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 coagulothrombopathies. 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 antimarial 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. Funduscopy revealed bilateral retinal pigment epithelial detachments 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 (0.93–1.05), Russell viper venom with platelets 1.12 (0.95–1.05)) and the presence of anticardiolipin antibodies (anticardiolipin IgG 46.3 gpl units (0–13.3)). Antinuclear antibodies were weakly positive at 1:20 with a homogenous/speckled pattern but anti-double stranded DNA, 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 aetiopathy of CSR remains obscure and angiography findings suggest that perfusion changes in the chorioapillaris may be responsible. Occlusive ocular vascular disease involving the retinal and chorioapillaris 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, both 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.

M T J COSTEN
Southampton Eye Unit, Tremena Road, Southampton
SO16 6DY
J A OLSON
Eye Department, Aberdeen Royal Infirmary, Aberdeen
AB25 2ZN
Correspondence to: Mr Costen
MTRJAC@aol.com
Accepted for publication 13 January 2000

5 Piccolino FC, Borgia L. Central serous chorioretinopathy and indocyanine 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 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, 6 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 papillary 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° endophoto-coagulation. 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 this infection in the immuno-
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.1,2 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 windscreen. 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 spectacles (Fig 1). The force was not sufficient to bruise her face. A shard from the spectator 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 2). 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 nylon to the cornea and 6/0 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...
Ocular ectodermal syndrome of epibulbar dermoid and cutaneous myxovascular hamartoma

EDITOR,—Ocular ectodermal syndrome is a congenital disorder manifesting classically with epibulbar dermoids and aplasia cutis congenita. Systemic abnormalities may also accompany ocular ectodermal syndrome.1 4 We describe the case of a newborn boy who presented with unilateral epibulbar dermoid and myxovascular hamartomas of the scalp. We believe that the association of epibulbar dermoid and myxovascular hamartomas of the skin represents an unusual variant of ocular ectodermal syndrome.

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 corneal lesion in the left eye was noted and the child was referred to the oculo-neurology 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 fundi 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>
<thead>
<tr>
<th>Table 1 Clinical findings documented in six reported patients with the ocular ectodermal syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
</tr>
<tr>
<td>Birth weight (kg)</td>
</tr>
<tr>
<td>Birth OFC* (cm)</td>
</tr>
<tr>
<td>Postnatal macrocephaly</td>
</tr>
<tr>
<td>Craniofacial anomalies</td>
</tr>
<tr>
<td>Parietal bossing</td>
</tr>
<tr>
<td>Frontal bossing</td>
</tr>
<tr>
<td>Skul defect</td>
</tr>
<tr>
<td>Location</td>
</tr>
<tr>
<td>Prominent eyes</td>
</tr>
<tr>
<td>Epicanthal folds</td>
</tr>
<tr>
<td>Flat nasal bridge</td>
</tr>
<tr>
<td>Ocular anomalies</td>
</tr>
<tr>
<td>Epibulbar dermoid</td>
</tr>
<tr>
<td>Other findings</td>
</tr>
<tr>
<td>Ectodermal anomalies</td>
</tr>
<tr>
<td>Skin hypopigmentation</td>
</tr>
<tr>
<td>Skin myxovascular hamartoma</td>
</tr>
</tbody>
</table>

*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.

Supported by the Paul Kayser International Award of Merit in Retina Research, Houston, TX (J Shields) and Eye Tumor Research Foundation, Philadelphia, PA.

KAAN GÜNDÜZ
CAROL L SHIELDS
YELENA DOYCH
Oncology Service, Wills Eye Hospital, Philadelphia, PA, USA

BRUCE SCHNALL
Pediatric Ophthalmology Service

JERRY A SHIELDS
Oncology Service

Correspondence to: Carol L. Shields, MD, Oncology Service, Wills Eye Hospital, 900 Walnut Street, Philadelphia, PA 19107, USA

Accepted for publication 28 January 2000

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 include asymmetry of the cornea 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.

Figure 1 shows the clinical appearance on 1 October 1999 is no further evidence of CNV recurrence; the VA improved to 6/9. After 9 and 12 months VA was 6/12. Figure 2 shows the flattening retinal fold (Fig 2B). On 5 November 1998 refraction VA was 6/15, a small zone of persistent CNV was treated. On 8 January 1999 refraction VA was 6/12. Figure 2C shows a flattening retinal fold and Figure 1C a zone of recurrent CNV which was treated. On 8 October 1999 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 BJO. We describe the 12 month follow up of a case reported in the press in December 1998.

CASE REPORT

Mr JB, a 70 year old retired dentist was referred from another unit on 31 August 1998 with AMD and a history of poor vision for 5 years in the right eye and 5 months in the left. Refraction visual acuity was right eye 5/60 (+1.50/+1.50), left eye 6/60 (+2.25 sphere), Snellen equivalent. Figure 1A shows a subfoveal pure classic CNV of less than 1 disc area in extent in the left eye. The right eye was affected by end stage fibrotic scar. After full discussion of risk/benefit of confluent laser and the experimental nature of surgery he underwent surgery on 24 September 1998 (DW). Scleral plication was achieved by using 14 radially disposed sutures applied to the superotemporal quadrant of the globe. A three port pars plana vitrectomy was carried out and a subtotal retinal detachment was induced by subretinal infusions via three posterior retinotomies. The retina was reattached with a fluid/air exchange and the fovea was manipulated to its final position with a small bore flute needle. A radial fold formed in the upper nasal aspect of the fundus.

Figure 2 shows the retina on the 7th postoperative day with the superonasal fold and Figure 1B the midphase fluorescein angiogram (FA) showing the CNV now located superior to the fovea. On 8 October 1998 confluent laser was applied to the CNV (SPH) (Fig 2B). On 5 November 1998 refraction VA was 6/15, a small zone of persistent CNV was treated. On 8 January 1999 refraction VA was 6/12. Figure 2C shows a flattening retinal fold and Figure 1C a zone of recurrent CNV which received repeat confluent laser. On 12 February 1999 the post-laser follow up FA (Fig 1D) shows no persistent leakage; the VA had improved to 6/9. After 9 and 12 months VA was 6/18, N6 (~0.75/+0.50; +3.00 add) with no further evidence of CNV recurrence; the clinical appearance on 1 October 1999 is shown in Figure 2D.

COMMENT

At present, foveal relocation surgery is the only therapeutic option which aims to improve the vision for patients with CNV caused by AMD. The case we report gained a
The importance of close angiographic monitoring is illustrated by the development of recurrent CNV, which was maintained at a good level after 12 months.

Although brimonidine is almost 1000-fold more selective for the α₂ than the α₁ receptor subtype, its ability to cause vasoconstriction and reactive hyperaemia demonstrates that it has some α₁ 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 α₁ mediated vascular reaction of the periorbital skin. Brimonidine caused skin blanching and a surrounding hyperaemia that was not limited to the trail of the teardrop, but included the entire check. While the half life of intrascleral pressure lowering effect of brimonidine is only 8–12 hours, the dermal vasoconstrictive effect of the drug persists for almost 24 hours.
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 evverting 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 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.

P HEYWORTH
J R O COLLIN
Moorfields Eye Hospital, London

P LUTHERT
Institute of Ophthalmology, London

Correspondence to: Mr P Heyworth, Moorfields Eye Hospital, City Road, London EC1V 2PD

susan.heyworth@virgin.net

Accepted for publication 27 January 2000

Figure 3 Photomicrograph of full thickness lid resection shows a basal cell carcinoma at the lid margin (superiorly) and along the conjunctiva (left). Lipid droplets are associated with an intense chronic inflammatory cell infiltrate (haematoxylin and eosin; original magnification ×27). The inset (from an adjacent section) shows the lipid droplets at higher magnification (×110).