Retinal periphlebitis in ulcerative colitis

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Summary: A man presented with unilateral visual blurring associated with bilateral retinal periphlebitis which was felt to be a complication of his biopsy-proven active ulcerative colitis. Retinal periphlebitis has been associated rarely with some forms of colitis but we can find no report of its occurrence in association with ulcerative colitis although other ocular inflammatory disorders are well recognized.

Introduction

Retinal periphlebitis is a well recognized manifestation of many diverse systemic diseases. Ocular disease is a well established extraintestinal manifestation of ulcerative colitis. However, to our knowledge retinal periphlebitis has not been described before in association with ulcerative colitis.

Case report

A 33 year old Indian man presented with three episodes of acute onset partial visual obscuration in his right lower visual field lasting 12, 2 and 5 hours respectively over a 10-day period. On the fourth occurrence the visual field defect has remained for the present follow-up period of 9 months. On examination his visual acuity was 6/18 in the right eye and 6/6 in his left eye. There was a right infero-nasal field defect. The optic discs were swollen and the retinal veins dilated and tortuous with peripapillary haemorrhages and cotton wool infarcts extending to the periphery. An ischaemic lesion supero-lateral to the right disc was felt to be responsible for his persisting field defect (Figure 1). There was no abnormality of the cornea, iris, lens and vitreous body in either eye. General examination was otherwise normal.

Ulcerative colitis had been diagnosed 4 years previously on a history of bloody diarrhoea and confirmed endoscopically with biopsy. His clinical course had a relapsing remitting pattern of bloody diarrhoea responsive both to steroids and mesala-

Figure 1 Right optic disc showing disc swelling, retinal venous dilatation and peripapillary haemorrhages with cotton wool infarcts.

zine. On this presentation his colitis was symptomatically dormant on maintenance mesalazine 1.6 g per day, prednisolone 7.5 mg per day and hydrocortisone acetate suppositories 125 mg at night. Any reduction below this regimen lead to a symptomatic relapse of his colitis.

Retinal periphlebitis was diagnosed on fluorescein angiography. No cause for the fundal appearances could be found on general investigation and review of his bowel condition showed active ulcerative colitis. He was treated with prednisolone 30 mg per day with an improvement in his right visual acuity to 6/6 over a 4 month period. However, there has been no change in his visual field defect in the current follow-up period. His bowel condition remains symptomatically quiescent on the same drug regimen.

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Investigations

Full blood count and a full clotting screen were normal. Serum viscosity was normal and haemoglobin electrophoresis showed A and A2 only. Serum biochemistry and protein electrophoresis were unremarkable. The cerebrospinal fluid was acellular with a normal protein and opening pressure. Immunoglobulin titres, cardiolipin IgG, C-reactive protein, C3 and C4 were also unremarkable. Serological titres for toxoplasma, brucella, syphilis, hepatitis B and Epstein-Barr virus showed no elevation suggestive of causality.

Fluorescein angiography (Figure 2) showed venous leakage in both eyes and areas of retinal ischaemia; one corresponding to the lesion mentioned previously. Computed tomographic brain scan with contrast enhancement was normal and venous digital subtraction cranial angiography showed no evidence of a venous thrombosis or any other abnormality. Chest X-ray was normal. Sig-moidoscopic review of his bowel revealed active mucosal inflammation. Heavy focal inflammation with crypt abscesses was seen histologically indicative of active ulcerative colitis.

Discussion

Ocular disease in association with ulcerative colitis was first described by Crohn in 1925 when he recorded two cases of corneal inflammation and conjunctivitis with ulcerative colitis.¹ It is now a seldom but well established extraintestinal complication with an incidence of 3.5–11.8% in various large series of inflammatory bowel

Figure 2 Peripheral retina in the late phase of fluorescein angiography showing patchy leakage of fluorescein from retinal periphlebitis.

disease.^{2–4} It can precede, coincide with, or follow the overt manifestations of the colitis with a tendency to flare during exacerbation of colitis.⁴ Ocular features commonly include conjunctivitis, episcleritis, iritis, corneal ulceration and keratitis.³ Central serous chorioretinopathy, occasionally with bullous retinal detachment, rarely occurs in association with ulcerative colitis.⁵ Retinal artery and branch artery occlusions also rarely occur.⁶

Retinal periphlebitis is characterized by peripheral retinal venous darkening and irregularity. The vein is generally dilated with narrowed sites along its course. Progression is marked by white periphlebitic foci which appear as scattered cuffing along venous segments in a random distribution. Retinal blot haemorrhages appear in the area of venous drainage. A number of diverse inflammatory conditions start as retinal periphlebitis or develop it as a secondary feature. In association with recurring vitreous haemorrhage an idiopathic form is known as Eales' disease.⁷ It is a well observed secondary phenomenon in uveitis. Peripheral retinal periphlebitis is also associated with multiple sclerosis.⁸ Other inflammatory diseases including sarcoidosis, syphilis, brucellosis and cryptococcal infection have rarely been associated.⁷

Retinal periphlebitis has a reported incidence as seen by fluorescein angiography of 13% in Behçet's disease.⁹ The colitis in Behçet's disease is often mistaken histologically as that of ulcerative colitis.¹⁰ However, the broader clinical context provides easy differentiation; the absence of mucosal, skin and uveal involvement in this case, besides the legion of lesser features as defined by Mason and Barnes, excludes Behçet's as a likely diagnosis.¹¹

Immunological mechanisms involved in the pathogenesis of ulcerative colitis are complex.^{12,13} An exact immunological relationship with any of the recognized ocular manifestations of ulcerative colitis is unknown. However, there is evidence that anterior non-granulomatous uveitis may be a manifestation of an exposure to a colonic antigen resulting in an immune complex type inflammation.¹⁴ The mural inflammatory changes seen in retinal periphlebitis appear to be initiated by immune complex deposition on the vessel wall. Plasma cells containing IgG and IgA have been found throughout chronically inflamed vessels with C3, IgG and IgA demonstrable immunohistochemically in the endothelium of episcleral veins.¹⁵

In this instance the fluorescein angiographic appearance was characteristic of retinal periphlebitis. Accordingly the diagnosis of his colitis was reviewed only to be confirmed histologically as ulcerative colitis, though with greater activity. We have found no previous report describing the association of retinal periphlebitis with ulcerative colitis.

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