LETTERS TO THE EDITOR

Central serous chorioretinopathy may be a manifestation of the primary antiphospholipid syndrome

EDITOR,—Central serous chorioretinopathy is thought to be an idiopathic disease and such cases are not usually investigated for an underlying cause. Bullous exudative retinal detachment is a recognised complication of central serous chorioretinopathy (CSR). There are a number of reports of exudative retinal detachments occurring in the context of generalised coagulopathies. We present a patient with central serous chorioretinopathy who on further investigation was found to have the primary antiphospholipid syndrome.

CASE REPORT
A 42 year old white man presented in December 1992 with difficulty in reading and a grey “circle” in his right central field of vision of 6 weeks’ duration. His past medical history consisted of widespread burns requiring skin grafting and complicated by septicemia 20 years previously. Apart from two short courses of antimalarial tablets within the past 6 years, he had been taking no other medication. He was a non-smoker, but had been a heavy drinker. Corrected visual acuity was 6/9 N12 right eye, 6/9 N5 left eye. Funduscop y revealed bilateral retinal pigment epithelial (RPE) disturbances with no dye leakage on fluorescein angiography. Over subsequent months, transient serous elevation of the neurosensory retina at the right macula was noted. In October bilateral pigment epithelial detachments were noted with visual acuities of right eye 6/24 N10, left eye 6/60 N48. A trial of oral prednisolone seemed to halt the deterioration in vision and subsequently the patient was given 1 g daily of intravenous methylprednisolone for 3 days. A subsequent autoimmune screen revealed a positive lupus anticoagulant test (Russell viper venom with Cephalin 1.61 (0.99–1.05), Russell viper venom with platelets 1.12 (0.95–1.05)) and the presence of anticardiolipid antibodies (anticardiolipin IgG 46.3 gpl units (0–13.3)). Antinuclear antibodies were weakly positive at 1:20 with a homogenous/spiked pattern but anti-double stranded DNA, complement, extractable nuclear antigens and rheumatoid factor were negative. Steroids were stopped and the patient was given aspirin.

On subsequent review the patient had developed a large bullous inferior exudative retinal detachment in the right eye (Fig 1). B-scan ultrasound revealed retinal detachment with presumed subretinal fibrosis (Fig 2). Eventually the retinal detachment resolved spontaneously with the patient taking aspirin alone. Visual acuity is currently right eye 1/60, left eye 6/36. The patient manages N8 with a reading loupe.

COMMENT
Idiopathic CSR tends to be a self limiting disease which can be complicated by the development of exudative retinal detachments. The exact aetiology of CSR remains obscure and green angiography findings suggest that perfusion changes in the choriocapillaris may be responsible. Occulsive vascular eye disease involving the retinal and choroidal vessels has been reported in up to 8% of patients with raised anticardiolipin antibodies. Serous detachment of the macula has been reported with the primary antiphospholipid antibody syndrome; however, other ocular and systemic features were always noted.

The presence of antiphospholipid antibodies (anticardiolipin and lupus anticoagulant) can result in abnormalities of coagulation leading to venous and arterial thrombosis although exactly why this happens is not fully understood. Putative mechanisms include inhibition of antithrombin and activated protein C dependent anticoagulant systems, inhibition of fibrinolysis, and interference with the anticoagulant properties of β2 glycoprotein I.

We postulate that some cases of presumed idiopathic CSR may reflect thrombosis of the choroidal circulation secondary to the primary antiphospholipid antibody syndrome. Further studies are warranted to prove or disprove this hypothesis.

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5 Piccolino FC, Borgia L. Central serous chorioretinopathy and idiopathic green angiography. Retina 1994;14:231–42.

Acute retinal necrosis following chickenpox in a healthy 4 year old patient

EDITOR,—Originally described by Urayama et al, acute retinal necrosis (ARN) is a part of a continuous spectrum of necrotising herpetic retinopathies where the clinical expression is determined by the immune status of the host. We report a case of unilateral ARN complicated by retinal detachment following chickenpox. This is unusual in its severity and an extremely rare occurrence in this age group. To our knowledge, this patient represents the youngest case of chickenpox associated ARN.

CASE REPORT
A previously healthy 4 year old boy presented with total retinal detachment secondary to ARN of his left eye 6 weeks after an uncomplicated chickenpox infection. He was attending his local ophthalmologist soon after chickenpox, having been initially referred by an optometrist, where he was treated for anisometropic amblyopia. His visual acuity at that time was 6/18 right eye and 5/60 left eye, and no detachment was noted. However, 4 weeks later, visual acuity in his left eye had dramatically reduced to perception of light secondary to a retinal detachment, and he was transferred to the vitreoretinal service.

Examination revealed a dense relative afferent pupillary defect (RAPD) and panuveitis in the left eye. There was a 250° giant retinal tear, peripheral retinal necrosis, and a pale optic disc (Figs 1 and 2). The retina was completely detached. The right eye was normal.

The patient underwent left vitrectomy with silicone oil tamponade and 360° endophotoagulation. Vitreous polymerase chain reaction (PCR) was negative for herpes simplex virus (HSV) and varicella zoster virus (VZV) but positive for cytomegalovirus (CMV). A repeat CMV PCR on the same vitreous sample was negative. Magnetic resonance imaging did not show any evidence of intracranial calcification suggestive of CMV or VZV. He had IgM to VZV only. A full immunological survey was normal. This included a normal CD4 count of 1.4 × 109/l (range 1.2–2.0 × 109/l).

He was treated with systemic aciclovir and topical steroid and mydriatic following surgery. Although the operation was anatomically successful and the retinitis had resolved, acuity remained at perception of light due to optic atrophy and was unchanged at 7 months. His right eye remained normal throughout. Prophylactic treatment to the fellow eye with systemic aciclovir 400 mg by mouth twice daily is planned for 12 months.

COMMENT
Uncomplicated primary VZV infection (chickenpox) presents initially as maculopapular and later vesicular dermal eruptions without any ocular or CNS manifestations. ARN is a rare consequence of VZV infection in the immuno-
competing. Previous case reports by Matsuo et al and Culbertson suggest that chickenpox associated ARN has good visual prognosis irrespective of the immune status. None of their patients had retinal detachments. However, this case had an unusually severe retinitis culminating in a total retinal detachment and a poor visual prognosis.

Determining the specific cause of ARN is important as it has implications as to the choice of antiviral therapy. CMV associated ARN has been reported in healthy individuals. This case had no risk factors, clinical feature, or serological markers of CMV infection. The first vitreous PCR for CMV DNA was thus falsely positive, as confirmed by a repeat PCR test. Since HSV studies were also negative and he had classic features of chickenpox, backed by serology; the ARN was attributed to primary VZV infection. A retinal biopsy was not performed in this case because of a clear history of chickenpox infection, although it would have been strongly considered had the patient exhibited bilateral progressive disease.

The mode of retinal infection is unclear. Possible routes include direct haematogenous spread to the retina, or indirectly to the brain with subsequent transneural spread to the eye shortly after the primary episode. The length of therapy is controversial. Treatment in the acute phase and prophylaxis for the fellow eye is by systemic aciclovir. However, since aciclovir is only effective against actively replicating virus, and cannot eradicate latent virus, any protective effect is debatable. Indeed, delayed involvement of the fellow eye may be as long as 34 years after the initial episode.

Zoster related ARN remains largely a disease of adults despite the fact that a majority of patients acquiring chickenpox are in the paediatric age group. Before this case, only two patients aged 6 years and 13 years were reported. Since chickenpox associated ARN is extremely rare in children, it would not be justified to treat them prophylactically. However, the physician should be aware of the possibility of ARN soon after chickenpox infection. Early detection of ARN and prompt treatment are required to reduce the risk of severe visual loss.

**CASE REPORT**

A 37 year old woman wearing, glass, frameless spectacles (Rodenstock R244C), collided with a lamp-post at around 15 mph (24 km/h). She was driving a small hatchback car and the impact was sufficient to damage the bonnet and windshield. The car was not fitted with air bags but the driver and passenger were wearing three point seat belts. The driver’s head fell forwards hitting the steering wheel and fracturing her face. A shard from the spectacles lacerated the patient’s left eye.

When the patient was seen in casualty a 14 mm, trans-corneal laceration was noted, which extended into sclera (Fig 1). The cut was clean and linear with no loss of corneal tissue. A total hyphaema was present with a brisk anterior uveitis with periarterior and retinal detachment.

Figure 1 Fundus drawing of left eye. Note the giant retinal tear within the necrotic retina.

Figure 2 Perioperative photograph of left eye showing total retinal detachment.

Globe perforation with frameless spectacles

**EDITOR,—Glasses are perceived to be a protective eye shield; however, when lenses shatter severe ocular injuries may result. Several reports discuss the relation of spectacle design to their safety, both the lens and the frame contributing spectacle stability. A trend for thin or absent frames may place some patients at increased risk of serious ocular injury. We present the case of a car driver who sustained a perforated globe when glass frameless spectacles fractured during a low impact collision.**

**CASE REPORT**

A 37 year old woman wearing, glass, frameless spectacles (Rodenstock R244C), collided with a lamp-post at around 15 mph (24 km/h). She was driving a small hatchback car and the impact was sufficient to damage the bonnet and windshield. The car was not fitted with air bags but the driver and passenger were wearing three point seat belts. The driver’s head fell forwards hitting the steering wheel and fracturing her face. A shard from the spectacles lacerated the patient’s left eye.

When the patient was seen in casualty a 14 mm, trans-cornea laceration was noted, which extended into sclera (Fig 1). The cut was clean and linear with no loss of corneal tissue. A total hyphaema was present with a brisk haemorrhage from the wound. Further examination at surgery revealed a clean laceration with loss of the crystalline lens and two thirds of the inferior iris; no intraocular foreign body was found. Primary repair was undertaken, prolapsed iris and vitreous were removed. The wound was closed with 10/0 Vicryl to the sclera. The patient’s postoperative recovery was covered with systemic antibiotics, topical steroids, and cycloplegics.

Following surgery the patient regained hand movement vision. Ultrasonography initially showed choroidal detachment and vitreous haemorrhage. The choroidal effusions settled over 10 days. Postoperative pressure spikes were controlled with Iopidine (apraclonidine 0.1%).

**REFERENCES**

plastic frames or a plastic liner to metal frames. There are several case reports of penetrating ocular injuries either from the spectacle frame or from shattered glass in patients travelling a modest speed. 12 13 Our case demonstrates the effect of a low speed collision on fragile spectacle with glass lenses, dramatically increasing the likelihood of ocular injury from airbag activation or relatively minor collisions. Although polycarbonates are often used in the manufacture of many frameless spectacles their mechanical strength has not been reported in the literature. This case raises doubts about the safety of these spectacles, especially when made with glass lenses.

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CASE REPORT
A newborn boy was noted at birth to have multiple serous purple blisters on the left scalp region, measuring 0.5 cm to 2.0 cm in base (Fig 1). Anterior fontanelle asymmetry was also recognised. The skin lesions were thought to be aplasia cutis congenita and a skin biopsy was performed. Histopathological examination of the biopsied skin revealed loose connective tissue in the dermis with an apparent absence of mature collagen, multiple blood vessels with thrombi, intimal hyperplasia and chronic inflammation, and few hair follicles, most consistent with pigmented myxovascular hamartoma.

At the 2 week examination, a cornal lesion in the left eye was noted and the child was referred to the ophthalmology service at Wills Eye Hospital for evaluation. On examination, the vision was fix light in both eyes. The anterior segment examination of the left eye revealed an elevated, pink vascular conjunctival mass in the superotemporal quadrant measuring 12.0 mm × 4.0 mm in base and extending onto the cornea. The epibulbar mass was not biopsied as it appeared consistent with a choristoma, most probably dermoid, and it demonstrated no change in appearance and no effect on visual acuity (Fig 2). The right eye and the fundus of both eyes were normal.

The patient was the product of a normal vaginal delivery and an uncomplicated pregnancy. His birth weight was 2863 g, length 49.7 cm, and occipitofrontal circumference (OFC) 43.2 cm. There was no maternal history of tobacco, alcohol, or drug use. The

Table 1 Clinical findings documented in six reported patients with the ocular ectodermal syndrome

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<td>Patient 1</td>
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<td>Aplasia cutis congenita</td>
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<td>Skin hyperpigmentation</td>
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<td>Skin myxovascular hamartoma</td>
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*Occipitofrontal circumference.
We believe that the association of atypical epibulbar dermoid and myxovascular hamartoma of the skin represents a different and extremely unusual variant of ocular ectodermal syndrome.

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COMMENT
Aplasia cutis congenita, which is the hallmark of ocular ectodermal syndrome, is the congenital absence of skin presenting heterogeneously as an ulcerated, eroded, scarred, or blistered area with alopecia, usually on the scalp near the vertex. Our patient was initially felt to have aplasia cutis congenita. However, pathological examination revealed pigmented myxovascular hamartoma of the scalp, characterised by the sporadic onset of asymmetric scalp lesions presenting with subcutaneous nodules. Our search of the literature failed to disclose any previous reports on the association of cutaneous myxovascular hamartoma and epibulbar dermoid, as described in our patient. We speculate that our case represents an extremely unusual variant of ocular ectodermal syndrome.

In addition to the epibulbar dermoid, other ocular findings in ocular ectodermal syndrome supported by the literature include corneal opacity and strabismus (Table 1). Besides the skin and eye findings, associated systemic findings in ocular ectodermal syndrome are numerous and can occur in several systems including the musculoskeletal system (Table 1). Cardiac anomalies in ocular ectodermal syndrome include atrial septal defect and neurological findings include psychomotor retardation, and seizures. Although the skin and eye findings are both relatively innocuous, knowledge of the more serious associated systemic disorders, such as seizures, psychomotor retardation and various congenital defects, should be recognised. In addition to the skin and ocular findings, our patient had parietal bone defect and anterior fontanelle asymmetry.

Foveal translocation with secondary confluent laser for subfoveal CNV in AMD: 12 month follow up

EDITOR—Foveal relocation surgery for the treatment of subfoveal choroidal neovascularisation (CNV) secondary to age related macular degeneration (AMD) has received significant publicity in the lay press and has given rise to some controversy reported in the BJ Ophthalmology. We describe the 12 month follow up of a case reported in the press in December 1998.

Table 1

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<td>Figure 1 Small purple blistered lesions on the scalp consistent with myxomatous hamartomas.</td>
<td>Figure 2 Oblong vascular connerosal dermoid in the superotemporal quadrant.</td>
<td>Figure 1 Fluorescein angiogram images before and after foveal relocation surgery (A) preoperatively, showing pure classic subfoveal CNV less than 1 disc area in extent; (B) 7 days postoperatively showing fovea located below CNV; (C) 5 months postoperatively showing recurrence; (D) 4 weeks after confluent laser to recurrence shown in (C) showing complete closure of CNV.</td>
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substantial improvement of vision which was at its best 6 months postoperatively but still maintained at a good level after 12 months. The importance of close angiographic monitoring is illustrated by the development of recurrent CNV, which is not unexpected in laser treated CNV.

Mr JB was one of the first patients to undergo foveal relocation as part of a pilot study performed with the approval of the Liverpool research ethics committee. The study performed with the approval of the Liverpool research ethics committee. The importance of close angiographic monitoring is illustrated by the development of recurrent CNV, which is not unexpected in laser treated CNV.

To further investigate this unusual dermatological reaction, a brimonidine eyedrop was placed on the patient's cheek in a circular pattern. The contact area blanched almost immediately. The blanching intensified over a period of 30 minutes, and erythema developed in the surrounding skin. The blanching and erythema lasted approximately 20 hours before it began to fade. Complete resolution occurred by 22 hours. Another drop of brimonidine produced a similar regional blanching with surrounding erythema when placed on the patient's back.

**COMMENT**

Although brimonidine is almost 1000-fold more selective for the $\alpha_2$ than the $\alpha_1$ receptor subtype, its ability to cause vasoconstriction and reactive hyperaemia demonstrates that it has some $\alpha_2$ mediated effects. Brimonidine is 23–32-fold more selective than apraclonidine, and conjunctival blanching has been reported as an infrequent local effect of brimonidine, whereas it is seen in up to 85% of patients using apraclonidine. Our patient demonstrated a pronounced $\alpha_2$ mediated vascular reaction of the periorbital skin. Brimonidine caused skin blanching and a surrounding hyperaemia that was not limited to the trail of the tear drop, but included the entire cheek. While the half life of intrasceral pressure lowering effect of brimonidine is only 8–12 hours, the dermal vasoconstrictive effect of the drug persists for almost 24 hours.

Supported in part by a grant from Research to Prevent Blindness.

**References**


**Figure 1 Linear skin blanching (arrows) after administration of brimonidine eyedrops.**

**Figure 2 Colour fundus photographs of follow up after foveal relocation surgery: (A) 7 days postoperatively showing fold extending from disc into superonasal retina; (B) 1 week after (A) showing confluent laser applied to CNV and fovea located inferiorty to laser; (C) 5 months postoperative CNV recurrence with blood (VA = 6/9); (D) 12 months postoperatively (VA = 6/18).**

The oil droplet sign

EDITOR,—Over the past 22 months we have collected a series of five patients with basal cell carcinomas of the lid who exhibit the oil droplet sign. The following case report is a representative example.

CASE REPORT
A 27 year old Zimbabwean male presented with a 14 month history of a progressively enlarging mass in the right lower lid.

Clinically there appeared to be a nodular basal cell carcinoma involving the lateral part of the right lower lid measuring 3.5 × 3.5 mm and involving the lid margin (Fig 1). There was no regional node involvement. On elevating the lid there were multiple transparent droplets beneath the tarsal conjunctiva measuring between 0.25 and 0.50 mm in diameter (Fig 2, see arrow). The droplets lay directly over the area of tumour involvement and extended beneath the fornical conjunctiva. The droplets contained a clear or straw coloured fluid which had an oily appearance.

A full thickness wedge resection with 4 mm margins was performed. The lid was closed directly.

A light photomicrograph (Fig 3) of the full thickness lid resection shows a basal cell carcinoma at the lid margin (superiorly) and along the tarsal conjunctiva (left). Lipid droplets are associated with an intense chronic inflammatory cell infiltrate. Note one large droplet very close to the surface.

COMMENT

We have noted this sign over many years and have collected five cases over the past 22 months. To our knowledge this sign does not appear in the literature. The sign has only been noted among cases of basal cell carcinoma and has not been associated with other tumours such as squamous cell carcinomas or meibomian gland carcinomas.

We propose that the lipid droplets represent trapped meibomian gland secretions which cannot drain through obstructed or infiltrated meibomian gland ducts. It is interesting to note that the droplets appear beneath the conjunctiva rather than forming collections within the tarsus as is seen with chalazion formation. The invasion and destruction of the tarsus by tumour (as opposed to benign obstruction) may offer a route of least resistance whereby the droplets appear beneath the conjunctiva.

There are insufficient numbers of cases to state whether this is exclusively a sign associated with basal cell carcinomas or whether it may appear among the rarer tumour types. The sign is however a good indicator of an infiltrative process and may assist the clinician in the diagnostic process.

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Acute retinal necrosis following chickenpox in a healthy 4 year old patient

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