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SHORT COMMUNICATION

Case report

Central serous retinopathy associated with Weber-Christian disease

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ABSTRACT: Purpose. To report an association between central serous retinopathy and Weber-Christian disease.
Methods. A case report.
Results. Central serous retinopathy was noticed in a patient with Weber-Christian disease possibly secondary to vasculitis involving the choriocapillaris.
Conclusions. Central serous retinopathy may be associated with an uncommon systemic condition like Weber-Christian disease. (Eur J Ophthalmol 1999; 9: 139-41)

KEY WORDS: Central serous retinopathy, Weber-Christian disease, Panniculitis

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INTRODUCTION

Weber-Christian disease (relapsing febrile nodular non-suppurative panniculitis) is an inflammatory condition of the adipose tissue occurring usually in middle aged women. The patients present with tender nodules most commonly on the lower limbs, which may be associated with systemic symptoms. The condition may resolve with symptomatic treatment. However treatment with systemic steroid and other immunosuppressive agents may be required to control the recurrent episodes. Ocular involvement is rare. We describe a patient who developed visual disturbance due to central serous retinopathy (CSR).

Case Report

A 51-year-old man who suffered from Weber-Christian disease was seen in the ophthalmology department in September 1997, with a history of visual disturbance in his left eye for 1 month. His visual acuity was 6/5 OD and 6/18 OS. Anterior segment of both eyes was normal. Fundus of the right eye was normal. His left eye showed a serous detachment of the macula, one and a half disc diameters in size. A central scotoma was demonstrable with Amsler chart. A clinical diagnosis of resolving CSR was made and fluorescein angiography performed. Choroidal and the arterial phase of fluorescein angiography showed an area of choroidal hypo-perfusion around the optic disc and the macula (Fig. 1). Multiple leaks that appeared in the early phase progressed during the course of the angiography fitting the description of 'ink-blot' leakage. There were two spots at the inferior margin of the CSR and two other near the supero-temporal arcade (Fig. 2). The right eye was normal. He was followed up without treatment. The CSR resolved spontaneously in four months and his visual acuity improved to 6/6 with residual retinal pigment epithelial changes at the macula. The diagnosis of Weber-Christian disease had been made in November 1993 after a biopsy of a nodule on his left thigh showed the characteristic features including foam cells. He was treated with oral prednisolone 40 mg per day that was tapered off to a maintenance dose of 5-10 mg daily. This was increased usually to 40 mg to control recurrent episodes which were 2-3 per year. Azathio-

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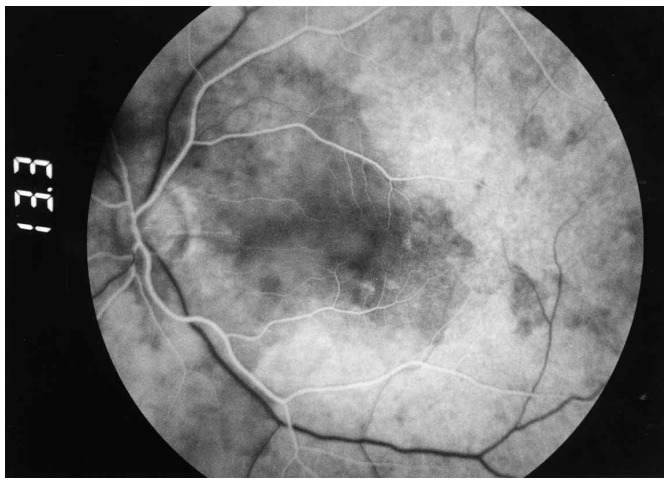

Fig. 1 - *Fundus fluorescein angiogram of the left eye showing choroidal hypo-perfusion around the optic disc and in the macula.*

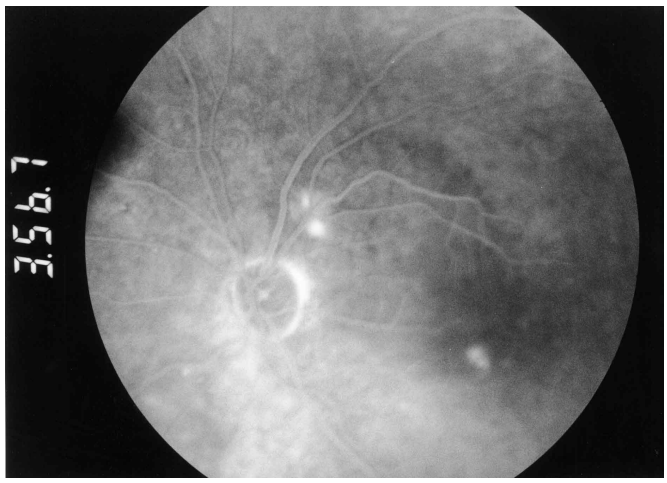

Fig. 2 - *Late phase of fundus fluorescein angiogram of the left eye showing the multi focal 'ink-blot' leaks.*

prine, methotrexate and cyclosporin were used as steroid sparing agents since February 1996. He also received supplementary treatment with folic acid and a biphosphonate. Nifedipine had been used to control his systemic hypertension since November 1996. Two months prior to the onset of visual symptoms he had a severe recurrence of panniculitis, associated with pleuro- pericardial effusion. This was controlled with prednisolone 40 mg, methotrexate 7.5 mg per week and cyclosporin 200mg per day in divided doses. Two weeks prior to the onset of visual symptoms he had another recurrence, which was treated similarly.

DISCUSSION

Pfeiffer first described the clinical and histopathologic characteristics of Weber-Christian disease in 1892 although it was not so named until 1936. In 1925, Weber named this condition relapsing nodular non-suppurative panniculitis. Christian added the word 'febrile' in 1928 (1). Ocular involvement is rare in this condition.

Klein (2) reported a 63-year-old woman who suffered recurrent episodes of anterior uveitis. She also developed an episode of central choroiditis, which was treated with adrenocorticotrophic hormone. Klein suggested the possibility of antigen-antibody reaction at the site of the vascular bed being responsible for this. Frayer et al (3) found ocular and adnexal changes in 3 patients with Weber-Christian disease. Two patients had nodular lesions adjacent to the limbus and the third had an eyelid lesion along with nodular episcleritis.

Freeman (4) reported macular involvement causing visual loss in a 62-year-old woman. She had bilateral macular haemorrhage and tortuous retinal arterioles in the macular region. Fluorescein angiography showed leakage at both maculae. The haemorrhages had subsided and visual acuity returned to 6/9 in 2 weeks. Recently Cook and Kikkawa (5) described a patient who presented with proptosis that was initially treated as orbital inflammatory disease. The patient presented several weeks later with proptosis in the other eye and for the first time associated with features of Weber-Christian disease. The proptosis was thought to be due to panniculitis of the orbital fat. Histopathologic features of Weber-Christian disease (1,6) include lobular and septal panniculitis, vasculitis along with the characteristic foam cells. Panush et al (1) in their detailed review of 15 patients with Weber-Christian disease, found that vasculitis was present in 32% of their patients. Eckstein et al (7) reported two cases of CSR associated with systemic lupus erythematosus. Although their patients did not show any abnormality of choroidal circulation, they discussed about a mechanism in which vasculitis of choriocapillaris may result in reduced choroidal blood flow with the breakdown of outer blood-retinal barrier leading to subretinal leakage of fluid. The presence of the choroidal hypo-perfusion and residual retinal pigment epithelial changes in our patient

supports the above theory and may explain the pathogenesis of CSR in him. Indocyanine green angiography if available to us could have provided more information on the choroidal circulation (8). Systemic corticosteroid usage may be another factor to be considered in the development of CSR in our patient. High dose of systemic prednisolone (100 mg) has been associated with development of CSR (9). Our patient had never received such a high dose but had been maintained on low dosage of prednisolone along with steroid sparing immunosuppressive agents. Nevertheless this association should be borne in mind. CSR typically occurs in men in the age group of 20- 45 years of age. Fortuitous occurrence of CSR and Weber-Christian disease is less likely here as our patient is not in the typical age group. The onset of visual symptoms was also preceded by a recurrence of panniculitis. Thrombotic microangiopathy has been reported in patients who received cyclosporin following transplantation. However the pathogenesis of this condition could be multi factorial, as successful reinstitution of cyclosporin therapy without recurrence of thrombotic microangiopathy has been reported (10). Its role in our patient is uncertain. We believe that the occurrence of CSR in our patient is related to activity of his Weber Christian disease itself. Although the visual prognosis for CSR is usually good, poor visual outcome may result from uncommon complications such as macular oedema, choroidal neovascularisation and retinal pigment epithelial atrophy (11). CSR may be an uncommon association with an uncommon systemic condition like Weber-Christian disease. This is to our knowledge the first report of such an association.

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