LETTERS TO THE EDITOR

Relapses of CMV retinitis after 2 years of highly active antiretroviral therapy

EDITOR,—Highly active antiretroviral therapy (HAART) can reduce HIV replication and increase the CD4+ cell count.¹ Some cytomegalovirus (CMV) disease relapses have been described within the first 3 months of HAART.² However, after this critical phase, quiescent retinitis and an unusually prolonged relapse free interval suggest restoration of immune functions. Maintenance therapy can be discontinued for the majority of HAART responders.³⁻⁶ The duration of CMV disease remission is actually unknown.

CASE REPORTS

A 29 year old, HIV infected woman was affected by CMV retinitis in the left eye in August 1996. Her CD4+ T lymphocyte count was 15 cells $\times 10^9/l$ at that time. CMV retinitis was treated with intravenous foscarnet. HAART was initiated in June 1996 (d4T, 3TC, and zidovudine). In June 1997, her CD4+ cell count increased to 400 cells $\times 10^9/l$ and the HIV-1 viral load became undetectable. CMV maintenance therapy was discontinued in December 1997 without any relapse until May 1999 when a relapse of CMV retinitis, associated with inferior retinal detachment, was noted. At that time, her CD4+ cell count was 380 cells $\times 10^9/l$, CMV viraemia was negative, but HIV viral load had increased to 31 000 copies/ml. The lymphoproliferative test against CMV antigens was negative. The patient was treated with intravenous ganciclovir and vitreoretinal surgery was performed. CMV DNA was found in the vitreous by polymerase chain reaction.

A 69 year old HIV infected man was referred to our department in November 1996 for bilateral CMV retinitis. CD4+ T lymphocyte count was 35 cells $\times 10^9/l$ at onset. Retinitis was treated with intravenous foscarnet. HAART (3TC, d4T, nevirapine) was initiated in September 1997. For 2 years, the HIV-1 viral load remained undetectable, and CD4+ cell count rose to 120 cells $\times 10^9/l$. In October 1998, the HIV viral load started to increase, reaching 6100 copies/ml. In May 1999, a relapse of CMV retinitis was diagnosed and treated successfully with intravitreal ganciclovir injections. At that time, the HIV viral load rose to 22 000 copies/ml, and CD4+ cell count was 180 cells $\times 10^9/l$. The lymphoproliferative test against CMV antigens was negative. CMV viraemia and pp65 antigenaemia always remained negative.

COMMENT

Several studies suggest that selected patients with healed CMV retinitis who have both immunological and virological response to HAART can temporarily discontinue maintenance therapy.³⁻⁶ In our patients on HAART, CMV retinitis was quiet for months, as long as the HIV-1 replication remained under control. CMV retinitis relapse occurred when the viral load started to increase even though the CD4+ T lymphocyte count remained stable (mean 280 cells $\times 10^9/l$). Autran *et al* have studied in vitro lymphocyte proliferation to

specific antigens such as CMV and reported positive effects of HAART on CD4+ T cell functions.¹ The CD4+ T cell reactivity to recall antigens that restore a certain immune competence is linked to the control of HIV replication.⁷ For our patients, the T cell reactivity to CMV antigens was negative. These immunological tests are not routinely performed and their usefulness in predicting CMV diseases is not established. Casado *et al* showed that a positive CMV polymerase chain reaction test is the most predictive test associated with the development of CMV diseases.⁸

In a prospective multicentre study, Mazon *et al* showed that HIV viral load is a predictive marker, independent of the CD4+ cell count.⁹ Our findings suggest that an increase in the HIV viral load increases the risk of CMV retinitis relapse despite a high CD4+ T lymphocyte count.

Our case reports emphasise the importance of close ophthalmological follow up in patients on HAART when the HIV-1 viral load starts to increase despite a CD4 T lymphocyte count of over 100 cells $\times 10^9/l$.

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A large rapidly growing pilomatixoma on a lower eyelid

EDITOR,—Pilomatixoma (also called benign calcifying epithelioma of Malherbe) is a rare, benign tumour originating from the matrix of the hair root, first described in 1880 by Malherbe and Chenantais.¹ Pilomatixoma can occur almost anywhere on the body but has a propensity to occur in the head and neck region, often involving the eyelid or eyebrow.² It is most commonly seen in children and

adolescents with a female predominance reported.^{2,3} Pilomatixoma is often misdiagnosed clinically and the correct diagnosis only established after excision and histological examination.³

This case report describes an unusual presentation of pilomatixoma of the lower eyelid following trauma. It also highlights some characteristic features of this tumour that can help the clinician differentiate it from other eyelid masses found in children.

CASE REPORT

A 4 year old girl presented to the eye casualty department with a lump on her left lower eyelid. She had been hit by a ball 3 months previously after which the lump had appeared. On examination, it measured 1 cm by 1 cm, had a red-blue discoloration, and was hard and painful to touch. Her ocular examination was otherwise normal.

An initial diagnosis of unresolving haematoma or possible cholesterol granuloma was made. She was listed for surgical exploration of the lump but before this could be carried out she re-presented because the lump had grown rapidly in size. It now measured 2 cm by 2.2 cm and still had the red-blue discoloration (Fig 1).

Excision biopsy was expedited. The skin-orbicularis muscle incision was made over the mass, inferior to the lateral aspect of the lower lid, and the tumour shelled out intact. The tumour was well circumscribed, nobbly hard, of yellowish white colour, and measuring 19 mm $\times$ 15 mm $\times$ 8 mm (Fig 2A and B).

Histopathology showed a pseudoencapsulated mass composed mainly of eosinophilic acellular material in which ghost cells were prominent, together with numerous small foreign body giant cell granulomata and considerable calcification. At the periphery there were focal areas of basaloid cells with little cytoplasm. Squamous maturation of these cells to form keratinous material was present (Fig 2C and D). The pathological diagnosis was pilomatixoma.

The child has made an uneventful recovery and remains free of tumour 2 years later.

COMMENT

Pilomatixoma typically presents as a firm, non-tender, subcutaneous nodule, adherent to the skin but not fixed to underlying tissue.⁴ In our case the lump was tender with bruising caused by preceding trauma. The association with trauma is unusual.

In their survey of 150 cases of pilomatixoma in a paediatric hospital Orlando and coworkers found no history of trauma or inflammation.⁵ However, a lesion that bruised easily on trauma has been described in a case report of an aggressive variant of pilomatixoma that grew over the parotid area in the face of a 4 year old boy.⁶

Figure 1 Large lower lid/cheek tumour visible on left side. Note blue discoloration and small area tethering to skin.

Figure 2 (A) Peroperative exposure of tumour. (B) Firm yellow nodular tumour. (C) This low power view shows islands of basaloid cells at the edge of the tumour which is formed mainly of keratinous debris with scattered areas of granulomatous inflammation and focal calcification responsible for fracturing of the section during cutting. (Haematoxylin and eosin, original magnification $\times 20$). (D) A high power view of the edge of the tumour showing basaloid squamous epithelium maturing to the right with an area of parakeratosis merging into ghost cells. An area of granulomatous inflammation with a giant cell of foreign body type is present in the upper part of the field. (Haematoxylin and eosin, original magnification $\times 200$)

Red-blue or blue discoloration of the skin is a more typical feature of pilomatrixoma⁸ and should help to differentiate it from inclusion and dermoid cysts which are the most common misdiagnoses.⁹ Inclusion cysts have a diffuse yellow colour when filled with keratin and are more likely to be softer and more fluctuant than pilomatrixoma. They are also rarer in children than in adults. The skin over dermoid cysts looks normal and can be moved freely over the lesions.

Capillary haemangiomas that arise in the subcutaneous tissue without involving the skin can have a blue-purple colour similar to pilomatrixoma but that lesion tends to be soft and compressible on palpation and growth is much faster than pilomatrixoma. A chalazion can become inflamed but does not show the red-blue discoloration of pilomatrixoma.

Rhabdomyosarcoma is the most common orbital malignancy of childhood that can present rarely as an eyelid mass with erythema of the overlying skin but no other symptoms.¹⁰ However, the tumour usually extends to the

deeper orbital tissues and is not restricted to the subcutaneous tissues.

CONCLUSION

Pilomatrixoma of the eyelid is often misdiagnosed clinically. But there are characteristic features of the lesion that can help clinicians differentiate it from other tumours seen in children. The tumour is usually rock-hard, freely mobile within the subcutaneous tissue, but lightly attached to the overlying skin which can have a blue or red-blue discoloration. The tumour may also present following trauma, as demonstrated by this case.

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Use of mitomycin C and r-tPA for the management of conjunctival membrane and cataracts in a child with conjunctivitis lignosa

EDITOR,—Ligneous conjunctivitis is a rare chronic pseudomembranous conjunctivitis that in some cases is associated with membrane formation in other mucous tissues. Involvement of the palpebral conjunctiva is the most common site of the disease although formation of bulbar membrane and even corneal membrane have been reported.¹ Induction of membrane formation may occur after minor trauma (for example, conjunctivitis, surgery, toxic agents).

The disorder has an AR inheritance and is caused by an impaired fibrinolysis related to deficient plasminogen type I. Recently causative mutations in the plasminogen gene have been identified in affected patients.²

Pathological findings disclose fibrin as the major component within amorphous hyalin-like eosinophilic material, fibroblastic proliferation, inflammatory cellular infiltration, and acid mucopolysaccharides.³

The treatment of the disorder has generally been unsuccessful, although spontaneous resolution may occur. Excision of the membrane (with cryocoagulation and/or autologous conjunctival graft or scleral graft) often worsens the conjunctivitis within a few days after surgery.

Treatment strategies that act on the different constituents found in the ligneous membrane have been proposed; topical proteolytic enzymes (hyaluronidase and chymotrypsin),⁴ fibrinolysin drops that cause fibrinolysis,⁵ and topical anti-inflammatory agents (corticosteroids and cyclosporin).⁶

In 1995 De Cock *et al*⁷ suggested the administration of topical heparin (antifibrin action) in combination with the above agents. The authors reported efficacy of their treatment although it was not consistent in all patients.

CASE REPORT

In an earlier report we discussed a boy who underwent eight consecutive unsuccessful excisions of a ligneous conjunctival membrane between August 1997 and February 1999.⁸ Surgery was combined with topical medications—chymotrypsin, hyaluronidase, dexamethasone, and heparin.

Research on type I plasminogen deficiency conducted by Schuster *et al* disclosed in our patient a decreased plasminogen activity (36% of normal activity) and a causative mutation in the plasminogen gene.⁹

On 12 March 1999 we performed another excision of the recurrent membrane and we applied mitomycin C for 3 minutes followed by an amniotic membrane graft. Adjuvant medical treatment consisted of the administration of systemic prednisone 1 mg/kg/day and topical heparin for 14 days. This treatment was successful (Fig 1) and since then the membrane has not recurred.

On ocular follow up examination we observed the formation of complicated SCP cataract.

On 27 January 2000 we performed a lensectomy with insertion of a PC-IOL and peroperatively we injected 25 µg tissue plasminogen activator (r-tPA) intracamerally, to prevent fibrinous effusion. During the first postsurgical week, slit lamp examinations showed the absence of reaction in the anterior chamber. The child was discharged from hospital on day 7. On control examination (day 9) slit lamp examination revealed multiple fibrin strands emerging from the pupil towards the cornea over 360° (Fig 2). The child was again admitted and received a protective shell to prevent eye robbing. This measure was followed by the disappearance of fibrin after 1 day. The PC-IOL restored the visual acuity.

Figure 1 (Top) Ligneous conjunctival membrane. (Bottom) The same child 1 month after surgery with mitomycin C.

Figure 2 Day 9 after cataract surgery. Notice multiple fibrin strands emerging from the pupil towards the cornea.

COMMENT

Up to now strategies that have been proposed for the treatment of conjunctivitis lignosa act on different aspects in the cascade of wound healing. We added two agents, mitomycin C and r-tPA, that had not yet been applied for this indication.

Mitomycin C is an antiproliferative drug that prevents the development of scar tissue and is widely used in ocular surgery. We propose to use mitomycin C in combination with heparin and corticosteroids in the treatment of ligneous conjunctivitis.

If intraocular surgery is needed in affected patients, we suggest using intracameral recombinant tissue plasminogen activator in order to stimulate fibrinolysis. The intracameral injection of 25 µg r-tPA has proved to be efficient and safe in the treatment of severe postoperative fibrinous reactions.¹⁰

Until causative treatment with pharmacological plasminogen is possible we believe that mitomycin C and r-tPA may be useful adjuvant agents respectively in conjunctival and intraocular surgery in patients with ligneous conjunctivitis.

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Annular granular corneal opacity: a rare corneal stromal dystrophy or degeneration?

EDITOR,—Very rare, subtle conditions of the cornea may be infrequently reported but can provide valuable insights into the workings and diseases of the cornea. We report an unusual appearance of corneal rings in an asymptomatic patient with no similar abnormalities detected in first degree relatives.

CASE REPORT

A 56 year old white female emmetrope (20/20 both eyes) presented with photopsia second-

Figure 1 Anterior segment photograph demonstrating corneal ring opacity in mid-peripheral stroma.

Figure 2 Anterior segment photograph demonstrating corneal ring opacity in mid-peripheral stroma.

ary to posterior vitreous separation. Bilateral, thin (<0.5 mm), grey-white, granular but continuous, mid-peripheral stromal corneal ring opacities were incidentally noted (Figs 1 and 2). The rings were 7–8 mm diameter and in cross section were “V”-shaped: widest nearest to Descemet’s membrane. The apex, anteriorly, was slightly displaced towards the centre of the cornea. The central and peripheral cornea was clear. Pachymetry and specular microscopy were unremarkable. The remainder of the anterior segment including drainage angle was completely normal. The patient had no previous history of eye disease or trauma.

Apart from bendrofluzide taken for hypertension, she took no other medicines. Systemic examination including hair, skin, and nails, was unremarkable. Full blood count, urea and electrolytes, liver function test, glucose, sedimentation rate, urate, copper, fasting lipids, and autoantibodies were normal. She was found to be heterozygous for factor V Leiden (present in 5% of the population).

Examination of all living first degree relatives (mother aged 89, brother aged 58, son aged 31, daughter aged 27) revealed no corneal abnormalities.

COMMENT

Within the literature we have found a total of six cases of bilateral corneal rings of a similar size and appearance to that which we describe; all without apparent aetiology.^{1–4} In four cases the rings are reported as predominantly located in superficial or anterior stroma.^{1–3,4}

However, in the two other cases, wedge-shaped ring opacities, most dense over Descemet's membrane are described, a feature common to those we observed.² The ages of the patients in these reports range from 25 to 80 and all investigations, sometimes exhaustive, were unremarkable. Where family members were examined, none demonstrated similar features and where patients were followed up (2–9 years), appearances remained stable. However, in one report, the rings were not observed during a ophthalmic examination 14 years previously, suggesting that they are not congenital in origin.¹

Another report of a unilateral, off-white, anterior stromal, mid-peripheral ring is of note because the patient was found to have elevated serum cholesterol.⁵ However, the opacity may have been a unilateral arcus.⁴

Of the six bilateral cases reported, only two describe the ring as wedge-shaped in cross section and predominantly in the posterior stroma² and closely resemble our case. With so few cases reported, over a wide range of ages, and without affected family members it is unclear whether the condition should be described as a dystrophy or a degeneration. It is important to report such rarities, so that further correspondence and reporting is encouraged which may lend insights into causes of more serious corneal abnormalities.

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Idiopathic tractional corectopia

EDITOR.—Congenital abnormalities of pupil position and shape are uncommon. Slit lamp examination will usually identify which of the various rare anterior segment developmental anomalies has caused the pupil appearance. Possible associations include iris coloboma, Axenfeld-Reiger anomaly, ectopia lentis et pupillae, persistent pupillary membrane, and hyperplastic pupillary membrane. Surgical intervention for the pupil abnormality is not normally required.

Idiopathic tractional corectopia is an isolated unilateral congenital pupil abnormality with a highly characteristic appearance (Fig 1). The four previously reported cases required no intervention or responded well to simple lysis of the fibrous strand that is characteristic of the condition.¹

We report a fifth case, where progression of the tractional process led to failure of initially successful conservative management and a surgical pupilloplasty was required as no benefit was obtained by cutting the fibrous strand.

CASE REPORT

A healthy male child was referred to the ophthalmology department soon after birth with a unilateral misshapen eccentric pupil. There

Figure 1 Preoperative appearance of the right eye. The white fibrous band from the pupil to limbus and the posterior embryotoxon-like corneal opacity are pathognomic of idiopathic tractional corectopia.

was no family ocular history. At initial examination he was found to have right corectopia with an oblique oval pupil and a fibrous band extending from the pupil margin to the endothelial surface of the peripheral superonasal cornea. The iris appeared otherwise normal: hypoplasia, polycoria, retroillumination defects, or ectropion uvea were not present. The corneal diameter and intraocular pressure were normal. A pupil opening was present with normal red reflex and limited view of the ocular fundus. The pupil did not dilate with topical mydriatic. Examination of the left eye was unremarkable. Ultrasound of the eyes was normal.

Over the next 5 months the pupil in the right eye became smaller and more eccentric. Occlusion therapy was commenced but the right eye became visually unresponsive. The pupil could be seen to offer no optical pathway by this stage (Fig 1). At surgery the fibrous band was cut with intraocular scissors but there was no effect on pupil morphology. Further attempts to mobilise the pupil threatened to disinsert the iris root superonasally. Therefore a central pupil was created by small multiple iris sphincter incisions. Postoperatively the eye settled well and without complication. At 6 months postoperatively the vision in the right eye had improved to 6/24 with aggressive occlusion therapy.

COMMENT

Early reports of isolated corectopia are not particularly useful owing to inadequate examination techniques and equipment.² Isolated corectopia was considered by Duke-Elder to be a bilateral condition.³

Scott Atkinson *et al*¹ reported four children with isolated unilateral corectopia, resulting from a white band that extended from the pupil margin to insert in a circumferential condensation of tissue on the endothelial surface of the peripheral cornea that superficially resembled an incomplete posterior embryotoxon. The condition was named "idiopathic tractional corectopia" (ITC). The pupil displacement may progress during the first months of life, possibly by further contraction of the fibrous band. The origin of the band is unknown. Children with ITC need to be monitored closely by monocular vision testing as they are at risk of amblyopia. Prompt laser or surgery may be indicated if vision reduces because of optical pupil occlusion.

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Subhyaloid or subinternal limiting membrane haemorrhage in meningococcal meningitis

EDITOR.—Subhyaloid or subinternal limiting membrane haemorrhage (sub-ILM) is usually caused by diabetic retinopathy,¹ hypertensive retinopathy,² retinal artery macroaneurysm,² Valsalva retinopathy,³ Terson's syndrome,² or blood dyscrasias.⁴ Bacterial meningitis has been associated with subhyaloid or sub-ILM haemorrhage.^{5,6} We report a case of subhyaloid or sub-ILM haemorrhage in a young boy with meningococcal meningitis, and discuss the possible mechanisms and the management of this condition.

CASE REPORT

A 13 year old boy was admitted to the paediatric ward with a 24 hour history of progressive drowsiness, headache, neck stiffness, nausea, and vomiting. On examination, he was confused, agitated, and pyrexial. There were petechial skin haemorrhages on both arms and legs. The blood pressure was normal and all peripheral pulses were present. There were no focal neurological signs. Funduscopy showed no obvious abnormalities. Lumbar puncture was contraindicated because of the risk of raised intracranial pressure. A clinical diagnosis of meningococcal meningitis was made. He was treated with intravenous benzylpenicillin and cefotaxime, and he was sedated and ventilated for 48 hours.

Gram stain and microscopic examination of smears from petechial skin lesions revealed intracellular Gram negative diplococci consistent with *Neisseria meningococcus* infection. Blood culture and polymerase chain reaction studies were negative. The prothrombin time was prolonged on the first two consecutive days to 19 seconds (normal 11.5–15 seconds). The activated partial thromboplastin time and the platelet count were normal. His condition improved rapidly. He was discharged home on day 6, and he returned to hospital for intravenous ceftriaxone therapy daily for another 4 days.

On day 9, he noticed poor vision in the right eye on inadvertently covering his left eye. Ophthalmological examination revealed visual acuity to be counting fingers right eye and 6/5 left eye. Anterior segments were normal. Funduscopy of the right eye revealed a round, well circumscribed, dome-shaped haemorrhage overlying the posterior pole consistent with a subhyaloid or sub-ILM haemorrhage (Fig 1). Conservative management was decided upon because of his young age, recent severe illness, and good vision in the left eye. By the fourth week, there was contraction in size of the haemorrhage. By the 16th week, the right pre-retinal haemorrhage had almost completely

Figure 1 Right fundus photograph showing a round, well circumscribed, dome-shaped haemorrhage overlying the posterior pole.

Figure 2 Fluorescein fundus angiography at 16 weeks showing the clearance of the haemorrhage with residual inferior rim of haemorrhage outlining the detached internal limiting membrane.

cleared and vision had improved to 6/5. Fundus fluorescein angiography revealed no retinal or choroidal abnormality (Fig 2).

COMMENT

As far as we know, there have been two previously reported cases of subhyaloid or sub-ILM haemorrhage associated with bacterial meningitis.^{5,6} The exact mechanism of this haemorrhagic response is not fully understood. Valsalva retinopathy has been suggested.⁵ Coughing and vomiting cause a rapid rise in intravenous pressure within the eyes. This can cause spontaneous rupture of superficial retinal capillaries.⁵

Elevation of intracranial pressure is a potential complication of meningococcal meningitis.⁷ A sudden increase in intracranial pressure can rupture the epipapillary and peripapillary capillaries, known as Terson's syndrome.² In this case, lumbar puncture was clinically contraindicated, but raised intracranial pressure may have occurred.

Clotting disorders have been reported to cause sub-ILM haemorrhage.⁸ The mild prolongation of the prothrombin time in this patient could be due to the consumption of coagulation factors as a result of the release of bacterial endotoxin.⁷ This mild clotting defect may have caused the preretinal haemorrhage in this case.

Spontaneous resorption of subhyaloid or sub-ILM haemorrhage caused by Valsalva retinopathy usually occurs without sequel.³ Pars plana vitrectomy has been recommended for dense preretinal haemorrhage resulting from diabetic retinopathy, as spontaneous resorption is usually prolonged in these patients.¹ Laser puncturing of the posterior hyaloid face or the internal limiting membrane

has been described as an alternative to vitrectomy.⁹ Ulbig *et al*¹⁰ recently reported the results of a series of premacular subhyaloid haemorrhages treated with pulsed Nd:YAG laser. Nevertheless, this procedure is not without complications. They reported one case of macular hole formation and one case of retinal detachment after the procedure.

We feel that premacular subhyaloid or sub-ILM haemorrhage caused by factors other than diabetic retinopathy may be managed conservatively in the first few months. Our case demonstrates that a good visual outcome can be achieved with conservative management.

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Reverse and converse ocular bobbing with synkinetic blinking and opsoclonus in a child with Epstein-Barr virus encephalitis after bone marrow transplant for MPS I

EDITOR.—Reverse ocular bobbing is an abnormal spontaneous eye movement in which the eyes move rapidly and conjugately upwards (fast phase), followed by a slow drift (slow phase) back to the primary position (that is, the reverse of ocular bobbing—fast conjugate downwards deviation, with a slow return up to the midline). This eye movement disorder may be seen in patients with viral encephalitis, metabolic encephalopathy, and in those with pontine lesions.¹ Converse ocular bobbing (also referred to as reverse ocular dipping or slow upward ocular bobbing), consists of a slow phase upwards, followed by a fast phase back to the primary position, and has also been reported in patients with viral or metabolic encephalopathy, and in those with pontine infarction.¹ Opsoclonus, also referred

to as "saccadomania" or "dancing eyes", is characterised by intermittent bursts of large amplitude high velocity multidirectional back to back saccades, and has also been reported in patients with viral encephalitis and metabolic encephalopathy, as well as in those with occult neuroblastoma and drug toxicity.² When these back to back saccades occur purely horizontally, they are known as "ocular flutter" and can be a stage of resolving opsoclonus. We report a case of converse ocular bobbing, reverse ocular bobbing, with synkinetic blinking, opsoclonus, and ocular flutter occurring in a patient with Epstein-Barr viral encephalitis.

CASE REPORT

A 2½ year old female with a delayed diagnosis of mucopolysaccharidosis type 1 (MPS I) underwent a bone marrow transplant (BMT). Preoperative assessment revealed hearing loss of 60-65 dB, marked ventricular dilatation, but no evidence of raised intracranial pressure. Her visual acuity was recorded as being 6/24 using Cardiff acuity cards, but it was felt that the vision may be better than this as the child was uncooperative during vision testing. She had moderate corneal clouding, and her retina and optic discs appeared normal. BMT preparative chemotherapy consisted of fludarabine/melphalan/antilymphocyte globulin, and the patient received an HLA matched unrelated donor bone marrow infusion, with additional graft versus host disease prophylaxis consisting of cyclosporin and methyl prednisolone. Fourteen days post-BMT, the neutrophil count had recovered (the BMT preparative chemotherapy induces a febrile neutropenia in these children, which typically recovers at 2-3 weeks post BMT), but the patient remained febrile on broad spectrum antibiotics and antifungal agents. Twenty nine days post-BMT, she became irritable, her pyrexia persisted, and her upper limbs became hypertonic. Her conscious level then deteriorated and computed tomograph (CT) scan revealed acute hydrocephalus with very large ventricles and an emergency external ventricular drain was inserted. Epstein-Barr virus (EBV) was detected in the cerebrospinal fluid by polymerase chain reaction (PCR), EEG showed generalised slowing with occasional sharp waves, and magnetic resonance imaging with gadolinium revealed extensive abnormal signals in the cerebral white matter. A diagnosis of EBV encephalitis was made. The patient was commenced on phenytoin in case there was underlying seizure activity. On examination of her eyes at that time, she was not fixing or following and had developed abnormal eye movements, which consisted of converse ocular bobbing in the initial phase (that is, slow phase up/fast phase down, see above). Each upward slow phase was accompanied by a blink. On observation 2 days later she had developed reverse ocular bobbing (that is, fast phase up/slow phase down) as oppose to converse bobbing. The upward fast phase was accompanied by a blink which she seemed to be attempting to overcome with her frontalis muscle which was overacting in synchronicity with the blinks. A repeat CT scan demonstrated resolving hydrocephalus and persistent cerebral oedema. Repeat lumbar puncture revealed decreasing EBV titres (as measured using quantitative PCR), which eventually became negative. There was a subsequent improvement in her condition, and her parents had noticed much less eye movement activity. However, on examination she was still demonstrating intermittent episodes of re-

verse ocular bobbing with synkinetic blinking, and now intermittent bursts of both opsoclonus and ocular flutter.

COMMENT

Ocular bobbing, dipping (also referred to as inverse bobbing—slow downward movement, followed by a fast upward movement to the primary position), and reverse bobbing have been reported on different occasions in the same patient,³ as have opsoclonus and ocular bobbing.⁴ Synchronism of inverse ocular bobbing (slow phase down/fast up) and blinking has been reported in a 7 year old female with severe cerebral trauma; the rapid phase of the eye movement was synchronous with phasic contraction of the orbicularis oculi.⁵

The above case of reverse and converse ocular bobbing with synchronous blinking, together with the later development of opsoclonus and ocular flutter in a child with viral encephalitis is the first such combination of eye movement disorders to be reported in one individual. The abnormal eye movements in this child may be a result of her encephalitis, mucopolysaccharidosis, or the treatment she received. It is difficult to determine which phenomenon or combination is responsible. When such abnormal eye movements occur either together or in isolation, viral encephalitis should be considered.

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Retinorhexis in macular translocation

EDITOR,—Age related macular degeneration¹ is one of the leading reasons for legal blindness in the western world.² Most therapeutic strategies, however, have so far been unsuccessful in restoring vision for the majority of patients. Recently, Robert Machemer suggested translocating the macula on healthy retinal pigment epithelium, outside the originally central region of the fundus.³ The method includes a circumferential peripheral retinotomy to mobilise the retina. The retinotomy has usually been performed by cutting the retina close to the ora serrata using scissors or the vitrectomy instrument.^{3–5} Using scissors can lead to complications since the underlying Bruch's membrane and the uvea can be injured. Using the vitrectomy

instrument for cutting the retina leads to a loss of retinal tissue since the vitrectomy instrument works by a combined action of suction and cutting. The purpose of this study was to report on an alternative technique for circumferentially mobilising the retina.

CASE REPORT

A 87 year old woman presented with long-standing rhegmatogenous retinal detachment with proliferative vitreoretinopathy, and an additional age related macular degeneration with a 2 mm by 3 mm large disciform subfoveal membrane. A three port pars plana vitrectomy was performed. After removal of epiretinal membranes and after pars plana phakectomy, the detached peripheral retina was grasped with a microforceps, and by gentle and repeated traction, the inner layer of the pars plana epithelium was separated from the outer layer of the pars plana epithelium. Additionally, a tear was produced between the inner pars plana epithelium and the non-pigmented epithelium of the pars plicata. By regripping and changing the position of the microforceps, the peripheral retina with the inner layer of the pars plana epithelium attached was circumferentially separated from the inner surface of the eye wall. Further surgical steps included retinal rotation, temporary injection of perfluorocarbon liquid to be replaced by silicone oil, and circumferential peripheral endolaser coagulation. The same procedure with peripheral retinorhexis was performed in the next patient aged 75 years and undergoing pars plana vitrectomy for macular rotation as treatment of age related macular degeneration.

COMMENT

One of the major complications of macular translocation for the treatment of age related macular degeneration is the development of proliferative vitreoretinopathy. Some of the reasons are the incision into the retina and the temporary detachment of the retina. Since the risk of proliferative vitreoretinopathy depends on the size of a retinal defect and on the area of exposed retinal pigment epithelium, one tries to perform the circumferential retinal incision with the least possible retinal destruction and least possible loss of retinal tissue. It may be accomplished by a retinorhexis using the technique described here. Retinal tissue is not lost since the tear in the tissue is located in the epithelium of the pars plana peripheral to the ora serrata. Consequently, retinal pigment epithelium is not, or only slightly, exposed. It may reduce the risk of postoperative proliferative vitreoretinopathy.

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Acute retinal necrosis presenting with scleritis and raised intraocular pressure

EDITOR,—Acute retinal necrosis (ARN) is a necrotising herpetic retinopathy (NHR), which commonly presents as a painless, rapidly advancing retinitis. We report a patient with ARN who was initially diagnosed with anterior scleritis.

CASE REPORT

A 50 year old woman presented to the casualty department with a 3 day history of intense pain and tenderness in her right eye. She was fit and well with no notable past ocular or medical history. The right eye had 6/12+2 Snellen visual acuity and normal Ishihara colour vision with no relative afferent pupillary defect. The intense hyperaemia did not blanch with 10% phenylephrine and demonstrated scleral oedema with red-free light. The intraocular pressure on the right was raised at 30 mm Hg. The left eye was normal, seeing 6/5.

The patient was treated with flurbiprofen 50 mg three times per day and an appointment was made for her in the scleritis clinic 3 days later when the pain had eased. However, she now noted a positive scotoma and funduscopy through moderate vitreous condensations revealed an inferior, pale area of retinitis and occlusive retinal vasculitis (Figs 1 and 2).

Figure 1 Acute retinal necrosis showing retinitis and vasculitis.

Figure 2 Acute retinal necrosis showing retinitis and vasculitis.

The clinical diagnosis of ARN was supported by a rapid increase in the area of retinal whitening over the next 2 days and confirmed by stabilisation in response to intravenous aciclovir treatment. Flurbiprofen 100 mg three times per day did not control her pain, and in view of optic disc swelling prednisolone 80 mg per day was prescribed together with dexamethasone 0.1% four times per day and cyclopentolate 1% twice per day. Betagan 0.5% twice per day was successful in controlling the intraocular pressure.

Three days after starting intravenous aciclovir the area of affected retina stabilised at about 5 disc diameters and argon laser was applied to wall off the area of necrotic retina from the posterior pole. The disc became gradually less swollen and increased vitreous activity dropped vision to 6/24. The dose of prednisolone was reduced slowly as the vitritis cleared. Final vision was 6/12 being limited by an epiretinal membrane.

COMMENT

Pain, redness and, sometimes, even scleritis may accompany ARN early in its course which should always be considered when these signs are present.^{1,2} The diagnosis of ARN is clinically defined as one or more areas

of retinal necrosis with discrete borders primarily located in peripheral retina, rapid circumferential progression, occlusive vasculopathy with arteriolar involvement, and inflammatory reaction in vitreous and anterior chamber.³

The one other disease that can have scleritis, high IOP, and retinitis is toxoplasmosis.^{4,5} Although polymerase chain reaction (PCR) for herpes and toxoplasma can distinguish the two, ARN was diagnosed clinically in view of the rapidity of advancement, rendering PCR unnecessary.⁶

Although herpes viruses may become active simultaneously in more than one site, the anterior scleritis was probably a local response to ARN.⁷

ARN should be considered in patients with raised intraocular pressure and scleritis because useful vision can be maintained with prompt treatment.

The authors have no proprietary interests in any product named in this report.

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