3 vertebrae. Patient was operated. After pathological investigation of lesion we diagnosed bone marrow involvement of sarcoidosis. Lung and the other organs systems were normal. A-74-year-old woman patient having complaints from malaise and fatigue admitted to our hospital, we detected pancytopenia and vertebral lesion than we applied bone marrow biopsy from the lesion. After bone marrow biopsy we diagnosed bone marrow involvement of sarcoidosis. In other investigations lung involvement with mediastinal lymphadenopathy was detected and the other organ systems were normal. Here we presented two cases diagnosed with bone marrow involvement of sarcoidosis which is seen rarely.

Keywords: Sarcoidosis, Bone Marrow Involvement, Granuloma

Introduction

Sarcoidosis is a systemic granulomatous disorder that most often involve lungs (> % 90) but can involve all organs and systems. Although hematologic anomalies can be seen in patients with bone marrow involvement, but it is not a rule. Though sarcoidosis is rarely seen in cytopenia etiology, it is a clinical circumstance that shouldn't be ignored and is seen seldomly.

Case 1

A-47-year-old woman patient having complaints from backache, difficulty in walking and weakness in bilateral lower extremities admitted to hematology clinic. The patient's vital parameters were normal. No pathology is diagnosed in physical examination. In the patient's laboratory investigation: Leucocyte: 4000/mm³, Hemoglobin: 8,8 g/dl, Platelet count: 237.000/mm³ were found. In biochemical surveys, Glucose:76 mg/dl, Ure:25 mg/dl, Creatinine:0,7 mg/dl, ALT:41 IU/L, AST:53 IU/L, Na:140mmol/L, K:4,1 mmol/L, CRP:19 mg/dl, Erythrocyte sedimentation rate:13 mm/h, Ferritine, Vitamin B12 and Folic acid levels were normal. In Lumbar MRI after contrast administration we detected lesion in L2-L3 vertebrae that cause environmental and heterogeneous contrast involvement (Figure 1). L2 portral and L3 total laminectomy operation was applied to the patient and during the operation, biopsy was perform from the lesion which was seen in MR. In pathologic investigation of biopsy material, a large number of noncaseating granulomas constituting groups from place to place and locally necrosis in bone marrow were detected and bone marrow involvement of sarcoidosis was diagnosed (Figure 2). Lung and the other organs systems were normal.

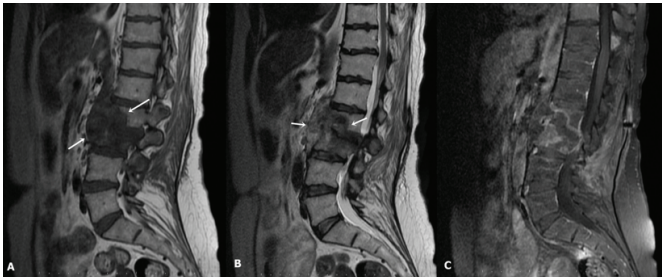


Figure 1: Sagittal T1A (A), sagittal T2A (B) ve sagittal kontrastlı sekanslarda (C) L2-3 vertebra korpuslarını ve intervertebral disk mesafesini tutan, periferik kontrast tutulumu gösteren sarkoidoz ile uyumlu granümatöz iltihabi reaksiyon izleniyor.

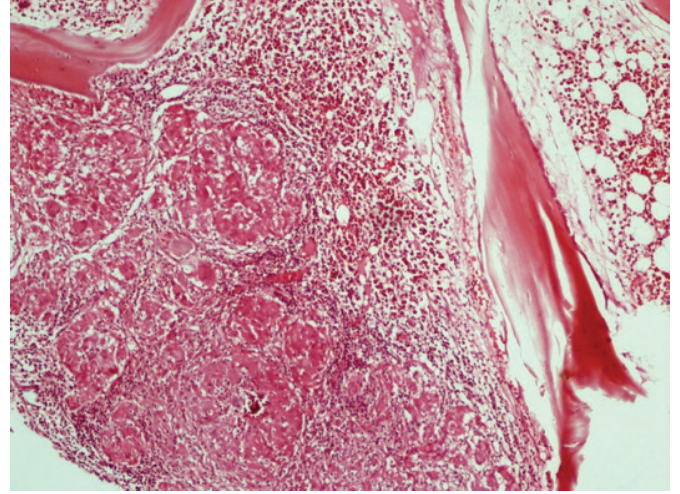


Figure 2: Kemik trabekülleri arasında izlenen kemik iliği mesafesinde, çok sayıda epitelioid histiositten oluşan, gruplar halinde nonkazeifiye granülom yapıları görülüyor. HE, x100

Case 2

A-74-year-old women patient complaining from malaise and fatigue applied to the hospital. The patient's vital parameters were normal. In physical examination, no pathology was detected apart from hepatosplenomegaly. Leucocyte: 2800/mm³, Hemoglobin: 10,1 g/dl, Platelet count: 53200/m³ were found. As biochemical, Glucose:121 mg/dl, Ure:20 mg/dl, Creatinine:0,9 mg/dl, ALT:7 IUL, AST:24 IU/L, Na:134 mmol/L, K:3,8 mmol/L, CRP:11 mg/dl, Erythrocyte sedimentation rate:44 mm/h were detected. Ferritine, Vitamin B12 and Folic acid levels were detected as normal. Bone marrow biopsy was applied to the patient. In pathologic investigation of the biopsy material, noncaseating granulomas forms that consisted of in different dimention from epithelioid histiocyte stained positive with CD68 in light hypocellular according to the age and bone marrow involvement of sarcoidosis was diagnosed (Figure 3). In other investigations lung involvement with mediastinal lymphadenopathy was detected and the other organ systems were normal.

Discussion

Sarcoidosis is a multisystemic granulomatous disorder that most often involve in lung, lymph node, spleen, liver, eye and skin. Although sarcoidosis involve most often in lungs it can involve in all organs, isolated lung external sarcoidosis frequency was detected in patients lower %101. In our cases only the bone marrow was affected. Granulomas is rarely detected

in bone marrow and it should be considered infectious circumstances such as tuberculosis, malignancy and sarcoidosis in the differential diagnosis². Whereas anjiyotensin converting enzyme (ACE) levels increase in sarcoidosis it is not pathognomonic³.

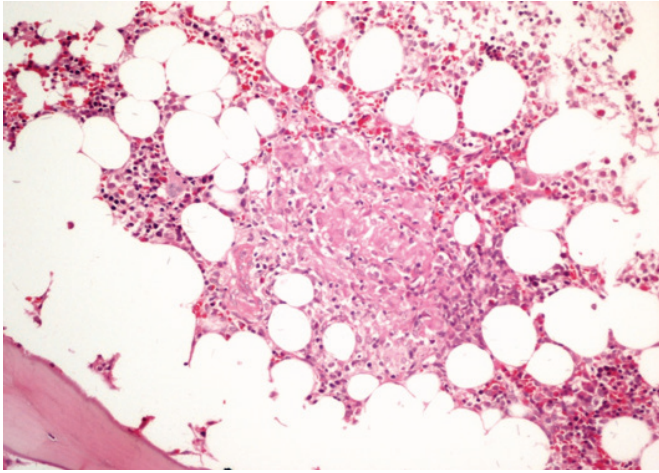


Figure 3: İzlenen her seriye ait hematopoetik hücreleri içeren kemik iliği mesafesinde, epitelioid histiositlerden oluşan non-kazeifiye granülom yapısı görülüyor. HE, x200.

Definite diagnosis of sarcoidosis is done with biopsy and recently PET-CT has given so beneficial conclusions in sarcoidosis diagnosis^{4,5}. In an investigation of Brockens de Hugo and his friends; 57 granulomas were detected in 9641 bone marrow biopsy and %20 of these granulomas was determined as sarcoidosis⁶. Bone marrow involvement in sarcoidosis doesn't conclude with defect everytime in hematologic parameters and owing to the this fact, bone marrow involvement in patients who haven't defects in hematologic parameters may be overlooked⁷. In both cases, due to unexplained anemia, we decided applying bone marrow biopsy and a great number of noncaseating granulomas was detected in consequence of the biopsy. Immunosuppressive drugs such as; Azathioprine, corticosteroids, infliximab and methotrexate are used in the treatment of sarcoidosis^{8,9,10}. In our cases we used corticosteroids in the treatment. The point we want to emphasize in our article is to take into consideration bone marrow involvement in revealing unidentifiable symptoms even though there aren't any anomalies in hematologic parameters.

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