

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

GABRIELLA DE SALVO, CONCETTA LI CALZI, MARIO ANASTASI, GAETANO LODATO

University Dept. of Clinical Neuroscience (DINEC), Ophthalmology Unit, University of Palermo, Palermo - Italy

PURPOSE. To describe a rare branch retinal vein occlusion (BRVO) followed by central retinal artery occlusion (CRAO) in a patient with Churg-Strauss syndrome (CSS).

METHODS. A 55-year-old man with a not yet diagnosed CSS developed a BRVO in the left eye and 1 year later a CRAO with painless and acute vision loss in the same eye. Medical history included bronchial asthma, history of allergy, eosinophilic pneumonia, bilateral pleuric and pericardial effusion, hypereosinophilia, and purpuric vasculitis.

RESULTS. CRAO in the left eye was diagnosed by retinal whitening and a cherry red spot with coexisting old BRVO evidenced by previous laser photocoagulation. Corticosteroids and cyclophosphamide therapy improved his general condition but no visual recovery occurred.

CONCLUSIONS. BRVO and CRAO can occur in the same eye in CSS. In the presence of systemic signs or symptoms, it is important to rule out systemic vasculitis in order to start appropriate immune-modulatory treatment thereby avoiding unnecessary mortality. (*Eur J Ophthalmol* 2009; 19: 314-7)

KEY WORDS. Branch retinal vein occlusion, Central retinal artery occlusion, Churg-Strauss syndrome, Systemic vasculitis, Allergic granulomatosis, Hypereosinophilia

Accepted: September 4, 2008

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 pleuric 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

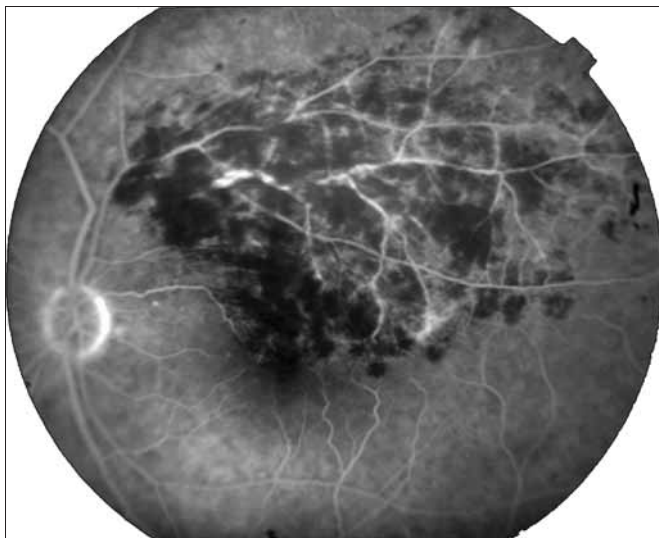


Fig. 1 - Fluorescein angiography of the left eye demonstrates small areas of retinal capillary nonperfusion in the distribution of the superotemporal vein and moderate intraretinal leakage of dye from branches of the occluded vein.

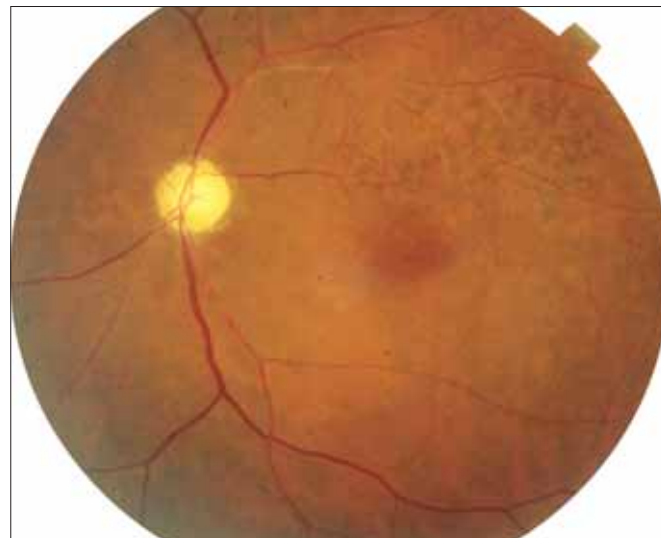


Fig. 2 - Color fundus photograph of the left eye shows optic disc atrophy, pigmented retinal scars due to superotemporal sector laser photocoagulation, as well as narrowed, sheathed retinal veins and arteries in the distribution of the occluded branch retinal vein.

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 in-

creased 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

vascular abnormality in various organ systems. They termed this condition “allergic granulomatosis and angiitis.” Histologically these “allergic granulomas” were composed of necrotic eosinophilic exudates, severe fibrinoid collagen changes, and granulomatous proliferation of epithelioid and giant cells. In 1990, the American College of Rheumatology (3) developed the following criteria for epidemiologic and therapeutic studies of CSS: asthma, eosinophilia greater than 10%, mono- or polyneuropathy, non-fixed pulmonary infiltrates, paranasal sinus abnormalities, and biopsy containing blood vessels with extravascular eosinophilia. 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 inflammatory 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 systemic vasculitis signs, including, after 1 year, central CRAO in the same eye. CSS is a leukocytoclastic systemic 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 followed by eosinophilic pneumonia. Since CSS invariably involves the lungs but affects a wide variety of other tissues 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 disease. 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 syndrome (11), hyperhomocysteinemia (12), lupus erythematosus (13), type 2 diabetes (14), interferon therapy (15), acquired immunodeficiency syndrome (16), thrombotic thrombocytopenic purpura (17), systemic non-Hodgkin lymphoma (18), acute lymphoblastic leukemia (19), infective endocarditis (20), and hormone replacement therapy (21). Knowing these causes, it is essential to make an early diagnosis and to not miss any underlying disease.

CONCLUSIONS

BRVO and CRAO can occur in the same eye in CSS. In the presence of systemic signs or symptoms, it is important to rule out systemic vasculitis in order to start appropriate 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 systemic 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 underlying systemic disease through a careful screening.

The authors received no financial support and have no proprietary interest in the subject or materials discussed in this article.

Reprint requests to:
Gabriella De Salvo, MD
Università degli Studi di Palermo
DINEC–Dipartimento di Neuroscienze Cliniche–Sezione di Oftalmologia
Via Liborio Giuffrè 13
90127 Palermo, Italy
gabrydsl@tiscali.it

REFERENCES

1. Jennette JC, Falk RJ, Andrassy K, et al. Nomenclature of systemic vasculitides: proposal of an International Consensus Conference. *Arthritis Rheum* 1994; 37: 187-92.
2. Churg J, Strauss L. Allergic granulomatosis, allergic angiitis, and periarteritis nodosa. *Am J Pathol* 1951; 27: 277-301.
3. Masi AT, Hunder GG, Lie JT, et al. The American College of Rheumatology 1990 criteria for the classification of Churg–Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum* 1990; 33: 1094-100.
4. Robin JB, Schanzlin DJ, Meisler DN, et al. Ocular involvement in the respiratory vasculitides. *Surv Ophthalmol* 1985; 30: 127-40.
5. Rosenthal G, Schnek M, Lifshitz T. Branch retinal vein occlusion in Churg–Strauss syndrome. *Clin Exp Ophthalmol* 2002; 30: 381-2.

6. Udono T, Abe T, Sato H, et al. Bilateral central retinal artery occlusion in Churg–Strauss syndrome. *Am J Ophthalmol* 2003; 136: 1181-3.
7. Hamann S, Johansen S. Combined central retinal artery and vein occlusion in Churg–Strauss syndrome: case report. *Acta Ophthalmol Scand* 2006; 84: 703-6.
8. Lanham JG. Churg-Strauss syndrome. *Br J Hosp Med* 1992; 47: 667-73.
9. Alp MN, Türkçü FM, Tola M, Kural G. Combined central retinal vein and branch retinal artery occlusion in a patient with coexisting heterozygous factor V and prothrombin 20210A mutations. *J Retina-Vitreous* 2007; 15: 133-6.
10. Tavola A, D'Angelo SV, Bandello F, et al. Central retinal vein and branch artery occlusion associated with inherited plasminogen deficiency and high lipoprotein(a) levels: a case report. *Thromb Res* 1995; 80: 327-31.
11. Levy J, Baumgarten A, Rosenthal G, Rabinowitz R, Lifshitz T. Consecutive central retinal artery and vein occlusions in primary antiphospholipid syndrome. *Retina* 2002; 22: 784-6.
12. Ozdek S, Yülek F, Gürelik G, Aydın B, Hasanreisolu B. Simultaneous central retinal vein and retinal artery branch occlusions in two patients with homocystinemia. *Eye* 2004; 18: 942-5.
13. Leibovitch I, Goldstein M, Loewenstein A, Barak A. Combined central retinal artery and vein occlusion in a patient with systemic lupus erythematosus. *Rheumatology* 2001; 40: 1195-6.
14. Arikan G, Saatci AO, Soylev MF, Kocak N. Bilateral combined cilioretinal artery and central retinal vein occlusion. *Ann Ophthalmol* 2006; 38: 135-7.
15. Rubio JE Jr, Charles S. Interferon-associated combined branch retinal artery and central retinal vein obstruction. *Retina* 2003; 23: 546-8.
16. Wen F, Chen X, Li H, Liao R, Wu D. Bilateral central retinal vein occlusions combined with artery occlusions in a patient with acquired immune deficiency syndrome. *Yan Ke Xue Bao* 2002; 18: 27-9.
17. Schwartz SG, Hickey M, Puliafito CA. Bilateral CRAO and CRVO from thrombotic thrombocytopenic purpura: OCT findings and treatment with triamcinolone acetonide and bevacizumab. *Ophthalmic Surg Lasers Imaging* 2006; 37: 420-2.
18. Shukla D, Arora A, Hadi KM, Kumar M, Baddela S, Kim R. Combined central retinal artery and vein occlusion secondary to systemic non-Hodgkin's lymphoma. *Ind J Ophthalmol* 2006; 54: 204-6.
19. Chan WM, Liu DT, Lam DS. Images in haematology. Combined central retinal artery and vein occlusions as the presenting signs of ocular relapse in acute lymphoblastic leukaemia. *Br J Haematol* 2005; 128: 134.
20. Kato T, Takeda Y, Matsuyama S, Mishima HK. Photo essay: combined occlusion of the central retinal artery and vein in a pediatric patient secondary to infective endocarditis. *Arch Ophthalmol* 2001; 119: 1868-9.
21. Murray DC, Christopoulou D, Hero M. Combined central retinal vein occlusion and cilioretinal artery occlusion in a patient on hormone replacement therapy. *Br J Ophthalmol* 2000; 84: 549-50.

Copyright of European Journal of Ophthalmology is the property of Wichtig Editore and its content may not be copied or emailed to multiple sites or posted to a listserv without the copyright holder's express written permission. However, users may print, download, or email articles for individual use.