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Sir,

Uveitis in a patient with common variable immunodeficiency

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Common variable immunodeficiency (CVID) is the most common primary immunodeficiency. It can occur at any age, and is characterised by recurrent bacterial infections, hypogammaglobulinaemia, and deficient antibody responses. It is thought to be acquired, and auto-immune disorders can complicate the clinical picture. Chronic uveitis is often thought to have an auto-immune basis, and we report here a CVID patient who developed multifocal choroiditis with pan-uveitis.

Case report

In August 1997, a 55-year-old emmetropic man who worked as a printer presented with a 3-month history of deteriorating right vision. In 1981 he had a splenectomy for a presumed lymphoma. Subsequent histology and immunoglobulin levels, and a retrospective history of recurrent infections, revised the diagnosis to granulomatous antibody deficiency (GAD), a variant of common variable immunodeficiency (CVID). Since then he has been receiving intravenous immunoglobulin and continuous penicillin prophylaxis.

Unaided visual acuity was 6/18 right and 6/6 left. Inflammatory flare and cells were noted in the anterior chamber and vitreous cavity on the right, and fundal examination revealed numerous peripheral chorio-retinal scars, as well as cystoid macular oedema (Figure 1). There was no evidence of retinal vasculitis, and the left eye was normal except for fine macular drusen.

He was treated with an orbital floor injection of depomedrone 40 mg, and topical steroid eye drops were also started. One month later there had been little clinical response, and the steroid injection was repeated. The uveitis and macular oedema persisted however, and a small inflammatory vitreous 'snowball' was also noted at this time. Further (systemic) immunosuppression was not felt to be justified because of his hypogammaglobulinaemia and previous splenectomy, and also because his other eye was normal.

He was seen again 18 months later when the right acuity had dropped to counting fingers level, and a prominent submacular scar had developed (Figure 2). Moderate intra-ocular inflammatory activity persisted, and similar though milder inflammatory change was now also documented on the left. Vision on that side was, however, well maintained at 6/6, and the clinical picture has remained unchanged since then.

Routine full blood count and plasma viscosity measurements have been repeatedly normal, and serum angiotensin converting enzyme levels have been in the normal range. Thoracic CT scanning had been carried out in 1999 and had revealed bronchiectatic change consistent with his known CVID. It is not possible to test for serology to toxoplasma and toxocara as this would only reflect the serology of the immunoglobulins administered.

Comment

CVID is the most common primary immunodeficiency, and is characterised by recurrent bacterial infections, hypogammaglobulinaemia, and deficient antibody responses. It can occur at any age, and is thought to be acquired: auto-immune disorders and auto-antibodies can complicate the clinical picture. Non-caseating granulomata mimicking sarcoidosis have also been described affecting the lungs, liver, spleen and skin, and appear to be responsive to steroid therapy.

Uveitis can be caused by infections such as syphilis, tuberculosis or toxoplasmosis, but is more commonly 'idiopathic' or associated with systemic diseases that are thought to have an auto-immune basis such as Behcets, systemic lupus erythematosus or Vogt–Koyanagi–Harada syndrome. In our patient the presence of scattered peripheral chorio-retinal scarring in addition to the uveitis suggests a diagnosis of multifocal choroiditis with pan-uveitis (MCP), a descriptive term used to characterise an ocular inflammatory disorder that is thought to have an auto-immune basis and that can be

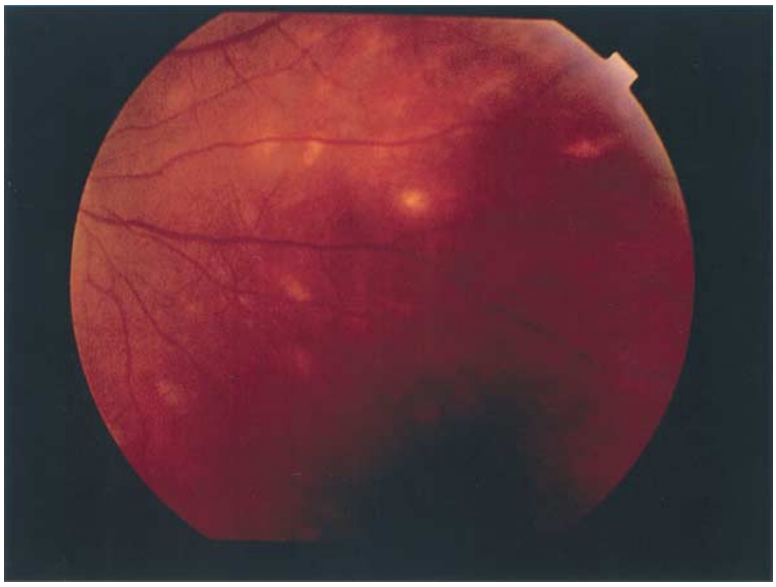

Figure 1 Fundus photo of nasal retina, right eye. Numerous small and atrophic chorio-retinal scars are visible.

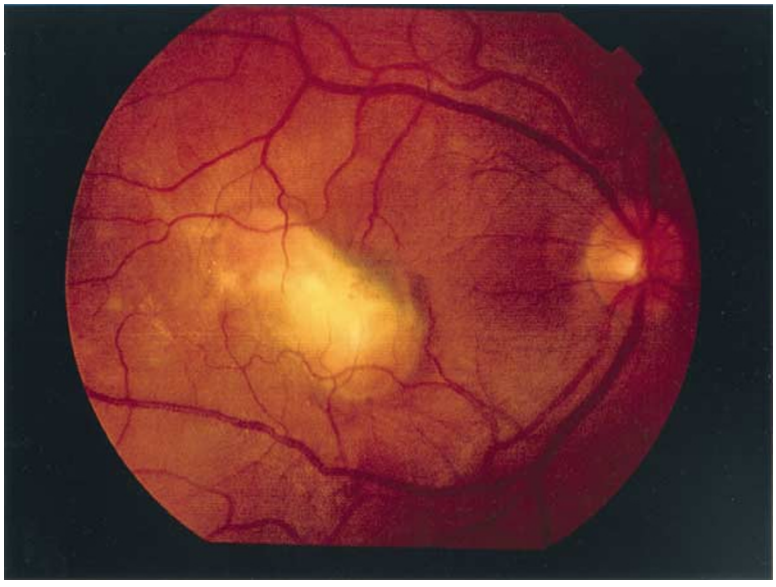

Figure 2 Fundus photo right eye, showing prominent white submacular scarring.

associated with vitiligo.[1] (Our patient has a small patch of vitiligo at the nape of his neck.) Response to steroids is variable, and high dose systemic treatment may be needed. The lack of clinical response to orbital floor injections of depomedrone in our patient is not uncommon.

Subretinal neovascularisation and consequent fibrovascular scarring is a recognised complication of MCP, and the prominent sub-macular scarring in our patient may be a result of this complication. It is, however, unusual to see such prominent fibrosis in a site apparently remote from any preceding inflammatory focus, and it is tempting to speculate that the subretinal mass is a choroidal granuloma. Such changes have previously been described in the central nervous system.[2]

MCP has been associated with sarcoidosis. In one study of 10 patients with MCP, all of whom underwent non-directed conjunctival biopsies. Non-caseating granulomata in the absence of acid-fast bacilli were

found in seven individuals.[3] Sarcoidosis may also complicate or mimic the clinical features of CVID.[4] However, we think it unlikely that sarcoidosis is present in this patient in view of the thoracic CT scan results, near normal serum angiotensin converting enzyme levels, and normal lymphocyte subset numbers.

It is probable that disturbed auto-immunity links CVID and MCP in this patient, and such an association has been suggested in a previous case report of a patient presenting with bilateral granulomatous anterior uveitis, optic disc swelling and multifocal areas of choroidal pallor.[5] Investigations led to the diagnosis of CVID, and the anterior uveitis responded to topical steroids. It is, however, not clear what happened to the optic disc and choroidal abnormalities. A further case report has linked CVID with uveitis following campylobacter enteritis,[6] though in this case the uveitis was not associated with peripheral chorio-retinal scars. Also, the uveitis was acute at onset, suggesting that it was predominantly anterior in location (iritis), and possibly secondary to an abnormal immune response following the enteritis itself (Reiters syndrome).[7]

Retinal vasculitis has also been linked with CVID in a case series of three children, all of whom had CVID and bilateral retinal vasculitis with optic nerve and macular oedema.[8] In two of these patients there was evidence of posterior uveitis as well. The authors point out that CD4+ cells of patients with CVID can produce normal amounts of interferon and may therefore be able to perpetuate an auto-immune response once initiated. In contrast to the above reports, in which the ophthalmic involvement occurred shortly after or even before the diagnosis of CVID, our patient's uveitis developed some 17 years after diagnosis, and throughout that time he had been receiving immunoglobulin treatment.

Patients with CVID are predisposed to a wide variety of illnesses that are thought to have an auto-immune aetiology. On the basis of this case report and others we suggest that MCP might be one such disease, and we would advocate prompt ophthalmic referral for any CVID patient experiencing blurred vision.

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Sir,

Mucocoele of the maxillary sinus and the eye

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Mucocoeles are common, expansile cyst-like lesions affecting the paranasal sinuses.[1] The majority occur in the frontal sinus, followed by ethmoid sinus, maxillary sinus, and sphenoid sinus. They grow gradually, expand in size, and rarely produce bony destruction of the sinus walls,[2] leading to orbital and ocular involvement. We report such a rare case.

Case report

A 69-year-old woman with 1-month history of right-sided facial numbness and constant aching was referred by her GP to see a neurologist. While waiting for the appointment, she developed transient double vision and mild right ptosis. She did not sustain any facial injury affecting the sinuses in the past, but gave the history of chronic right maxillary sinusitis and Caldwell–Luc