Churg-Strauss Syndrome: A Case Resembling Erythema Multiforme: Differential Diagnosis

Eriteme Multiformeye Benzeyen Churg-Strauss Sendromlu Bir Olgu

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ABSTRACT A 44-year-old female patient presented to our outpatient clinic with breathing difficulty, abdominal pain, articular pain and skin eruptions. Her story included bronchial asthma, for which she has been monitored for 10 years. The patient was diagnosed as Churg-Strauss syndrome (CSS) based on clinical, laboratory and histopathological findings. Treatment with corticosteroid 60 mg/day was initiated. The patient's lesions improved after the treatment.

Key Words: Churg-Strauss Syndrome; erythema multiforme

ÖZET Kırk dört yaşında bayan hasta nefes darlığı, deri döküntüsü, karın ve eklem ağrısı şikayetleriyle kliniğimize başvurdu. Hastanın öyküsünden 10 yıldan beri bronşiYal astım tanısıyla takip edildiği öğrenildi. Klinik, laboratuVar ve histopatolojik bulgular sonucunda hastaya Churg-Strauss sendromu (CCS) tanısı konuldu. Başlanılan kortikosteroid 60 mg/gün tedavisi sonucunda hastanın lezyonları iyileşti.

Anahtar Kelimeler: Churg-Strauss Sendromu; eritema multiforme

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Churg-Strauss syndrome (CCS), also known as allergic granulomatosis and necrotizing angiitis, was first described by Churg and Strauss in 1951.[1] CSS is a systemic necrotizing vasculitis that influences various organs, especially lungs, and involves small to medium-sized arteries, veins, capillaries and venules.[2] The disease is characterized by bronchial asthma, systemic vasculitis, peripheral eosinophilia and perivascular granulomatosis.[3]

A 44-year-old female patient presented to the Dermatology Department of Fırat University School of Medicine with rash and itching that had started on her hands, and fatigue. Medical history revealed that she has had asthma for 10 years. She had no diseases prior to the occurrence of lesions and did not use any medication, except for inhaler short term ß2-mimetic. In the system query of the patient, there was respiratory distress, abdominal pain, knee and elbow aches.

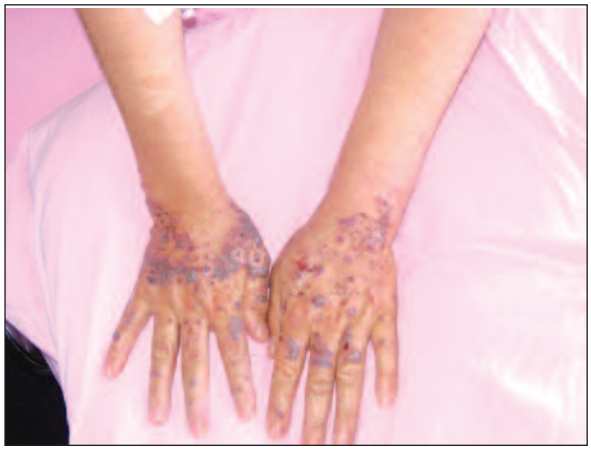

FIGURE 1: Erythematous plaques resembling erythema multiforme and bullae with hemorrhagic content.

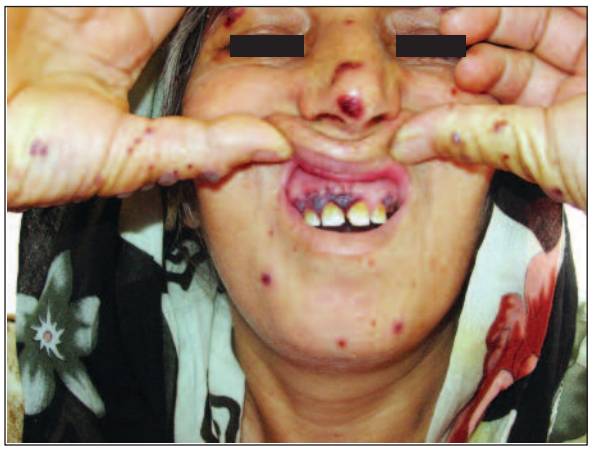

FIGURE 2: Hemorrhagic bullae and eroded areas on oral mucosa, especially on the soft palate and gingivae of the patient.

Physical examination revealed that the blood pressure was 110/80 mmHg, body temperature was 36°C and pulse rate was 80/min. Hemorrhagic bullae on the dorsal part of both hands and ears, and hemorrhagic crusts and eroded areas on the oral mucosa, especially on the soft palate and gingivae were detected (Figures 1 and 2). Other system examinations were normal. The patient had no ocular involvement.

The results of laboratory tests were as follows: white blood cell count: 23.600/mm³, eosinophiles in the differential: 36.4% (0.9-6%), total Ig E: 676 IU/L, CRP: 93.7 mg/L (0-5 mg/L) and erythrocyte sedimentation rate: 40 mm/h. The remaining biochemical parameters in the blood were normal; C3, C4, ANA, P-ANCA, C-ANCA, parvovirus-B19, CMV, coxackie, EBV and antibodies for viral hepatitis were negative. Urine, blood and lesion cultures did not yield any microorganism. While no pathology was detected in the postero-anterior chest X-ray and respiratory function tests, high frequency resonance thoracic tomography showed consolidation areas on the right lung and frosted glass view, and nasal polyps were found in the paranasal sinus tomography.

In the histopathological examination of the punch biopsy material obtained from the dorsal surface of the hand, intensive inflammatory cell infiltration and extensive collagen degeneration was detected on the dermis under multi-layer flat epithelium, where epithelioid histiocytes, with palisade order but without significant granuloma formation, as well as many eosinophils, neutrop-

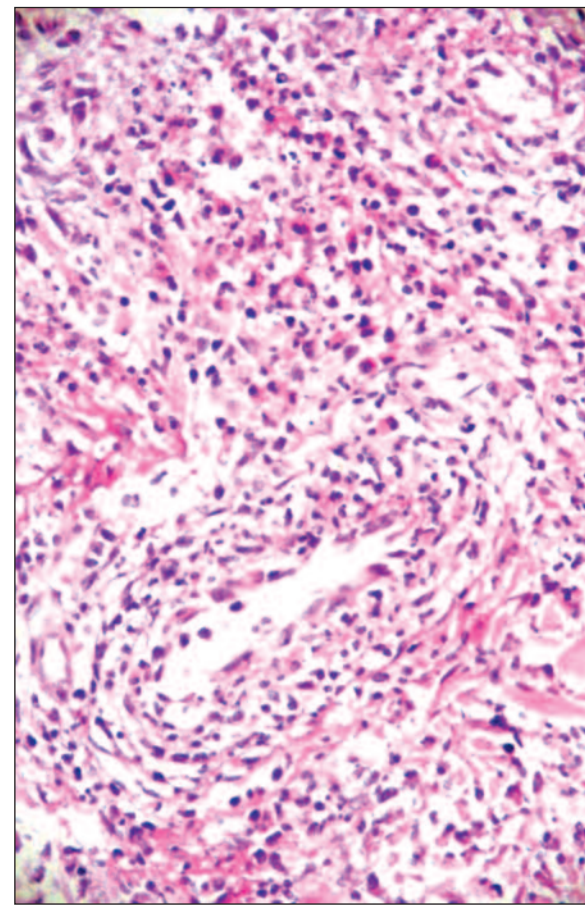

FIGURE 3: Epithelioid histiocytes with palisade arrangement as well as numerous eosinophils, neutrophils and nuclear residues, endothelial swelling and erythrocyte extravasation on vascular walls in the dermis. HE X 200.

hils and nuclear residues were observed. Occasional neutrophils, nuclear residues, endothelial swelling and erythrocyte extravasation were observed on vessel walls, which was consistent with CSS (Figure 3).

CSS diagnosis was made based on clinical, laboratory and histopathological findings and systemic corticosteroids 60 mg/day as well as topical corticosteroid therapy was initiated. The patient was transferred to the gastroenterology clinic on day four of hospitalization with the suspicion of gastrointestinal system hemorrhage, due to melena. The patient started to receive azathiopurine 100 mg/day on the fifth day following the discontinuation of systemic steroid therapy. Endoscopic examination revealed gastric ulcer. The patient is currently being monitored.

DISCUSSION

CCS, also known as granulomatous necrotizing vasculitis, is a systemic vasculitis, which primarily affects the respiratory system. The disease is more common in women and at around the age of 50.[4,5] Although the etiology of the disease is unknown leukotriene receptor antagonists like zafirlukast, which is used in asthma therapy, azithromycine and cocaine are suspected.[1,4-6] Cases following hepatitis-B vaccination were reported.[1] There was no medication use, except for inhaler short-term ß2-mimetic in the history of our case.

In 1990, the American Rheumatology Association suggested the diagnostic criteria for the disease as bronchial asthma, presence of more than 10% eosinophilia in the differential blood smear, mononeuropathy or polyneuropathy, imaging of temporary pulmonary infiltrates in PA chest X-ray, paranasal sinus pathologies and eosinophilic infiltration outside the vessels in skin biopsy material. Presence of at least four of the six criteria is considered diagnostic.[5] Our case who met four of the criteria was diagnosed as CSS.

Cutaneous involvement is observed in approximately 40% of the patients.[4] Straus et al classified the skin signs in 3 categories as maculopapular lesions resembling erythema multiforme, hemorrhagic lesions ranging from petechiae to ecchymosis and cutaneous-subcutaneous nodules.[7] The previous erythema multiforme-like cases in the literature were described as targetoid lesions or bullous targetoid lesions.[8-10] In our case, cutaneous eruption consisted of large, red, urticated round plaques with dusky centers. Targetoid lesions resembling erythema multiforme at the onset later changed to hemorrhagic bullae with a necrotic presentation and healed leaving a scar.

Histopathological examination of skin biopsies shows palisaded granulomas with basophilic necrosis at the center and giant cells, neutrophils and eosinophils lined up in the periphery.[4] Crotty et al who classified cutaneous lesions in 3 histopathological categories reported 50% extravascular granulomas, 33% leukocytoclastic vasculitis and 17% cutaneous polyarthritis nodosa.[11,12] Our patient's histopathological signs were in the form of leukocytoclastic vasculitis accompanied by eosinophilic infiltrations, which were in agreement with CSS.

Although the literature includes reports of polymorphic skin lesions like ulcers at the tips of the digits, necrosis, hemorrhagic bullae, urticarial and livedoid erythema as skin signs in CSS, we have not met any CSS case whose onset was with erythema multiforme resembling lesions.[13-15] Therefore, we decided to publish our case to draw attention to both vasculitis, which is used in the differential diagnosis of erythema multiforme, and CSS.

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