Idiopathic keratoconus in a patient with congenital rubella syndrome

Editor,—Keratoconus is very rarely found in patients with congenital rubella syndrome (CRS). The only known aetiology of keratoconus associated with CRS is eye rubbing.1 We report here a patient with CRS who developed idiopathic keratoconus.

CASE REPORT
A 24 year old woman with CRS was examined for progressive visual blurring of 2 years' duration in her left eye. The diagnosis of CRS was based on the maternal history of rash and fever in the first trimester of pregnancy, cataract, and microphthalmia in the right eye, bilateral rubella retinitis, nystagmus, deafness, congenital heart disease.

Available medical records dated from the time she was 1 year old. At that time, extracapillary cataract surgery was performed in the right eye. She had esotropia in the right eye and poor fixation. Apart from pigmentary changes in the fundus, the left eye was normal. A follow-up examination when she was 15 documented nystagmus, esotropia, and microphthalmia in her right eye, and bilateral rubella retinopathy. Visual acuity was RE light perception; LE 6/9 (with −4.0 −1.0 D sph). Two other examinations at the ages of 18 and 21 gave similar results.

On present examination, visual acuity with spectacles was LE 6/60. Retinoscopy revealed high, irregular myopic astigmatism with scissoring of the red reflex. Central keratometry showed irregular mires which could not be superimposed; 4 dioptrie irregular astigmatism was found. A conical reflection on the nasal cornea was obtained by shining a penlight from the nasal side (Rizzuti’s sign). Bulging of the lower eyelid on downgaze was also observed (Munson’s sign). Slit lamp examination revealed an inferiorly located ectatic protrusion of the central cornea with retinal and anterior stromal scars. As a result, the diagnosis of mild keratoconus was made. Treatment with a Softperm contact lens was performed and visual acuity recovered at 20/50.

Pseudouveitis as a manifestation of hyperlipidaemia

Editor,—We report a case of unilateral anterior pseudouveitis in a diabetic retinopathic eye, as a manifestation of hyperlipidaemia. High levels of lipids were detected in the aqueous humour and the anterior flare resolved only after successful control of diabetes and hyperlipidaemia was obtained.

CASE REPORT
A 44 year old man was referred for anterior uveitis not responsive to a 5 day topical steroid therapy. His past medical history was significant for non-insulin dependent diabetes mellitus, systemic hypertension, hyperlipidaemia, and obesity.

At presentation, the patient complained of pain and redness in his left eye. Visual acuity was 20/25 in the right eye and light perception in the left eye. Examination of the right eye disclosed only mild diabetic retinopathy. Slit lamp examination of the left eye showed hyperaemia of the conjunctiva, and an intense milky flare obstructing the view of the iris and the fundus (Fig 1). It was difficult to detect the presence of cells in the anterior chamber because of the flare. There were numerous keratic precipitates nor hypopyon or iris neovascularisation. Intraocular pressures in both eyes were within normal limits. B-scan ultrasonography was unremarkable.

Results of laboratory tests showed hyperglycaemia (24 mmol/l), a twofold increase in normal cholesterol levels (15 mmol/l), and a 22-fold increase in normal triglyceride levels (38 mmol/l). Results of laboratory examination including HLA typing, angiotensin converting enzyme, serologies for herpes simplex virus (HSV) and lues, purified protein derivative (PPD) skin test, and chest and saccroiliac x-rays, were not contributory. A β and pre-β lipoproteinaemia pattern was established by serum electrophoresis. Analysis of the aqueous humour of the left eye disclosed no cells, and high levels of proteins (32 g/l), cholesterol (3.4 mmol/l), and triglycerides (4.9 mmol/l).

Visual acuity recovered at 20/50 and the flare resolved after lipidemia and diabetes had been controlled by oral fibrates and insulin, respectively. Fundus examination of the left eye showed mild keratic neuropathy including microaneurism, hard exudates, and many paravascular retinal haemorrhages. Six weeks later, fundus examination of the left eye showed evidence of central venous occlusion (CRVO).

COMMENT
Although clinical presentation including pain, peribulbar conjunctival hyperaemia, and anterior chamber flare, was consistent with acute anterior uveitis, many features favoured a strong relation between the metabolic disorders and the occurrence of the flare. These clues included the milky appearance of the flare, the results of anterior chamber paracentesis, the lack of response to topical drugs, and a 22-fold increase in normal triglyceride levels.
Accepted for publication 15 April 1998

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Accepted for publication 15 April 1998

Letters

Detection of anticytomegalovirus antibody synthesis in the anterior chamber in Vogt–Koyanagi–Harada syndrome

Editor,—Vogt–Koyanagi–Harada (VKH) syndrome has long been suspected to be a consequence of autoimmunity or hypersensitivity against melanocytes. Nevertheless, to date no certain aetiological factors have been reported.

CASE REPORT

A previously healthy 30 year old Portuguese woman was admitted complaining of progressive visual loss in both eyes, over 2 weeks, associated with headaches and pain on eye movements and meningeum. Symptoms appeared 24 hours after the beginning of a flu-like syndrome. The patient had no past history of ocular trauma or surgery. On examination, the visual acuity was 20/100 in both eyes. Slit-lamp examination revealed 2+ cells in both anterior chambers with an uncosed partial vein occlusion or an episode of transient iridocyclitis rapidly cured by topical steroid therapy.

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Another mechanism by which microbial organisms can induce ocular autoimmunity is by the presence of homology between some sequences of their proteins and the sequence of uveitogenic determinants of ocular antigens. Recently, it has been shown that there is a similarity between self and foreign antigens called “molecular mimicry”; may induce a cross reaction with one or more self antigens sharing determinants with the foreign agents; this suggests that infectious agents can act as a trigger of organ specific autoimmunity in predisposed individuals. Among presumed triggers that could initiate the immune process, viruses may play a role in the pathogenesis of VKH syndrome. Indeed, various authors have reported the presence of Epstein-Barr virus genome in the cerebrospinal fluid or in the vitreous of patients with VKH syndrome, although it is difficult to establish the exact role of ubiquitous viruses in specific diseases. Our patient had an anti-CMV antibody synthesis in the anterior chamber and an acute CMV infection.

This case illustrates the possible implication of CMV in the aetiology of VKH syndrome; nevertheless, other known or unknown viruses may be involved in the pathogenesis of this syndrome in predisposed individuals.

Presented at the Fourth International Symposium on Uveitis, October 1997, Yokohama, Japan.

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Accepted for publication 21 April 1998


Bilateral small retinal infiltrates during rickettsial infection

EDITOR—Rickettsiae may cause heterogeneous retinal manifestations, the most characteristic of which are vascular lesions, periphlebitis, retinal haemorrhages and oedema, central retinal thrombosis, and papilloedema. We present the first case in which rickettsiae led to small retinal white infiltrates.

CASE REPORT
A previously healthy 25 year old woman had a fever up to 40°C of unknown origin during a trip to Queensland, Australia in August. Five days after the onset of fever a generalised maculopapular rash and a black lesion on the forehead (eschar) were visible, without a known history of a tick bite. The patient was transferred to our hospital with the signs of general illness. Extensive examination for infectious disease including eye examination was initiated.

Visual acuity was 20/25 in both eyes. Slit lamp examination revealed bilateral conjunctival hyperaemia. Moderate inflammation of
the virulence in both eyes was visible. Fundus examination disclosed bilateral small white infiltrates within the neurosensory retina, practically without involvement of the choroid, mainly in the posterior pole (Fig 1). In the right eye, a tiny well defined haemorrhage near the temporal inferior artery was seen. Fluorescein angiography of both eyes showed punctuate hypofluorescence corresponding to the white infiltrates (Fig 2), staining of the optic discs, but no signs of vasculitis. Indocyanine green (ICG) angiogram, visual field examination, and colour tests were unremarkable in both eyes.

Results of the following laboratory tests were pathologic: erythrocyte sedimentation rate (86 mm in the first hour), platelets (135 × 10^9/l), CRP (15 mg/dl), GOT (165 U/l), GPT (127 U/l), GGT (49 U/l), and LDH (527 U/l). Abdominal ultrasound sonography revealed an enlarged spleen (160 mm). The main serological finding was a definite increase of the Weil-Felix reaction from 1:160 (OX19, OX2) to 1:2280 (OX 19, OX2) and 1:160 (OX K) to 1:1280 (OX 19, OX2) and 1:160 (OX K) within 1 week. Serology for brucella, salmonella, Yersinia enterocolitica, Leisteria monocytogenes, Mycoplasma pneumoniae, Francisella tularensis, Mycobacterium tuberculosis, Coxiella burnetii, Borrelia burgdorferi, Toxoplasma pallidum, hepatitis A, B, C, HIV, rubella virus, rubella virus, Epstein–Barr virus, cytomegalovirus, rubella virus, rubella virus, varicella zoster virus, cytomegalovirus, herpes simplex I and II, Candida albicans, Cryptococcus neoformans, Aspergillus, toxoplasmosis, and chlamydia were all negative.

During antibiotic treatment with doxycycline 200 mg once daily for 14 days and clarithromycin 500 mg twice daily for 5 days, all clinical symptoms subsided and the Weil-Felix reaction decreased to 1:640 (OX 19, OX2) and 1:80 (OX K). The visual acuity increased to 25/20 in both eyes. Four weeks after this treatment all laboratory tests were normal. Ophthalmic re-examination and angiography 3 weeks after initial presentation revealed complete improvement; 3 weeks later there was a complete resolution of retinal dots, and 6 months later findings were normal with no chorioterinal scars.

COMMENT

Based on patient history, clinical signs, and serology we concluded that the illness presented was associated with rickettsiae. Within the genus Rickettsia, small obligate intracellular bacteria, three groups of antigenically related organisms are known: spotted fever, typhus, and scrub typhus. The former is less apparent during feeding. In general, rickettsiae invade endothelial cells, resulting in destruction of smooth muscle fibres, thrombosis, and extravasation of blood into the tissues. The papilloedema also observed in our patient, is supposedly based on microvessel destruction. Further, rickettsiae may cause round cell infiltration in the perivascular region (Fig 1). Slight optic atrophy, vacuolation of the retina, and an enlarged macula were observed with sparing of the macular regions. The complete resolution without any residual scars and normal ICG angiograms practically excluded choioretinitis. Further, this rickettsia did not meet the fluorescein angiographic criteria of white dot syndrome, since no hyperfluorescence was observed. In conclusion, rickettsiae may lead to white dots in the retina and, thus, should be considered in the differential diagnosis.

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Accepted for publication 15 April 1998
mained uninvolved until his death (due to respiratory failure) at 18 months' follow up.

Aqueous humour analysis revealed low Goldmann–Witmer coefficients (<3.0) and no DNA of varicella zoster virus (VZV), herpes simplex virus, and cytomegalovirus (polymerase chain reaction negative) was detected. High serum IgG levels (1:8192) against VZV might have contributed to the low Goldmann–Witmer coefficients.

COMMENT

Our patient was immunocompromised by the cutaneous non-Hodgkin's lymphoma and the accompanying therapy, but in contrast with AIDS patients CD4 counts were, albeit low, within the normal limits. Characteristically for the Sézary syndrome however, the CD4/CD8 ratio was elevated due to low counts of CD8 cells. Compared with HIV seropositive patients or AIDS patients with PORN, our patient responded well to aciclovir treatment. Furthermore, notwithstanding the optic atrophy, he retained useful visual acuity and retinal detachment did not develop. As suggested earlier, the degree and probably the type of immune deficiency might be responsible for the differences in clinical picture, and therapy outcome.

Five cases with PORN in HIV seronegative, but otherwise immunocompromised, individuals were reported. In contrast with our patient, none of these cases displayed all typical features of PORN as described originally in AIDS patients: two patients had moderate or pronounced intraocular inflammation, and the third patient showed ophthalmological lesions resembling bilateral acute retinal necrosis syndrome. Two other cases were only briefly mentioned but not described. Recently, PORN was described in a HIV seronegative patient following bone marrow transplantation with poor visual outcome despite preventive antiviral treatment; however, this patient had low CD4 counts.

Our case demonstrates that PORN may develop in HIV seronegative, immunocompromised patients with normal CD4 but low CD8 counts.

Sponsors/grants: none.

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Accepted for publication 21 April 1998