Branch retinal vein occlusion followed by central retinal artery occlusion in Churg-Strauss syndrome: unusual ocular manifestations in allergic granulomatous angiitis

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INTRODUCTION

Retinal artery and vein occlusion in the same eye is unusual. When this occurs, a systemic disorder such as vasculitis should be suspected. We report a patient with a consecutive branch retinal vein occlusion (BRVO) and central retinal artery occlusion (CRAO) in the same eye who was subsequently found to have Churg-Strauss syndrome (CSS).

MATERIALS AND METHODS

In May 2003, a 55-year-old man with a 6-year history of allergic asthma (pellitory) and kidney cysts was referred for a fluorescein angiography (FA) for a BRVO in the left eye. Visual acuity was 20/20 in the right eye and 20/25 in the left. Funduscopy of the right eye was normal. In the left eye, retinal hemorrhages with retinal thickening were noted along the superotemporal arcade. FA confirmed BRVO in the left eye (Fig. 1) so that argon laser photocoagulation and acetylsalicylic acid therapy (100 mg once a day) was prescribed. In August he went to an internist for a suspected eosinophilic pneumonia. Computed tomography (CT) of the chest revealed an exudative alveolitis and bilateral pleural and pericardial effusion which were treated with prednisone. One year later, he was seen by a neurologist for hemorrhagic ictus with syncope, dysarthria, and right facial-brachial-crural hemiparesis. Duplex Doppler ultrasound was obtained and showed arterial endothelial calcification of the carotid bulbs, left > right. CT and magnetic resonance angiography (MRA) of
the brain revealed an intraparenchymal hemorrhage in the left lenticular nucleus and internal capsule. He started on oral nimodipin and atorvastatin with satisfactory functional recovery. In June 2004, he was seen again in our department for sudden visual loss in the left eye. Ocular examination showed that the best-corrected visual acuity (BCVA) was 20/20 in the right eye and light perception in the left. Slit-lamp examination and intraocular pressures (IOP) were normal but mydriatic left pupil with relative afferent pupillary defect (RADP) was seen. Fundus examination of the right eye showed grade 2 hypertensive retinopathy; funduscopy of the left revealed a single peripapillary flame hemorrhage temporally, laser photocoagulation spots along the superotemporal vein, narrowing of the arterial vessels, a macular “cherry red spot,” and ischemic edema of the retina. The picture was consistent with CRAO; an FA was not obtained due to patient refusal. Due to his systemic problems, he was hospitalized. He experienced mild hyperthermia (37.5 °C) and bilateral purpuric vasculitis in legs and hands, moderate hepatomegaly, left inguinal hernia, slight heart murmur, hyperexcitability of the right osteotendinous reflexes, and hypoexcitability of the left with slight deficiency strength of the left upper limb. His laboratory data showed marked elevation of white blood cell count (WBC 13,690 per mm³) with eosinophils at 54.4% (EO 7,450 per mm³), and an increased value of fibrinogen (515 mg/dL) and of the inflammatory indexes (ESR: 28 mm per hour and CRP: 13,17 mg/dL). Chest X-ray showed accentuation of bronchial pattern with micronodular pulmonary infiltrates. The following criteria – asthma bronchialis, previous eosinophilic pneumonia, hypereosinophilia, recurrent thrombotic events with a vasculitis origin, and purpura – established the diagnosis of CSS (1-3). The patient was started on oral prednisone (75 mg/day progressively tapered to 12.5 mg/day) and cyclophosphamide (150 mg/day) with regression of the purpuric lesions and the mild hyperthermia, normalization of the inflammation index, and improvement in general conditions. In April 2008, the patient was stable on a maintenance dose of prednisone of 5 mg/day but he had only light perception in the affected eye. Eighteen months later, optic atrophy was prominent in the left eye (Fig. 2).

**DISCUSSION**

Churg-Strauss syndrome, or allergic granulomatosis and angiitis, is a systemic vasculitis (1) first described by Churg and Strauss in 1951 (2). They reported 13 patients with severe asthma with a "strikingly uniform clinical picture" including fever, hypereosinophilia, and evidence of
BRVO followed by CRAO in Churg-Strauss syndrome

vascular abnormality in various organ systems. They
termed this condition “allergic granulomatosis and angio-
itis.” Histologically these “allergic granulomas” were com-
posed of necrotic eosinophilic exudates, severe fibrinoid
collagen changes, and granulomatous proliferation of ep-
ithelioid and giant cells. In 1990, the American College of
Rheumatology (3) developed the following criteria for epi-
demiologic and therapeutic studies of CSS: asthma,
eosinophilia greater than 10%, mono- or polynuropathy,
non-fixed pulmonary infiltrates, paranasal sinus abnor-
malities, and biopsy containing blood vessels with ex-
travascular eosinophils. The presence of four of the six
criteria yielded a sensitivity of 85% and a specificity of
99.7%. Ocular manifestations are rarely reported (4-7)
and include conjunctival granuloma, panuveitis, orbital in-
flammatory pseudotumor, ischemic optic neuropathy, and
retinal artery and vein occlusion. In the case we report,
BRVO was the first sign of CSS followed by other sys-
 temic vasculitis signs, including, after 1 year, central
CRAO in the same eye. CSS is a leukocytoclastic sys-
temic small-vessel vasculitis which predominantly affects
 small vessels. Its arteritic phase commonly develops
within 3 years of the asthma onset (8). In our patient,
the vasculitis took 6 years from the prodromal asthma
to appear and its first sign was an unusual BRVO fol-
lowed by eosinophilic pneumonia. Since CSS invariably
involves the lungs but affects a wide variety of other tis-
sues and organs including the eye, we strongly suggest
to always check for systemic pathologies such as CSS
in patients with retinal vessels occlusion and lung dis-
 ease. This is important to make an early diagnosis and
to avoid scenarios like that experienced by our patient:
hemorrhagic ictus and CRAO. Moreover, CSS is only
one among the systemic underlying diseases which can
lead to consecutive or simultaneous retinal vein and
artery occlusions in patients younger than 65 years.
Previously reported rare cases showed other systemic
causes liable for combined retinal vein/artery occlusions
such as thrombophilia (9,10), antiphospholipid syn-
drome (11), hyperhomocysteinemia (12), lupus erythe-
matosus (13), type 2 diabetes (14), interferon therapy
(15), acquired immunodeficiency syndrome (16), throm-
botic thrombocytopenic purpura (17), systemic non-
Hodgkin lymphoma (18), acute lymphoblastic leukemia
(19), infective endocarditis (20), and hormone replace-
ment therapy (21). Knowing these causes, it is essential
to make an early diagnosis and to not miss any underly-
ing disease.

CONCLUSIONS

BRVO and CRAO can occur in the same eye in CSS. In
the presence of systemic signs or symptoms, it is impor-
tant to rule out systemic vasculitis in order to start appro-
priate immune-modulatory treatment, thereby avoiding
unnecessary mortality. Therefore, in patients younger than
65 years with an occlusive retinal vessel episode and lung
disease, we strongly suggest to always check for sys-
temic vasculitis such as CSS. This is to avoid a delayed
diagnosis which can lead to more severe complications.
Furthermore, in patients with combined retinal artery/vein
occlusion, we recommend to always consider any under-
lying systemic disease through a careful screening.

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