A good visual recovery in a patient with central retinal artery occlusion associated with Churg-Strauss syndrome: case report

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ABSTRACT

Background: Churg-Strauss syndrome (CSS) is a rare systemic necrotizing vasculitis of unknown etiology that affects small-to-medium-sized vessels. A multisystem disease that mostly affects the lungs and the skin. In addition, it could affect the renal, cardiovascular, gastrointestinal, central, and peripheral nervous systems. Ocular involvement in the CSS is rare. The reported ocular manifestations include corneal ulcer, uveoscleritis, conjunctival granuloma, orbital inflammatory pseudotumor, amaurosis fugax, retinal artery occlusion, ischemic optic neuropathy, oculomotor nerve palsy, and trochlear nerve palsy.

Case Presentation: A 53-year-old man was admitted with sudden visual loss. CSS was diagnosed earlier, based on asthma, eosinophilia, peripheral neuropathy symptoms, and pulmonary infiltrates. His vision was completely lost in the left eye. Unilateral central retinal artery occlusion (CRAO) was observed by fluorescence angiography.

Results: The treatment modality used in our patient was steroid pulse therapy, and his vision improved 24 hours later. This outcome might emphasize the importance of steroid pulse therapy since it may be effective for CRAO in CSS patients.

Conclusion: This case report described a CSS patient with associated CRAO, in whom steroid pulse therapy improved his visual acuity. Therefore, since intensive treatment should be administered as early as possible to improve the prognosis of vision, rheumatologists should be aware of the presence of CSS-associated CRAO, and they should refer those patients to ophthalmology as soon as possible.

Keywords: Churg-Strauss syndrome (CSS), central retinal artery occlusion (CRAO).

Introduction

Churg-Strauss syndrome (CSS) is a rare systemic necrotizing vasculitis of unknown etiology that affects small-to-medium-sized vessels and is associated with severe asthma and blood and tissue eosinophilia. CSS is a multisystem disease that mostly affects the lungs and the skin. Also, it could affect the renal, cardiovascular, gastrointestinal, central, and peripheral nervous systems. Patients with this syndrome could present with several clinical features, including severe allergic asthma attacks, allergic rhinitis/sinusitis, skin nodules, impaired mental status, pericarditis, and Gastrointestinal (GI) involvement. Ocular involvement in the CSS is rare. The reported ocular manifestations include corneal ulcer, uveoscleritis, conjunctival granuloma, orbital inflammatory pseudotumor, amaurosis fugax, retinal artery occlusion, ischemic optic neuropathy, oculomotor nerve palsy, and trochlear nerve palsy. In this report, we will discuss a rare case of CSS in which the patient presented with central retinal artery occlusion (CRAO).

Case Report

On December 10, 2018, a 53-year-old male presented in the infectious disease department because of suspicion of non-tuberculous mycobacteria. He presented with
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respiratory symptoms, cardiac symptoms with high Troponin I and normal cardiac catheter, and neurological symptoms with left leg peripheral neuropathy symptoms. He had a history of myocarditis, and he quit smoking 15 years ago. Also, he has a family history of asthma. Before that, he came in October for investigations, and it was found that he has high White blood cell (WBC) reaching 70, high ESR, and high eosinophils. Also, a bone marrow biopsy was done and sent to Mayo Clinic, and it showed no hematological malignancy. Imatinib sensitive mutations, which Fluorescence in situ hybridization (FISH) did, and a gene sequence found in an abnormal chromosome 22 of some people with certain forms of leukemia (BCR-ABL) were all negative. A rheumatology workup was done and showed high Rheumatoid factor (RF) but no other rheumatic symptoms. He was given Prednisone and responded well. Then the infectious disease department concluded that this case was non-pathogenic. They put him off steroids, and he was doing well. On December 23, 2018, in the hematology department, Complete blood count (CBC) was done and showed average eosinophils count, leukocytosis, and neutrophilia, which could be secondary to bladder infection and cystitis. The patient was asymptomatic. Then a few days later, his eosinophils level became 14,000. Moreover, it was shown that he is having Raynaud Phenomenon. Then he was given 100 mg of Imatinib. After a week, he developed constitutional symptoms and went to the Emergency Room (ER), they did Chest X ray (CXR), which showed that there were changes in his right lung, there was no fever, cough, nor sputum. After that, they increase the dose of Imatinib to 200 mg. On January 24, 2019, he had a sudden loss of vision. The patient described his condition as a sudden darkening of image and slow improvement after a few minutes, but he still sees things only near to the face in the left eye. He mentioned that he has a history of wearing glasses. Complete examination details are provided as supplementary data.

A few days later, the vision improved to 20/20 in both eyes (Figure 1, 2). After that, he visited the rheumatology department on February 6, 2019, and it was shown that he had let foot drop began 10 days ago multiple mono neuritis multiplex with hypereosinophilia and weight loss, and negative bone marrow biopsy for malignancy of primary hypereosinophilic syndrome. The diagnosis was most likely Antineutrophil cytoplasmic antibodies (ANCA) negative Churg Strauss syndrome. On February 10, 2019, his abdominal CT scan showed evidence of small bowel perforation. Exploratory laparotomy and small bowel resection were done for him. Even though there was only one area of perforation, the entire length of the small bowel was affected by vasculitis. After that, this case was complicated by a severe attack of Cytomegalovirus (CMV) myocarditis that necessitated an ICU admission. Echocardiogram (ECHO) at that time showed severely compromised cardiac function with an Ejection fraction (EF) of 10%-15% subsequently. The patient was extubated and sent to the progressive care unit. On February 24, 2019, the patient started to complain of a new-onset abdominal pain. Then a CT scan showed three new small bowel perforations. He was a poor candidate for surgery because of his very poor cardiac status. His condition was explained to the patient and his family, but they insisted on going for surgery at high risk. Shortly after this surgery, the patient suddenly went into Pulseless electrical activity (PEA), which an anesthesiologist managed. Once the pulse was regained after prolonged resuscitative measures, the patient was sent to the ICU, where he had cardiac arrest shortly afterwards and died.
Discussion

We described a case of ANCA negative CSS, which was complicated by CRAO. Patients mostly present with several clinical features in this syndrome, including severe allergic asthma attacks, allergic rhinitis/sinusitis, skin nodules, impaired mental status, pericarditis, and GI involvement [1]. Ocular involvement in the CSS is rare. The reported ocular manifestations include corneal ulcer, uveoscleritis, conjunctival granuloma, orbital inflammatory pseudotumor, amaurosis fugax, retinal artery occlusion, ischemic optic neuropathy, oculomotor nerve palsy, and trochlear nerve palsy [2]. Our patient presented with asthma, eosinophilia, peripheral neuropathy symptoms, and pulmonary infiltrates. These manifestations satisfied the American College of Rheumatology 1990 criteria for the classification of CSS. Moreover, leukocytosis, the elevation of Erythrocyte sedimentation rate (ESR), high eosinophils level, and positive rheumatoid factor further confirmed the diagnosis of CSS. Akella et al. [3] explained the ophthalmic manifestations of CSS. In order of most frequent presentation to least frequent, these include central retinal artery or vein occlusion, ischemic optic neuropathy, conjunctival nodules, orbital myositis, proptosis, dacryoadenitis, retinal vasculitis/infarcts/edema, cranial nerve palsy, and amaurosis. In this patient, the ocular manifestation he presented with was a sudden unilateral CRAO, which was diagnosed by fundoscopic findings (retinal whitening with a cherry-red spot). CRAO is an ophthalmic emergency characterized in general by the presence of acute, devastating, and unrecoverable sight loss. The treatment of this condition is usually done by IV or intra-arterial thrombolysis to dissolve the clot, hyperbaric oxygen therapy to increase blood oxygen tension, vasodilators to increase blood oxygen content, or by decreasing the Intraocular pressure (IOP) with medications or surgeries [4]. There has been a previous report about a CSS patient without ANCA who developed unilateral CRAO. Steroid pulse therapy (methylprednisolone at 1 g daily for 3 days) followed by combined treatment with prednisolone (30 mg/day) and cyclophosphamide (150 mg/day) was administered to this patient. After treatment, his visual acuity recovered to 20/30 in 1 month, and no recurrence has occurred for 1 year [5]. However, previous reports showed poor visual outcomes in CRAO associated with CSS. It could be due to low-dose of systemic corticosteroids (prednisolone ≤ 60 mg) or delay in establishing steroid pulse therapy [6,7,8]. Our patient’s treatment modality was also steroid pulse therapy, and his vision improved 24 hours later. This outcome might emphasize the importance of steroid pulse therapy since it may be effective for CRAO in CSS patients. Also, we want to emphasize the importance of the time of starting the treatment. This means that the onset and the start of treatment, but not the treatment regimen, might have influenced the outcome of the patients. Therefore, intensive treatment should be given as soon as possible.

Conclusion

In summary, this case report described a CSS patient with associated CRAO, in whom steroid pulse therapy improved his visual acuity. Therefore, since intensive treatment should be administered as early as possible to improve the prognosis of vision, rheumatologists should be aware of CSS-associated CRAO and should refer those patients to ophthalmology as soon as possible.

List of Abbreviations

- CRAO  Central retinal artery occlusion
- CSS  Churg-Strauss syndrome

Conflict of interest

The authors report no conflict of interest.

Funding

None.

Consent for publication

Consent was taken verbally from the patient’s relatives since the patient is dead.

Ethical approval

Ethical approval was granted by King Abdullah International Research Center in May 18, 2021.

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Supplementary Data

On examination

C/O sudden loss of vision 1 hour before
Right eye (OD)  20/20
Left eye (OS)   CF 4 Ft  CF 4 Ft
OD  16
OS  15
LE relative afferent pupillary defect (RAPD)
Cornea-clear ou
Ac-deep, quiet
Iris-normal
Lens-clear
Fundus undilated
Attenuation of the vessels
Segmentation of the blood column in the arteries/cattle trucking in the superior arcade
Cheery red spot
Pale retina

Impression and recommended plan of care

CRAO left eye
Acute care
Ocular massage with gonio lens given
Acetazolamide 500 mg stat
Cosopt stat
Oct/Fundus photo done
Findings confirmed

Plan
Intermittent ocular massage given
Consult hematology consulted
Advise to give inj methylprednisolone and oral steroids and follow-up in-clinic sunday

Next visit on 25/01/19

On examination: his visual acuity was 20/20 in the right eye and 20/40 in the left eye. IOP was 15 in the right and 17 in the left eye.

Anterior segment

Conjunctiva: normal in both eyes
Cornea: clear in both eyes
AC-deep, quiet
Pupils: brisk
Iris: normal
Lens: clear

Fundus

The left eye arteries have no filling defect, good orange hue fundus, no retinal edema
Healthy retina
An excellent red reflex of the fovea

Impression

CRAO associated with transient blindness.