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.

(Asthma Allergy Immunol 2009;7:74-78)

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

Received: 13/03/2009 • Accepted: 13/04/2009

ÖZET

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...
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-axetil 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 10³/µL, and platelet count 543 x 10³/µL. Sedimentation rate was 90 mm/hour. Percentage of eosinophils in white cells was 36%. On admission total eosinophils count was 4100/mm³ which increased to 6400/mm³ 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 (FEV₁: 2.03 L 64% predicted; FVC: 2.88 L 74% predicted; FEV₁/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 lobes 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.](image-url)
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 presentati-
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.

REFERENCES