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SHORT COMMUNICATION

Case report

Hemiretinal vein occlusion in Wegener's granulomatosis

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PURPOSE. *To report a case of hemiretinal vein occlusion in a patient with Wegener's granulomatosis.*
METHOD. *Case report.*
RESULTS. *Retinal vein occlusion may occur in patients with Wegener's granulomatosis even in the absence of active inflammation affecting the retinal venules.*
CONCLUSIONS. *Wegener's granulomatosis should be considered in the diagnostic workup of young patients with major retinal vascular occlusion and concurrent history of pulmonary and/or renal disease. (Eur J Ophthalmol 2003; 13: 722-5)*

KEY WORDS. *Retinal vasculitis, Retinal vein occlusion, Wegener's granulomatosis*

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INTRODUCTION

Wegener's granulomatosis is characterized by necrotizing granulomas of the upper and lower respiratory tract together with glomerulonephritis (1). Widespread disseminated vasculitis involving small arteries and veins has also been described. The retinal vessels also fall into the category of small vessels and their involvement in this disease is thought to be due to focal necrotizing vasculitis (1). We report a case of hemiretinal vein occlusion without any evidence of associated intraocular inflammation in a known case of Wegener's granulomatosis.

Case report

A 40-year-old man with a known history of chronic obstructive airway disease for the past 2.5 years had presented with shortness of breath of 15 days duration in March 2000. Bilateral rhonchi were present and the patient was started on bronchodilators. One month later the patient presented with hemoptysis and fever. The patient also had a history of arthralgia. A chest x-ray showed parenchymal infiltrates in both lungs with middle lobe consolidation on the right side. A contrast enhanced computerized tomogram (CECT) showed cavitatory lesion on the right with multiple bilateral nodules. Sputum for acid-fast bacilli was negative. His blood urea and serum creatinine were also deranged. A PR3 ANCA was done, which came out positive (10.8 EU/ml; normal <7.0). A diagnosis of Wegener granulomatosis was thus made. There was no upper respiratory tract involvement. CECT of the abdomen showed a splenic infarct. The patient was given pulse cyclophosphamide and dexamethasone and then put on oral steroids. His renal function gradually improved and the chest x-ray showed resolution of the infiltrates. Initially the patient was given oral prednisolone 1 mg/kg body weight that was gradually tapered and for the past 3 months the patient was on 20 mg prednisolone OD. The patient was found to have

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developed hypertension during the course of his therapy and was started on antihypertensives on which his blood pressure was controlled. The patient was not a diabetic and was a nonsmoker.

After 1 year, the patient presented with a history of painless diminution of vision in the right eye for the past 2 months. His visual acuity was 6/18 in the right eye and 6/6 in the left eye. The anterior segment was normal. The intraocular pressure in both eyes was 17.3 mmHg. In the right eye the media was clear, and the inferior half of the disc was obscured by hemorrhages. The inferior division of the retinal vein was dilated and tortuous and there were florid hemorrhages confined to the inferior half of the fundus extending from the disc to the equator. There were also a few soft exudates (Fig. 1). The other eye fundus was normal. Fluorescein angiography showed multiple areas of blocked fluorescence corresponding to the hemorrhages, dilatation of the capillary bed, some microaneurysms, and small areas of capillary nonperfusion. The dilatation and tortuosity of the inferior retinal vein was further highlighted on the fluorescein angiogram. There was no evidence of retinal neovascularization or macular edema (Fig. 2). A diagnosis of right hemiretinal vein occlusion of the nonischemic type was made. Hematologic investigations revealed hematocrit 45%, hemoglobin 12 gm%, total leukocyte count 7300, differential leukocyte count polymorphs 66, and lymphocytes 34.

As the vascular occlusion was of the nonischemic type and there was no significant macular edema, the therapy of the patient was not changed and he was asked to follow up every 3 months. The features of vascular occlusion slowly subsided. At 6-month follow-up the patient showed significant resolution of retinal hemorrhages.

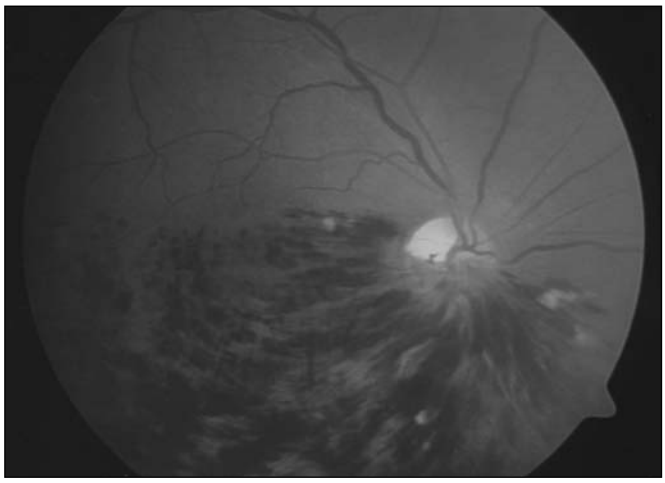

Fig. 1 - *Fundus photograph showing florid hemorrhages confined to the inferior half of the retina.*

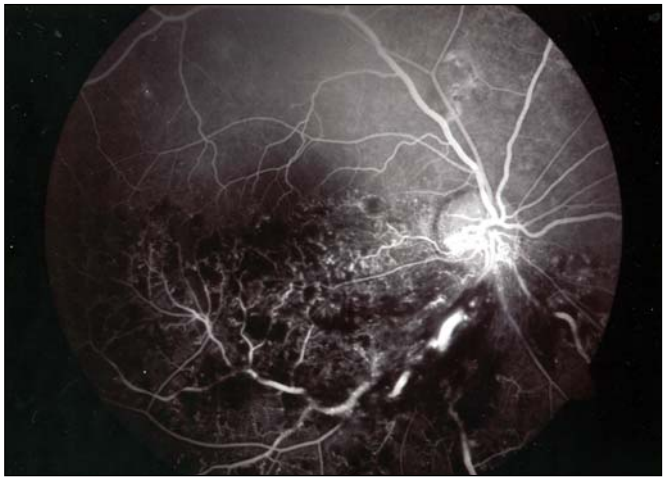

Fig. 2 - *Venous phase fluorescein angiogram showing blocked fluorescence corresponding to the hemorrhages and absence of neovascularization and edema.*

DISCUSSION

Wegener's granulomatosis involves the eye in 28 to 58% of cases, with ocular involvement being the primary manifestation in up to 16% of cases (2). The disease commonly involves the orbit, causing proptosis. Other common manifestations are marginal keratitis and necrotizing scleritis. Retinal involvement is rare, with its reported incidence varying from 1 to 13% in various case series (1, 2). Chorioretinitis, macular edema, retinal epithelial pigmentary changes, retinitis with cotton wool spots, acute retinal necrosis, peripheral vasculitis, central retinal artery occlusion, and exudative retinal detachment are some of the retinal manifestations described (3).

On a MEDLINE search using the key words Wegener's granulomatosis and retinal vein occlusion we found only two reports of major retinal vein occlusion. Of these, one report including three cases of retinal vein occlusion was published in a non-ophthalmic journal and the other report was a retrospective diagnosis made after renal biopsy and a review of ocular histopathol-

TABLE I - CLINICAL DETAILS OF PATIENTS WITH WEGENER'S GRANULOMATOSIS WITH MAJOR VASCULAR OCCLUSION OF RETINA

No.	Author	Year	No. of cases	Diagnosis	Renal disease	Lung disease	Upper respiratory tract	Arthralgia	Vision	Other ocular features	ANCA
1	Spalton et al	1981	1	CRVO with NVG	Positive	-	-	-	PI negative	Keratitis other eye	No record
2	Stavrou et al	1993	3	CRVO (2) BRVO (1)	All three cases	One case	One case	No record	6/18 - 6/6	Episcleritis in two cases	All three cases positive
3	Venkatesh et al (current case)	2003	1	Hemi retinal vein occlusion	Positive	Positive	-	Positive	6/18	-	Positive

CRVO = Central retinal vein occlusion; NVG = Neovascular glaucoma; BRVO = Branch retinal vein occlusion.

ogy. To our knowledge, hemiretinal vein occlusion has not been reported in this disease.

All three patients of the case series by Stavrou et al were of the complete type of Wegener granulomatosis; i.e., they had renal involvement (4). Involvement of lung and upper respiratory tract and presence of arthralgias were variable features. The visual acuity of the patients ranged from 6/18 to 6/6, which implies that the vascular blocks were probably nonischemic in nature. The other patient described by Spalton et al had one eye enucleated for thrombotic glaucoma (1). Eighteen months later the patient developed malaise, high erythrocyte sedimentation rate, and marginal keratitis and uveitis in the other eye. His renal biopsy showed proliferative glomerulonephritis and a review of ocular pathology of the enucleated eye showed perivascular leukocytic infiltration in the bulbar conjunctiva, episclera, and anterior sclera. There was no inflammation of the retinal vessels. A diagnosis of Wegener granulomatosis was considered based on the renal biopsy and ocular histopathology.

Our patient had Wegener's granulomatosis and was positive for PR3 ANCA. He developed hemiretinal vein occlusion without evidence of inflammation of any retinal vessels. In the previously reported cases of retinal vascular occlusion in this disease there also is no description of presence of retinal vessel inflammation. Presence of any other evidence of intraocular inflammation such as vitreous cells or retinal exudation is also lacking in these reports. The available histopathology of the case report by Spalton et al also does not reveal any retinal vessel inflammation (1). Therefore, although the pathology in this disease is a focal necrotizing vasculitis, the absence of intraocular inflammation in the reported cases of major retinal vascular occlusion could be due to the inflammatory vascular block occurring in the laminar or retrolaminar portion of the optic nerve head that may not be clinically evident. In contrast to the previously reported cases, our case had more widespread involvement, including that of the lung and joints, apart from the kidney and the eye.

Our patient developed hypertension during the course of his therapy with steroids. However, as the time between development of ocular symptoms and the onset of hypertension was only around 6 months, we do not believe that the vascular block was due to hypertension. In addition, the patient's fundus did not reveal any hypertensive changes.

In conclusion, retinal vascular occlusion can occur in patients with Wegener's granulomatosis even in the absence of clinically detectable intraocular inflammation. In young patients with major retinal vascular occlusion, a history for concurrent pulmonary and renal symptoms must be elicited, as their concurrent involvement would suggest a diagnosis of Wegener's granulomatosis. Early diagnosis and treatment is important in reducing the high mortality and morbidity associated with this disease.

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