Churg-Strauss syndrome is a rare disease with systemic vasculitis and hypereosinophilia. It is accompanied by allergic rhinitis and asthma. Herein, we present a case of Churg-Strauss syndrome in a family medicine outpatient clinic. This report aimed to emphasize that if patients diagnosed with asthma have cough and/or hemoptysis that cannot be controlled despite interventions such as inhalation, corticosteroid therapy, montelukast treatment, and anti-histamine therapy, vasculitic diseases involving the lung may be considered.

**Keywords:** Churg-Strauss syndrome, vasculitis, vasculitic syndrome involving the lung, family medicine

### Case Report

A 51-year-old woman was referred to the family medicine outpatient clinic in our hospital. She had a cough history spanning the past 1 month and complaint of newly developing bloody sputum. She was receiving inhaled corticosteroid therapy and montelukast + antihistamine therapy for asthma. However, she had a dry cough that persisted despite using two cycles of antibiotics for the past several months. Systemic examination revealed no pathology. Her vital findings were stable, but she was febrile. Rallies were observed in the lower left lobe. Saturation was found normal.

Laboratory examinations of the patient revealed the following data: peripheral smear leukocyte 9,000/mm³, eosinophil 1,500/mm³, hemoglobin 10.7 g/dL, hematocrit 34%, platelet count 413,000/mm³, C-reactive protein 8.64 mg/L, erythrocyte sedimentation rate 42 mm/hour, glucose 89 mg/dL, glycated hemoglobin A1c (HbA1c) 5.8%, BUN 12 mg/dL, creatinine 0.9 mg/dL, total protein 7.9 g/dL, total bilirubin 0.4 mg/dL, albumin 4.9 g/dL, cholesterol 192 mg/dL, triglyceride 104 mg/dL, uric acid 27 mg/dL, ALT 10 IU/L, AST 17 IU/L, ALP 67 IU/L, GGT 17 IU/L, LDH 361 IU/L, and IgE 699 IU/mL.

Antinuclear antibody, proteinase-3, antineutrophilic cytoplasmic antibody (ANCA), myeloperoxidase antineutrophilic cytoplasmic antibody (MPO-ANCA) were found negative. Chronic sinusitis and nasalpolyps were detected in the ear-nose-throat examination and paranasal CT. No pathology was detected in ECG, echocardiography, CT, and abdominal USG examinations of the patient. Chest X-ray revealed low lung ventilation in the lower left [Figure 1]. CT of the thorax showed a nodular mass, ~4 cm in diameter, adjacent to the diaphragm on the basal segment of left lobe [Figure 2a and 2b].

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Endobronchial lesion was not detected in bronchoscopy, and PET-CT showed involvement in favor of malignancy in the mass. The patient was consulted to undergo video-assisted thoracic surgery (VATS). Biopsy taken with VATS was reported as leukocytoclastic vasculitis where pulmonary infarction and eosinophils were evident. The D-dimer and pulmonary artery pressure were found to be normal since pulmonary thromboembolism was not considered clinically significant in the patient. However, pulmonary CT-angiography was performed for exclusion purposes, and pulmonary thromboembolism was excluded. Bilateral lower extremity venous Doppler US was reported as normal. The patient was diagnosed with rheumatology and vasculitis. Azathioprine (AZT) at 2 mg/kg/day and methylprednisolone (MP) at 1 mg/kg/day were started. It was observed that all clinical and radiological findings disappeared in the patient when called for control at 3 and 6 months after starting the treatment. Treatment was continued with 8 mg/day MP and 100 mg/day AZT. The patient was readmitted with a cough complaint in the 18th month of treatment. Newly developed pericardial effusion was detected on further examinations of the patient.

With the recommendations of a rheumatologist, the doses of MP and AZT were increased. Colchicine was added to the treatment. In the follow-up of the patient, pericardial effusion was found to have completely disappeared and the cough symptom was fully controlled. The patient is still under follow-up.

Discussion

Eosinophilic granulomatosis with angiitis is a rare disease consisting of systemic necrotizing vasculitis that affects small and medium vessels and causes blood and tissue eosinophilia and severe asthma. It is also known as CSS. It is mainly characterized by vasculitis accompanied by eosinophilia, which affects the respiratory system. Vascular lesions are in the form of polyarteritis nodosa or microscopic polyangitis. However, granulomas and eosinophils accompany the vasculitic process in CSS. In less than half of the patients, MPO-ANCA (p-ANCA) positivity is detected.

Our patient had vasculitis characterized by asthma, peripheral blood eosinophilia, paranasal sinusitis, pulmonary infiltration and extravascular eosinophils in pulmonary tissue. The history of asthma, higher than 10% eosinophilia and allergy are the three essential criteria met in this case. CSS tends to undergo three stages: first stage with symptoms of non-specific asthma and allergic rhinitis, second stage with hypereosinophilia with eosinophilic pneumonitis organoenteritis, and third stage with systemic vasculitis. These three periods/phases may not always occur sequentially and may present simultaneously in some cases.

Cardiac involvement in 60% of patients accounts for half the deaths associated with the syndrome. Myocardium, pericardium and endocardium can also be involved. It has been reported that heart disease is more common in ANCA-negative patients. ANCA was also observed negative in our patient. When AZT and MP doses were decreased, cardiac involvement appeared as pericardial fluid. However, when drug doses were increased, pericardial fluid could be brought under control. Lung is the most commonly involved organ in CSS. Pulmonary infiltration seen in about 90% of patients is one of the 6 ACR criteria used in the diagnosis of CSS. Generally, temporary, patch-like, diffuse parenchymal changes, or nodules have been reported.

In our case, pulmonary necrosis area (diameter, 4 cm) was detected in thorax CT. In addition, the symptoms of hemoptysis were developed due to pulmonary necrosis. Various case studies have documented that rheumatic complaints, such as arthralgia and myalgia, which were also present in our patient, occur in half of CSS patients and frequently occur during the vasculitic period of CSS. Since there are no large randomized, controlled studies comparing different treatment methods in this disease, it is difficult to determine the optimal treatment. Although corticosteroids provide dramatic improvement as in our case, immunosuppressive treatment and prednisone treatment every other day are recommended in patients with severe multisystem involvement. We also found improvement in our patient after AZT and MP treatment.

This report emphasizes that vasculitic disease involving the lung (especially CSS) should be considered in patients diagnosed with asthma and having cough and/or unmanageable hemoptysis despite treatment.

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Figure 1: Lung ventilation was low in the lower left in chest X-ray

Figure 2: (a) Parenchymal window, CT of the thorax showed a mass in the nodular form, approximately 4 cm in diameter, adjacent to the diaphragm on the basal of left lobe; (b) Mediastinal window
**Statement of ethics**

The patient gave her informed consent before publication of MS and before start of study.

**Ethics committee approval**

Ethics committee approval was received from the Ethics Committee of Private Yunus Emre Hospital (Ethics Committee approval No: 27/07/2020-37).

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Merkel PA, Monach PA. Systemic necrotizing arteritis. In: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K, editors. Fitzpatrick’s Dermatology in General Medicine. 8th ed. McGraw-Hill; 2012. p. 2013-28.

2. Suster S, Moran CS. Akciğer biyopsilerinin yorumu. In: Dizbay Sak S, editor. Istanbul: Nobel Tıp Kitabevleri; 2015. p. 65-85.

3. Choi YH, Im JG, Han BK, Kim JH, Lee KY, Myoung NH. Thoracic manifestation of Churg-Strauss syndrome: Radiologic and clinical findings. Chest 2000;117:117-24.

4. Jagadeesh LY, Sangle SR, Verma H, D’Cruz D. Alveolar haemorrhage in eosinophilic granulomatosis and polyangiitis (Churg-Strauss). Clinical Rheumatology 2014;33:1177-9.

5. Lanham JG, Elkon KB, Pusey CD, Hughes GR. Systemic vasculitis with asthma and eosinophilia: A clinical approach to the Churg-Strauss syndrome. Medicine 1984;63:65-81.

6. Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP, et al. The American college of rheumatology 1990 criteria for the classification of Churg-Strauss syndrome. Arthritis Rheum 1990;33:1094-100.

7. Guillevein L, Lhote F, Gayraud M, Cohen P, Jarrousse B, Lortholary O, et al. Prognostic factors in polyarteritis nodosa and Churg-Strauss syndrome. A prospective study in 342 patients. Medicine 1996;75:17-28.

8. Çiledağ A, Denzi H, Eledağ S, Özkal C, Dü zgün N, Erekul S, et al. An aggressive and lethal course of Churg-Strauss syndrome with alveolar hemorrhage, intestinal perforation, cardiac failure and peripheral neuropathy. Rheumatol Int 2012;32:451-5.

9. Fishman AP, Fishman AP. Pulmonary Diseases and Disorders. New York: McGraw-Hill; 1988.

10. Nurşen Duzgun. Churg-Strauss Sendromu. Available from: http://ichastıklaririmatoloji.medicine.ankara.edu.tr/wpcontent/uploads/sites/680/2014/02/Churg-Strauss-Sendromu.pdf.