Sinus Histiocytosis with Septic Embolism and Infective Endocarditis

Septik Emboli ve Enfektif Endokardit ile Seyreden Sinüs Histiositoz Olgusu

Rosai Dorman Hastalığı / Rosai Dorfman Disease

Elif Torun Parmaksız[1], Ergün Parmaksız[2]
[1]Dr Lütfi Kırdar Kartal Eğitim ve Araştırma Hastanesi, Göğüs Hastalıkları Kliniği,
[2]Başkent Üniversitesi İstanbul Hastanesi, Nefroloji Kliniği, Türkiye

Özet

Rosai Dorfman hastalığı olarak da bilinen benign histiositoz ilk olarak hastalığa adını veren Rosai ve Dorfman adlı araştırmacılar tarafından tanımlanmıştır ve günümüze kadar birkaç olgu bildirimi yapılmıştır. Lenfadenopatinin nadir görülen bir nedenidir. Bu yazıda bir sinüs histiositozu olgusu klinik, laboratuar ve biyopsi bulguları ile tanımlanmış ve daha önce bildirilen olgularla karşılaştırılmıştır. Olgumuzu diğerlerinden ayıran özellikler nadir görülen bir tanı oluşunun dışında sadece aksiler lenf bezlerinin tutulmuş olması, böbrek yetmezliği, enfektif endokardit ve septik embolinin eşlik etmesidir.

Anahtar Kelimeler

Sinüs Histiositozu; Rosai Dorfman Hastalığı; Septik Emboli; Enfektif Endokardit

Abstract

Rosai and Dorfman were the first to describe benign histiocytosis, also known as Rosai Dorfman disease. Since then, several case reports have been reported. This entity is a rare cause of lymphadenopathy. The clinical, laboratory, and biopsy findings of a sinus histiocytosis case is discussed and compared with previously reported cases. The distinguishing features of this case are isolated involvement of the axillary lymph nodes, accompanying chronic renal failure, infective endocarditis and septic embolism besides the rarity of the diagnosis of sinus histiocytosis.

Keywords

Sinus Histiocytosis; Rosai Dorfman Disease; Septic Embolism; Infective Endocarditis

DOI: 10.4328/JCAM.1128 Received: 10.06.2012 Accepted: 05.08.2012 Printed: 01.11.2015 J Clin Anal Med 2015;6(6): 844-846
Corresponding Author: Elif Torun Parmaksız, Dr. Lütfi Kırdar Kartal Eğitim ve Araştırma Hastanesi, Göğüs Hastalıkları Kliniği, Cevizli, İstanbul, Türkiye.
T.: +905062425356 E-Mail: dreliftorun@yahoo.com

Introduction

Rosai and Dorfman described a clinicopathological entity called "sinus histiocytosis with massive lymphadenopathy", better known as Rosai-Dorfman disease in 1969 (1). The first four cases reported were all children, with a disease characterized by massive lymphadenopathy, fever, hypergamaglobulinemia, increased erythrocyte sedimentation rate and leucocytosis [1-3]. Mediastinal, axillary and inguinal lymph nodes can also be affected and extranodal sites are involved in about 25% to 40% of cases [3].

The benign and self-limited course of the disease distinguishes it from malignant reticuloendotheliosis [2]. No specific treatment is required [4].

In this report we describe a case of sinus histiocytosis accompanied by infective endocarditis and septic embolism; to our knowledge, this is the first such case reported in literature.

Case Report

A 66 year-old female patient presented with a history of relapsing fever for 2 months. There had been mild cough and slightly disturbing chest pain. She had a history of chronic renal failure for 7 years and was on routine hemodialysis 3 times weekly. She was found to have severe anemia and had been receiving transfusions for 5 months. Her past history revealed a gastrointestinal system malignancy and chemotherapy 30 years ago. She had no other comorbidities. She had been thoroughly evaluated for anemia to rule a malignancy by Nephrology department in previous month. The abdominal computed tomography(CT), gastroscopy and colonoscopy remained nondiagnostic. The thoracic CT was found to be normal except for nonspecific millimetric parenchymal nodules. Bone marrow biopsy demonstrated hypercellular bone marrow. According to transthoracic echocardiography ejection fraction(EF) was 58% and there was mild mitral regurgitation and tricuspid regurgitation. The patient had a history of betalactam group antibiotic use. On admission, thoracic CT demonstrated diffuse, multiple, bilateral pulmonary parenchymal nodules of various sizes (Figure 1). The radiological differential diagnosis included infection or metastatic malignancy. Bronchoscopically no endobronchial lesion was detected, bronchial washings were negative for bacteria, fungi, acid-fast bacilli or atypical cells. She had a painless palpable lymph node of 1 cm on left axillary region. The excisional biopsy of the lypmh node revealed dilated sinuses filled with histiocytes with abundant pale eosinophilic cytoplasm. The histopathological diagnosis diagnosis was sinus histiocytosis with focal lymphoid proliferation (Figure 2).

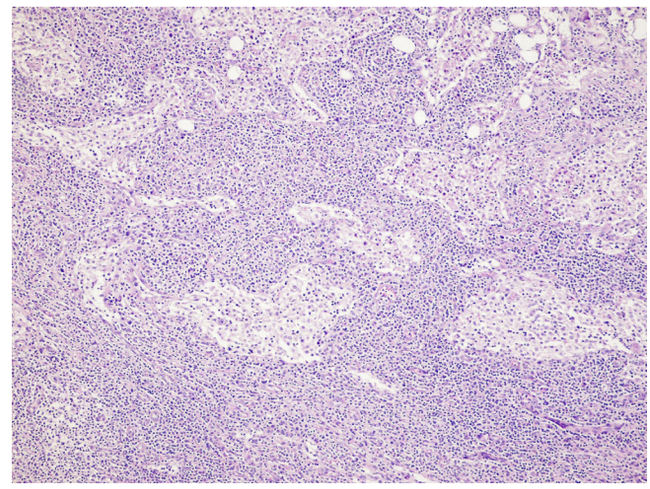


Figure 2. Lenf nodunun patolojik kesiti

Meanwhile the clinical condition worsened by high fever, fatigue, arthralgia, nausea and vomiting . Laboratory findings were as follows: hemoglobin:7.77g/dl, hemotocrit:22.1 %, mean corpuscular volume:88.6 fL, leucocytes:8830/ul, plataelets:181000/ul, C-reactive protein:313 mg/l, eritrocyte sedimentation rate:124/hour, blood urea nitrogen:42 mg/dl, creatinine:4.81 mg/dl, sodium:134 mmol/l, potassium:4 mmol/l, albumin:2.8 g/dl, other biochemical tests were within normal limits. Connective tissue markers, c-ANCA and p-ANCA were negative. Another thoracic

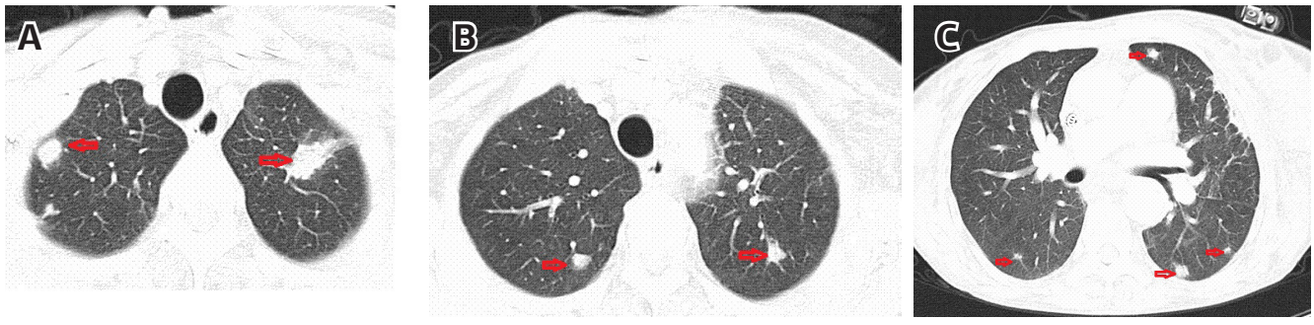


Figure 1. Bilateral akciğerlerde dağınık yerleşimli çeşitli boyutlarda nodüler lezyonlar (A,B,C).

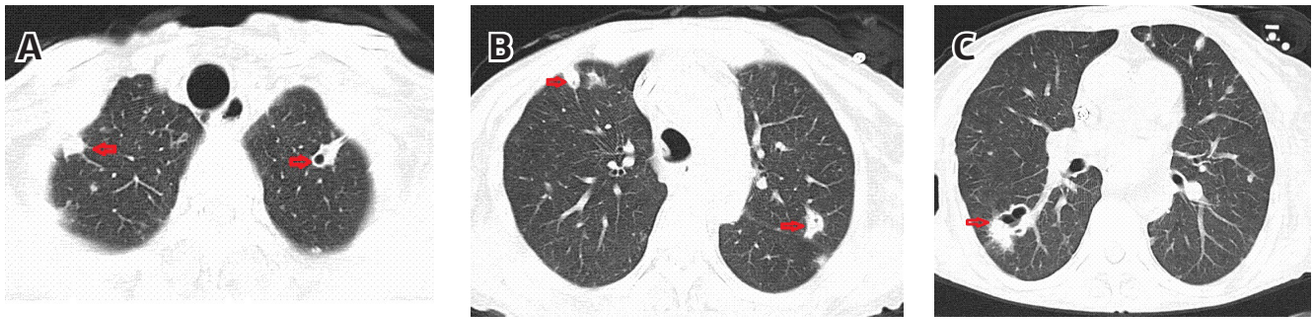


Figure 3. Akciğerlerdeki nodüllerde kavite oluşumları (A,B,C)

CT was taken and pulmonary nodules were found to increase in size and number, with cavities in some of them (Figure 3). Bronchoscopy was repeated and culture of the bronchial washings revealed methicillin sensitive Staphylococcus aureus. As clinical, radiological and microbiological findings pointed out septic embolism, echocardiography was repeated. Transthoracic echocardiograhy was suspectful for a 1,6x1 cm vegetation on atrial side of the tricuspid valve attached to the septal leaflet. Transesophageal echocardiography demonstrated 3 vegetations; first one 2x0.9 cm, mobile, on the catheter, extending into the superior vena cava; second 3x0.8 cm, mobile, originating from the tricuspid valve annulus; and third 0.8 cm, mobile, on right atrial lateral wall.

The patient was put on appropriate antibiotics with diagnosis of infective endocarditis and septic embolism, followed by surgery. All the clinical picture resolved and the patient was free of symptoms.

Administration of antibiotics prior to the exact diagnosis seem to mask the severe clinical picture of septic embolism and prevent bacterial growth on cultures. Due to their location, the vegetations could not be visualized by transthoracic echocardiography and the diagnosis remained obscure. The evaluation of fever of unknown origin requires careful and close follow-up accompanied by semi-invasive or invasive investigations, especially in patients with risk factors such as indwelling catheters.

Discussion

Sinus histiocytosis is a diffuse, lymphoproliferative disorder, mostly effecting children or young adults, although patients in elder ages have also been described [3]. Our case seems to be among the oldest cases described.

The disease has no known gender, ethnic or socieconomic predilection [5] . The etiology is generally unknown, although some cases have been linked to Ebstein Barr virus, cytomegalovirus, Brucella, Klebsiella or human herpes virus 6 [4-6]. Associated symptoms and signs may differentiate according to the size, site and number of lymph nodes involved or may be constitutional, such as fever and weight loss [3,4]. The affected extranodal sites may include skin and soft tissues, upper respiratory tract, orbit, testicle, kidney, thyroid, small bowel, breast, and bone [7].

Although early descriptions concluded that nearly every case was marked exclusively by cervical lymph node involvement, other organ systems may be affected. More than 90% of cases with sinus histiocytosis have cervical lymphadenopathy [8]. Axillary lymph nodes are not commonly involved in sinus histiocytosis. In evaluation of 2250 axillary lymph nodes removed from 487 autopsies, Tsakraklides et al found the incidence of sinus histiocytosis as 1.7% [9]. Another distinguishing feature of our case is involvement of axillary lymph nodes without cervical lymph node invasion.

Based on the fact that the laboratory findings include anemia, leukocytosis and increased ESR [3] , findings of our patient were consistent with literature.

In literature, cases of pleural effusion, mediastinal lymph node calcification, airway disease, cystic and interstitial lung disease [5,10] have been reported, however no association between infective endocarditis or septic pulmonary embolism has been described. As the severity of clinical picture was hidden by previous antibiotic use, the diagnosis of sinus histiocytosis was reached even before the underlying disease.

Considering the benign and self-limiting course of the disease, treatment is not required in most cases. Treatment is generally reserved for life-threatening complications or disfiguring enlarged lymph nodes. Corticosteroids [10] , chemotherapeutic agents such as etoposide, methotrexate, 6-mercaptopurine [11] , cyclophosphamide, rituximab [12], either alone or in combination, are among therapeutic agents used so far in treatment of Rosai Dorfman disease. Surgical excision and radiotherapy have also been tried [4] .

Some case reports in our country reveal a case with axillary, scalene and hilar lymphadenopathy [13] and another case with mediastinal and hilar lymph nodes [14] .

In conclusion, key distinguishing features of this case are isolated involvement of the axillary lymph nodes, accompanying chronic renal failure infective endocarditis and septic embolism besides the rarity of the diagnosis of sinus histiocytosis. This disease should be kept in mind in differential diagnosis of enlarged lymph nodes when no other predisposing disease coul be found.

Competing interests

The authors declare that they have no competing interests.

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How to cite this article:
Parmaksız ET, Parmaksız E. Sinus Histiocytosis with Septic Embolism and Infective Endocarditis. J Clin Anal Med 2015;6(6): 784-6.