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CASE REPORT

Churg-Strauss syndrome presented as Pseudo-Foster-Kennedy syndrome: a case report

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A 52-year-old woman with bronchial asthma, frequent sneezing, and a skin lesion has suddenly developed poor vision for 15 days in the left eye. Her presenting vision was 6/18 and perception of light was positive in the right and left eyes, respectively, with mild pallor in the right optic disc and disc swelling in the left eye. She had a high eosinophil count, positive ANCA, and a skin biopsy in favor of Churg-Strauss disease. The patient was diagnosed with left optic neuropathy as a result of arteritis, which is a component of Churg-Strauss syndrome (CSS), and was treated with Inj. Methylprednisolone pulse therapy, followed by oral prednisolone but preceded to no perception of light in the left eye after 7 days of follow-up. Although CSS is a rare disease, routine checkup by an ophthalmologist can save vision and identify the high-risk group for proper referral to prevent unwanted morbidity and mortality.

Keywords: Churg-Strauss syndrome, asthma, skin allergy, optic disc swelling, methylprednisolone

Introduction

Churg-Strauss (CSS) is one of the inflammatory diseases affecting blood vessels of medium and small size. It is also known as eosinophilic granulomatosis with polyangiitis (EGPA), allergic angitis, and Charge-Strauss vasculitis. The current name of the disease is EGPA. Two physicians, namely, Churg and Strauss, first described this syndrome as asthma, hypereosinophilia, and multi-system vasculitis in 1951 (1). It is a rare disease, and its incidence is 2.4–6.8 per million annually (2). This disease can affect person of any age but is commonly diagnosed between 30 and 50 years old and has a female predominance (3). It most commonly affect the nose, sinuses, lungs, renal system, heart, intestine, and peripheral nerves, but ocular involvement is rare (4). Ophthalmologic manifestations that are found in the literature are keratitis, granuloma in the conjunctiva, inflamed orbit, uveoscleritis, amaurosis fugax, arterial occlusion of retina, ischemic neuropathy, and paralysis of the third and fourth cranial nerves (5).

Our CSS case was presented as a Pseudo-Foster-Kennedy syndrome, and we are sharing our experience.

Case report

A 52-year-old woman developed vision loss in her left eye for 15 days, which was not gradual. She was non-diabetic, non-hypertensive but had bronchial asthma, frequent sneezing with nasal blockage, and skin allergy. She presented with 6/18 and perception of light vision in her right and left eyes, respectively, without any improvement in refraction. Her other ocular examination was normal except for the fundus. Posterior segment examination revealed mild pallor of the right optic disc and disc swelling in the left eye (**Figure 1**). Systemic examination showed some skin lesions in both hands and legs (**Figure 2**) and a few rhonchi in lungs. No other abnormality was found in cardiac, gastrointestinal, and nervous systems. Her previous biopsy report from a skin lesion showed Churg-Strauss disease. Routine blood examination showed a normal leukocyte

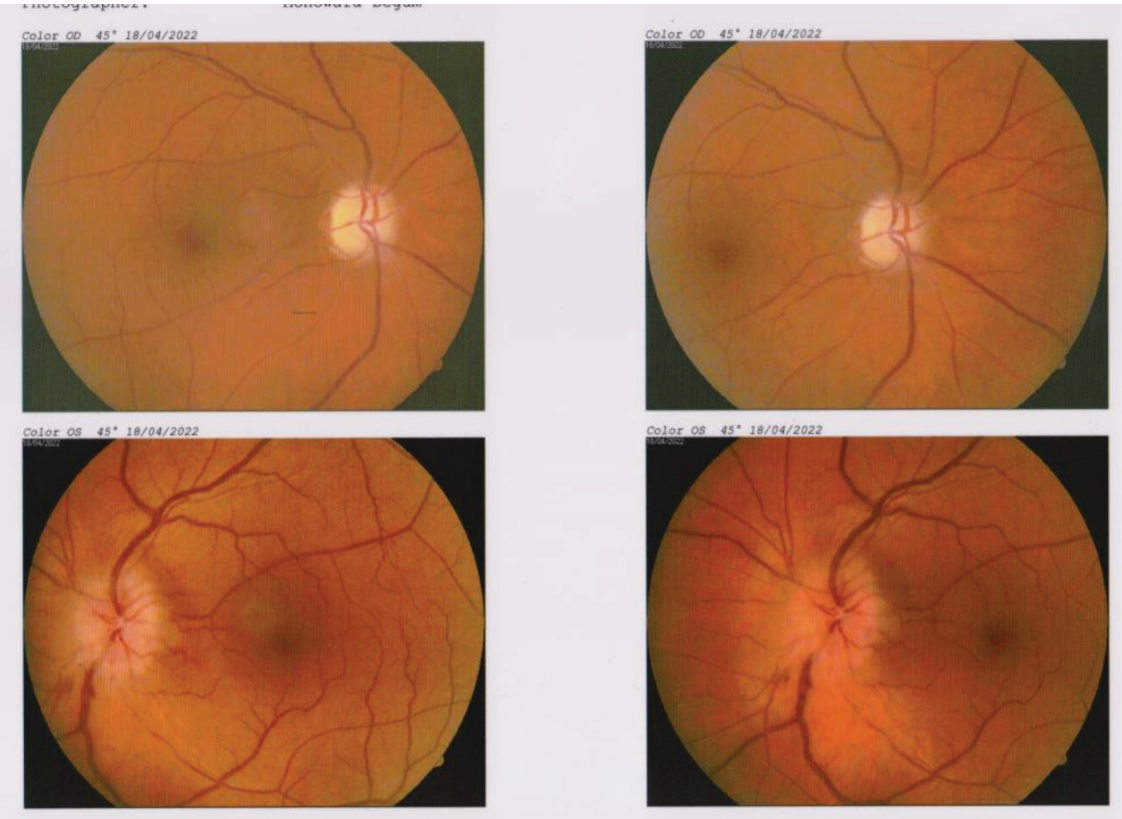

FIGURE 1 | Right optic disc is pale, which indicates optic atrophy due to previous inflammation. **Left** optic disc is swollen, indicating inflammation of the optic nerve.

count with eosinophilia, which was 32%. Her chest x-ray, ECG, echocardiogram, serum creatinine, and CT scan of the brain were normal except for p-ANCA. Her visual field test also showed peripheral field defect in the right eye (**Figure 3**). She was diagnosed with right-sided optic atrophy and left-sided optic neuropathy as a result of arteritis, which is a component of CSS. She was treated with Inj. Methylprednisolone pulse therapy, 1 g intravenously once daily for 3 subsequent days. After 3 days, she received prednisolone orally at a dose of 1 mg/kg body weight, with gradual tapering to save sight in the left eye. After 7 days of follow-up, there was no improvement in left eye vision and there was no perception of light, but in the right eye it was 6/9. As she presented with right eye optic atrophy and left eye disc swelling without any brain lesion, the case seems to be Pseudo-Foster-Kennedy syndrome. She was also referred to a medicine specialist for further evaluation and additional need for cytotoxic medication. In the next 1 year, the patient was followed up at regular intervals, and no other ocular disturbance was reported.

Discussion

The underlying pathology of CSS is not well understood and multiple organs may be affected. Most patients had a history of bronchial asthma and adult-onset asthma should be suspected of CSS. This disease passes through three distinct clinical phases, but it may not always follow the subsequent phases, which sometimes makes the diagnosis of the disease difficult. CSS initially begins with atopic allergy and asthma. This disease may even start in children. Among the three phases, the first one continues up to months or a year before stepping into the next phase. Granulomatous inflammation and infiltration with eosinophils are the criteria of the second phase. Clinical presentation of the second phase may be pneumonia, gastroenteritis, orbital pseudotumour, etc. In the third phase, medium- and small-sized blood vessels are affected by necrotizing vasculitis, which causes harm to the eye along with other vital organs like the skin, gastrointestinal tract, renal system, and heart (1). CSS can be misdiagnosed as vasculitis, which is the prime characteristic and can affect multiple systems. Most patients (50%) presented with skin lesions, and about 30% may present either renal, gastro-intestinal, or peripheral nerve involvement (6). Cardiac involvement is about 10%, but myocarditis and coronary arteritis are the primary causes of morbidity and mortality (1).

There are other vasculitis like Wegener's granulomatosis, hypereosinophilic syndrome, and microscopic polyangiitis that may confuse (**Table 1**) (7). Proper history, clinical examination, and investigations are needed for diagnosis,

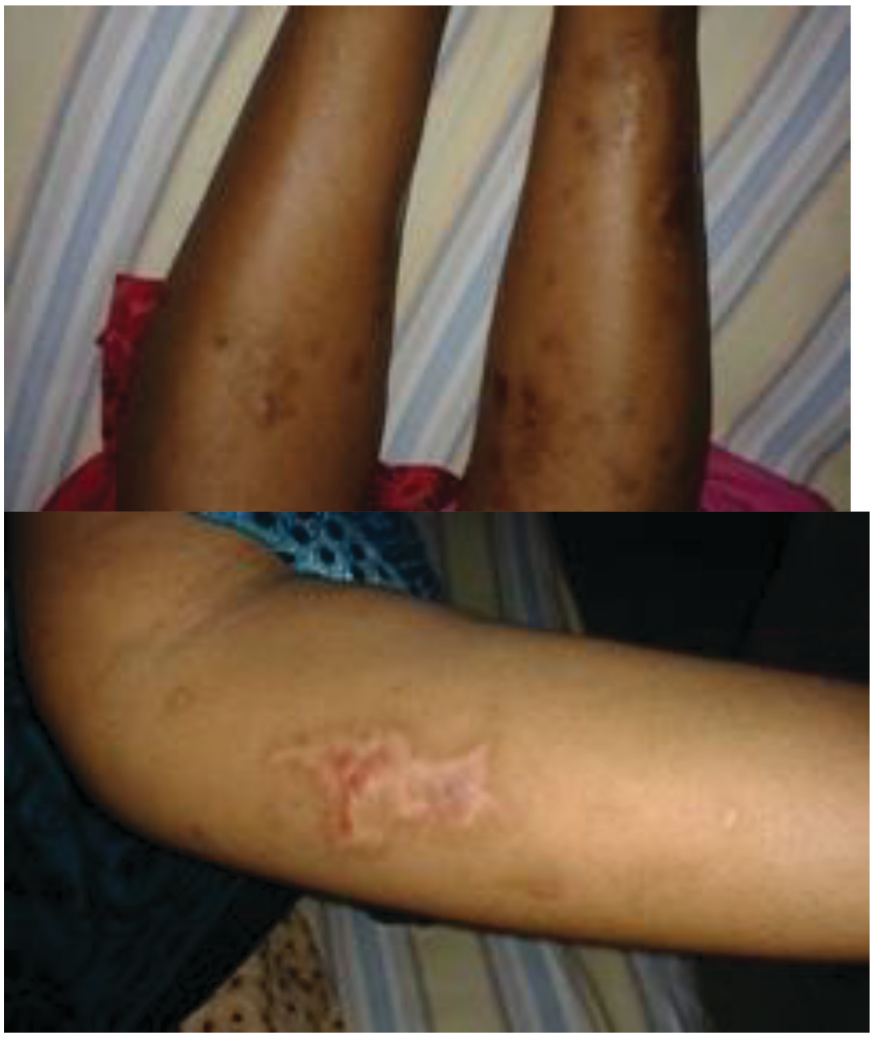

FIGURE 2 | Multiple skin lesion in the legs and the hand which is one of the hallmarks of Churg-Strauss syndrome.

and histopathology is the confirmatory diagnostic tool. To diagnose CSS, six criteria were set by the American College of Rheumatology in 1990, and four should be positive (8). According to Masi et al. (8) these criteria are 99.7% specific and 85% sensitive if four or more are positive. These criteria are as follows:

(1) Bronchial asthma
(2) >10% eosinophilia in peripheral blood film
(3) Paranasal sinusitis
(4) Lung infiltrations
(5) Histologically proven vasculitis along with extravascular eosinophils
(6) Mono neuritis multiplex or polyneuropathy

Classical triad is necrotizing arteritis, eosinophilic infiltration, and extravascular granuloma, but for a definite diagnosis, histopathology is indicated. In our patient, out of six, five criteria, namely, asthma, eosinophilia, sinusitis, optic neuropathy, and a histopathologically proven skin lesion were present.

Ocular manifestation of CSS is rare, and according to Takanashi et al. (5) it may have two varieties: orbital pseudotumor and ischemic vasculitis. Takanashi hypothesized that both varieties actually indicate granulomatosis and angiitis, which are two important criteria of the disease. Our patient had no peripheral neuropathy but presented with optic nerve involvement, which represents involvement of the central nervous system. The literature search showed that CNS involvement is the major cause of mortality, but it is a rare incidence (6).

Some authors found a relationship between ANCA and CSS. Serum ANCA, especially p-ANCA, may be positive in 70% of cases of CSS (9). There are also relationship between myeloperoxidase antibody (MPO) and CSS, and about 50–80% of samples with a p-ANCA pattern will have MPO antibodies. A literature review showed ANCA is usually present in ischemic vasculitis, indicating the third stage of the disease, which is a possible risk for sudden vision loss. Systemic steroid can be used as prophylaxis for sight- and life-threatening conditions of the third phase (5). Although steroid is the mainstay of treatment, 20% of patients may

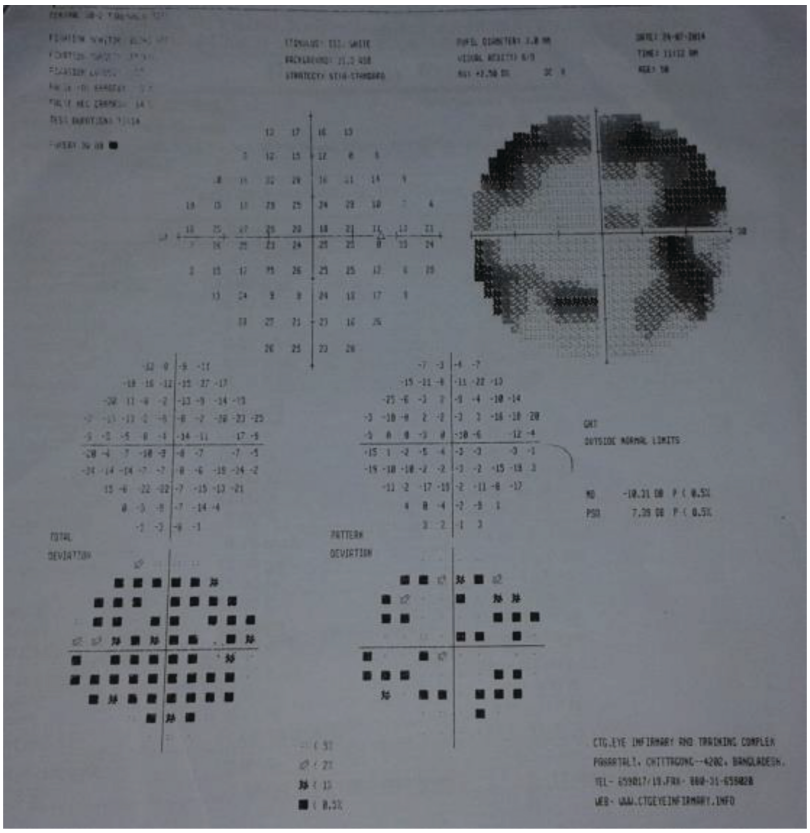

FIGURE 3 | Visual field test showed peripheral field defect in the right eye.

TABLE 1 | Distinguishing features of Churg-Strauss syndrome (CSS) from other differentials.

	CSS	HES	WG	MPA
Asthma	+	–	–	–
Eosinophilia	+	+	–	–
Sinusitis and rhinitis	+	–	–	–
Lungs involvement	+	+	+	+
Heart involvement	+	+	Rarely	Rarely
Skin involvement	+	+	+	+
Vasculitis	+	–	+	+
Eosinophilic infiltration	+	+	–	–
Granuloma	+	–	+	–
ANCA	Yes (50–80%, myelo-peroxidase)	–	Yes (90%, proteinase 3)	Yes (60%, myelo-peroxidase and 30% proteinase)

Adopted from Sinico and Bottero (7).
CSS, Churg-Strauss syndrome; HES, hypereosinophilic syndrome; WG, Wegener's granulomatosis; MPA, microscopic polyangiitis.

need cytotoxic drugs to reduce the progression of the disease. The combination of pulse intravenous corticosteroid and cytotoxic medicine may be needed if major life-threatening organs are involved (10). In our case, we started treatment with methylprednisolone, but her vision deteriorated to no perception of light. Due to high risk criteria, the patient was sent to an internist for other systemic evaluation and proper treatment.

Conclusion

Even if CSS is a rare disease, routine referral to an ophthalmologist would be beneficial for patients. Regular checkups, counseling, and early treatment during emergencies by an ophthalmologist can save the vision. An ophthalmologist also has a vital role in identifying high-risk patients, and timely referral can reduce unwanted morbidity.

Author contributions

Both authors listed have made a substantial, direct, and intellectual contribution to the work, and approved it for publication.

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