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OLGU SUNUMU CASE REPORT

A case with eosinophilic lung diseases: chronic eosinophilic pneumonia or Churg-Strauss syndrome?

Eozinofilik akciğer hastalığı olan bir olgu: Kronik eozinofilik pnömoni mi; Churg-Straus sendromu mu?

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ABSTRACT

Churg-Strauss syndrome is a rare disorder characterized by hypereosinophilia and systemic vasculitis which usually occurs in patients with asthma and allergic rhinitis. We, here, presented a case with Churg-Strauss syndrome with no extra-pulmonary presentation despite involvement in peripheral nervous system as in the form of peripheral mononeuropathy in femoral nerve.

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Key words: Chronic eosinophilic pneumonia, Churg-Strauss syndrome, eosinophilia, vasculitis, peripheral mononeuropathy

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ÖZET

Allerjik granülomatöz anjitis veya Churg-Strauss sendromu hipereozinofili ve sistemik vaskülit ile seyreden astım ve allerjik rinitli olgularda ortaya çıkan nadir bir hastalıktır. Burada periferik sinir sisteminde femoral sinirde periferal mononöropati şeklinde ekstrapulmoner tutulumu olan Churg-Strauss sendromu tanılı bir olgu sunulmuştur.

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Anahtar kelimeler: Kronik eozinofilik pnömoni, Churg-Strauss sendromu, eozinofili, vaskülit, periferik mononöropati

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Allergic and granulomatous angiitis or Churg-Strauss syndrome (CSS) is a rare disorder characterized by hypereosinophilia and systemic vasculitis which usually occurs in patients with asthma and allergic rhinitis[1-5]. Vasculitis commonly affects the lung, skin, peripheral nerves, heart and gastrointestinal tract. The clinical features of systemic vasculitis depend on the organs involved and, in turn, organ involvement is largely influenced by the size of the

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affected blood vessels[1-5]. The diagnostic work-up should be tailored to the clinical situation and geared toward a tissue or angiographic diagnosis, bearing in mind that the findings from these studies are not always pathognomonic[1-5]. We present a case who was diagnosed as CSS and has extrapulmonary system involvement without any clinical signs and symptoms which leads a difficulty in differential diagnosis.

CASE REPORT

A 50-year-old non-smoker man presented with shortness of breath, dry cough, chill and fever lasting for 10 days. He had been admitted to another health-care centre for these symptoms and had been prescribed cefuroxime-axetyl 500 mg, tablet, twice daily for 10 days. Because of progression in the symptoms, he applied to our clinic. The patient had been followed with diagnosis of asthma for 7 years in our clinic. His first asthma attack had occurred just after nasal polypectomy. He had mild persistent asthma and been taking 250 µg fluticasone daily for the last 6 months. He had no previous use of montelukast or zafirlukast. There was no history of cardiac, skin, neurological or gastrointestinal symptoms on admission. On physical examination, results of vital signs were as follows: blood pressure: 130/80 mmHg, body temperature: 36.8°C, respiration rate: 20/minute, pulse rate: 96/minute and rhythmic. Bilateral rare rhoncus were detected at the lung. The rest of the physical examination was unremarkable.

Results of laboratory investigations were as follows: haemoglobin 13 g/dL, white blood cell count 18.6 x 10^3/µL, and platelet count 543 x 10^3/µL. Sedimentation rate was 90 mm/hour. Percentage of eosinophils in white cells was 36%. On admission total eosinophils count was 4100/mm^3 which increased to 6400/mm^3 during follow-up period. Basic biochemical profile was in normal range. Serum total IgE level was increased (225 kIU/L). Serum complements levels (C3 and C4); IgM and rheumatoid factor levels were in normal range whereas an increased levels in C-reactive protein (CRP), IgA and IgG were detected. Pulmonary function test showed an obstructive pattern (FEV_1: 2.03 L 64% predicted; FVC: 2.88 L 74% predicted; FEV_1/FVC: 68%). Stool examinations for ova and parasites were negative. Repeated sputum smear was negative for acid-fast bacilli (AFB) and sputum culture results were also negative. Skin prick test with common inhaled allergens including house dust mites, cockroaches, moulds, cat and dog and pollens was negative.

Chest X-ray revealed bilateral non segmental and patchy infiltrates (Figure 1A). Bronchoscopic examination showed normal airways. Computed tomographic (CT) scan of the chest demonstrated septal thickness and ground glass appearance on the both upper lobs and middle lob (Figure 2A). He had also mucosal thickening on CT scan of paranasal sinuses (Figure 3). A minimal pericardial effusion was also noticed

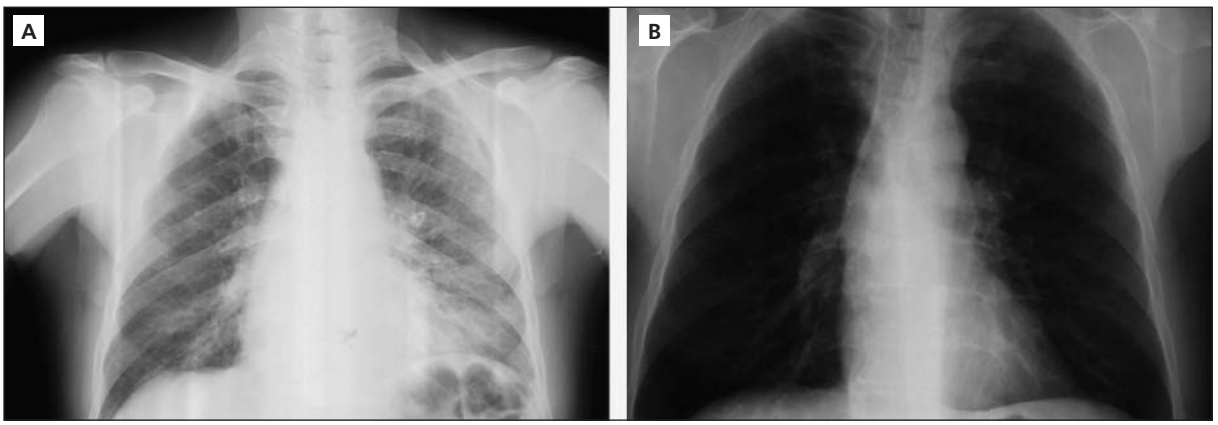

Figure 1. (A) The chest X-ray of the case on admission (B) after corticosteroid treatment.

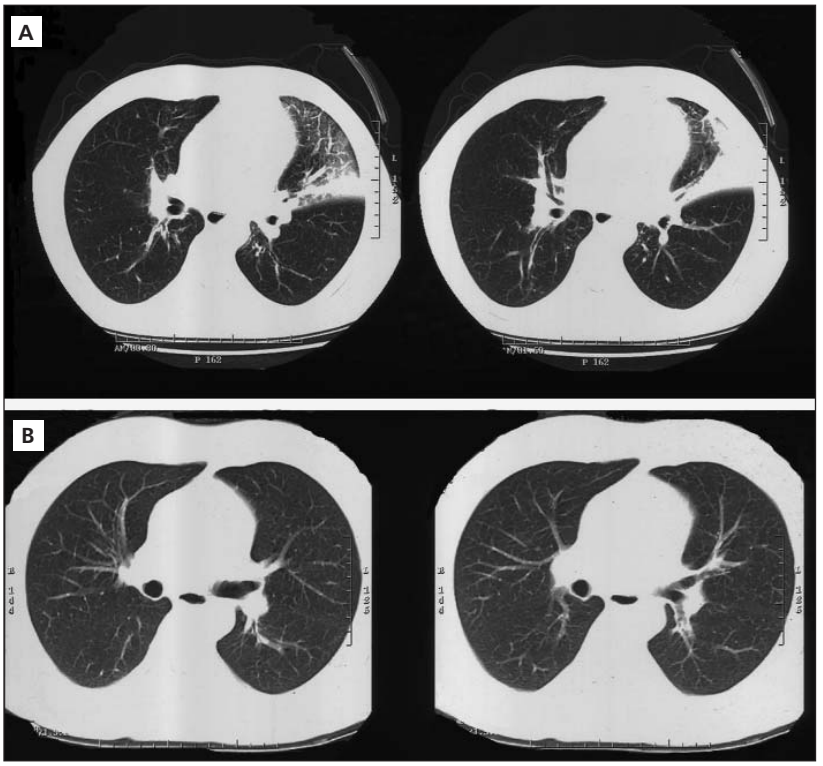

Figure 2. (A) The computed CT scan of the chest on admission (B) after corticosteroid treatment.

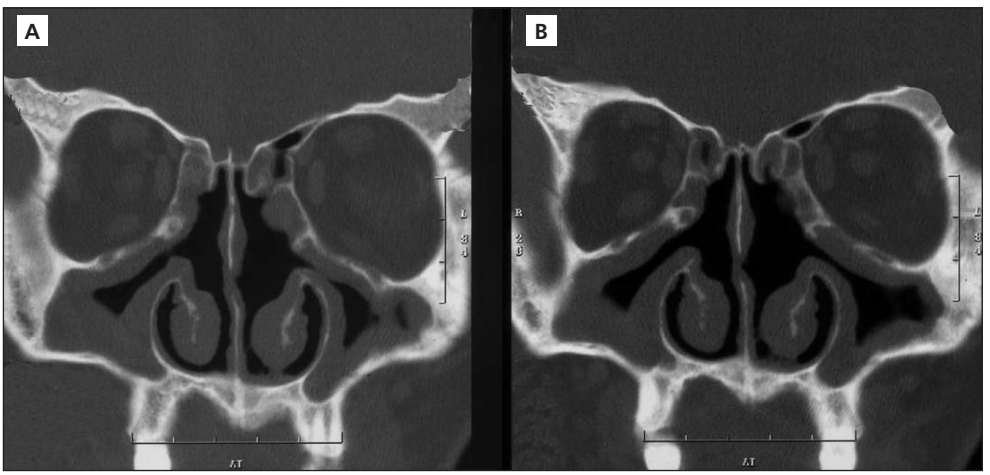

Figure 3. The computed CT scan of paranasal sinuses.

in CT scan of the chest. BAL and bronchial brush microscopy showed marked eosinophilia. Transbronchial lung biopsy demonstrated focal groups of eosinophil leucocytes in mixed inflammatory cells around the suspicious vascular structure. BAL culture was negative.

Although on the basis of persistent peripheral eosinophilia, infiltrates on chest X-ray, high percentage of eosinophils in the BAL and bronchial brush microscopy, and presence of asthma without any extrapulmonary manifestation, initially chronic eosinophilic pneumo-

nia (CEP) was accepted as the most likely diagnosis. However, as CT scan of the chest revealed minimal pericardial effusion, further investigations of heart as well as other systems was performed for the possibility of a diagnosis of CSS despite absence of extrapulmonary system symptoms and findings. Echocardiography showed minimal pericardial effusion, and no diagnostic procedure was able to perform in order to document the etiology of effusion. On further systemic evaluation, there was no pathologies on skin, eye and gastrointestinal system evaluations. Neurological examination showed a hypoesthesia at the side of right thigh despite no complaint reported by the patient. Electroneuromyography findings were compatible with damage of nerve of cutaneous lateral femoral.

According to the American College of Rheumatology criteria for the classification of CSS, as the patient met five out of 6 criteria (eosinophilia > 10% on differential white blood cell count, asthma, paranasal sinus abnormality, non-fixed pulmonary infiltrates on chest X-ray, and mononeuropathy), he was diagnosed as CSS[2]. Oral corticosteroid treatment (methylprednisolon) was commenced 1 mg/kg/day for the treatment. At the first week of the treatment, a dramatic decrease in blood eosinophil count was observed as well as a significant improvement in symptoms and chest X-ray and CT scan of the chest findings (Figure 1B and 2B). On the following weeks, corticosteroid dose was gradually tapered according to clinic and laboratory findings. After 1 year of corticosteroid treatment, the medication was stopped and now he has no relapse of the disease.

DISCUSSION

We, herein, presented a case with CSS with no extrapulmonary presentation despite involvement in peripheral nervous system as in the form of peripheral mononeuropathy in femoral nerve. CSS and CEP are eosinophilic lung disorders with a common respiratory presentations and chest findings[2-7]. The initial presentations, clinical findings of chest and X-ray appearance are quite similar in both diseases. Both diseases have a subacute presentation and constitutional symptoms including fever, malaise and weight loss which have been reported in both genders and all ages. History of asthma, allergic rhinitis, and nasal polyps, and peripheral eosinophilia are common characteristics of both CSS and CEP. However, the main difference between CSS and CEP is the involvement of extrapulmonary systems such as skin, heart, and nervous system in CSS during vasculitic phases whereas CEP doesn't have major extrapulmonary manifestations[2-7]. CEP also has been suggested to be early manifestations of CSS[8-10]. In our case, although the initial clinical presentation was reflecting the diagnosis of CEP, incidentally discovering of pericardial effusion causing for further evaluation of the patient for the probability of extrapulmonary involvement finally led us to diagnose the subject as CSS.

Neurologic system involvement is one of the characteristic features of the vasculitic stage of CSS and occurs approximately in 75% of the subjects[11,12]. Mononeuritis multiplex is the typical neuropathic manifestation of acute systemic vasculitis. Other neuropathic manifestations included peripheral polineuropathy or mononeuropathy. Involvement of the central nervous system was reported to be rare. Particular mention should be made of the frequent devastating involvement of the nerves which produces severe tingling, numbness, shooting pains and severe muscle wasting/power loss in the hands or feet[11,12]. The subjects with CSS and having neuropathic manifestation had been reported to have moderate to severe symptoms[11]. However, the patient presented here was asymptomatic on admission and during follow-up period, although he had peripheral mononeuropathy.

In this case, we didn't have a positive biopsy finding for CSS. However, transbronchial biopsy revealed focal groups of eosinophil leucocytes in mixed inflammatory cells around the suspicious vascular structure. A biopsy pro-

vided from sinuses was also not available as the patient did not accept this procedure. Furthermore, the patient also met the 5 out of 6 criteria of American College of Rheumatology for the classification of CSS, (eosinophilia > 10% on differential white blood cell count, asthma, paranasal sinus abnormality, non-fixed pulmonary infiltrates on chest X-ray, and mononeuropathy), so, he was diagnosed as CSS .

In conclusion, CSS should always be seek in the differential diagnosis of CEP as asymptomatic affected systems could be possible. As the treatment and the course of both diseases differ, at least detailed neurologic and skin evaluations should be performed in cases presented like CEP.

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