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

Eriteme Multiformeye Benzeyen Churg-Strauss Sendromlu Bir Olgu

**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, karn ve eklem ağrışi şikayetleriyle kliniğimize başvurdu. Hastanın öyküsünden 10 yıldan beri bronşiyal astım tanıstıla takip edildiği öğrendi. Klinik, laboratuvar ve histopatolojik bulgular sonucunda hastaya Churg-Strauss sendromu (CSS) tanısı konuldu. Başlamanın kortikosteroid 60 mg/gün tedavisi sonucunda hastanın lezyonları iyileşti.

**Anahtar Kelimeler:** Churg-Strauss Sendromu; eritema multiforme

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

A 44-year-old female patient presented to the Dermatology Department of Firat 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 β2-mimetic. In the system query of the patient, there was respiratory distress, abdominal pain, knee and elbow aches.
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 histocytes, with palisade order but without significant granuloma formation, as well as many eosinophils, neutrop-
hils and nuclear residues were observed. Occasion-
lar neutrophils, nuclear residues, endothelial swell-
ing and erythrocyte extravasation were observed
on vessel walls, which was consistent with CSS (Fi-
gure 3).

CSS diagnosis was made based on clinical, la-
boratory and histopathological findings and system-
ic 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 mele-
na. The patient started to receive azathiopurine 100
mg/day on the fifth day following the discontini-
uation of systemic steroid therapy. Endoscopic ex-
amination revealed gastric ulcer. The patient is
currently being monitored.

DISCUSSION

CSS, also known as granulomatous necrotizing vas-
culitis, is a systemic vasculitis, which primarily af-
fected 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 le-
ukotriene receptor antagonists like zafirlukast,
which is used in asthma therapy, azithromycin and
cocaine are suspected.1-6 Cases following hepatitis-
B vaccination were reported.1 There was no medi-
cation use, except for inhaler short-term ß2-mimetic
in the history of our case.

In 1990, the American Rheumatology Associ-
ation suggested the diagnostic criteria for the dis-
ease 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 infil-
tration outside the vessels in skin biopsy material.

Presence of at least four of the six criteria is consi-
dered diagnostic.5 Our case who met four of the cri-
teria was diagnosed as CSS.

Cutaneous involvement is observed in approxi-
ately 40% of the patients.4 Straus et al classified
the skin signs in 3 categories as maculopapular
lesions resembling erythema multiforme, hemorr-
hagic lesions ranging from petechiae to ecchymo-
sis and cutaneous-subcutaneous nodules.7 The
previous erythema multiforme-like cases in the lit-

erature were described as targetoid lesions or bul-

lous targetoid lesions.8-10 In our case, cutaneous

eruption consisted of large, red, urticated round
plaques with dusky centers. Targetoid lesions re-
ssembling erythema multiforme at the onset later
changed to hemorrhagic bullae with a necrotic pre-
sentation 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 histopat-

hological categories reported 50% extravascular
granulomas, 33% leukocytoclastic vasculitis and
17% cutaneous polyarthritis nodosa.11,12 Our pati-

ent’s histopathological signs were in the form of le-

ukocytoclastic 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 ha-

ve not met any CSS case whose onset was with ery-

thema multiforme resembling lesions.13-15 There-

fore, we decided to publish our case to draw atten-
tion to both vasculitis, which is used in the diffe-

rential diagnosis of erythema multiforme, and CSS.
REFERENCES


