The retinal appearance suggested possible cartilage, as shown in Figure 2A, which displays hyper-reflective echolucent areas temporally (A). Similarly, Figure 2B illustrates a highly reflective echo inferotemporally (B), while Figure 2C shows hyper-reflective echo persisting at low gain (C), indicating possible cartilage. A conservative approach was taken, with an initial period of observation and occlusion therapy.

Case report

An 18-month-old girl presented with a distorted pupil apparently following a fall downstairs some 4 weeks previously. The right pupil was displaced inferotemporally, with a localised area of ectropion uveae and underlying segmental lens opacity. An urgent EUA confirmed the clinical findings. The retina and ora serrata were normal on funduscopy but there was the vague suggestion of a subtle whitish pars plana abnormality inferotemporally. Visualisation of this area was however hampered by the lens opacity. B-scan was normal but high resolution scan showed an inferotemporal peripupillary iris cyst with irregular thickening of the peripheral iris. The ciliary body appeared normal but a triangular opacity of moderate reflectivity arose from the inferotemporal pars plana and extended along a thickened anterior hyaloid face (fig 1). In the absence of a definite mass the tissue changes were thought possibly to be secondary to trauma, perhaps of a penetrating nature. It was decided to manage her conservatively with an initial period of observation and occlusion therapy.

Unfortunately, she was lost to follow up and re-presented 4 months later with a right mature cataract. A repeat EUA was performed and B-scan now showed a partial posterior vitreous detachment with cellular vitreous. High resolution scan showed extensive increased irregular echoes of variable reflectivity extending from the pars plana to the ciliary body and along the anterior hyaloid face to the posterior lens. Small echolucent areas were present as well as one area of hyper-reflectivity inferotemporally. Although there was no absolute shadowing posterior to this hyper-reflective area, it remained highly reflective at low gain suggesting possible cartilage (fig 2A–C). Lens aspiration was performed revealing a dense, vascularised cyclitic membrane and underlying white fibrous tissue circumferentially around the pars plana and ciliary body though no distinct mass. Inferotemporally this structure was cystic with an area of hard chalky tissue, corresponding to the bright echo on scan, which was again thought probably to represent cartilage. A diagnosis of possible medulloepithelioma was made and confirmed on subsequent histological examination of tissue biopsies and vitreous cytology. Unfortunately, during surgery the retina developed an inferotemporal retinal dialysis requiring vitrectomy and inevitably some retrolenticular tissue was lost to further histological analysis during this procedure. Subsequent enucleation and extensive histological examination of the globe confirmed the diagnosis of malignant medulloepithelioma, although no heterologous elements could be identified in the available tissue.

Comment

Limited information exists regarding the echographic features of medulloepithelioma.** We had the rare opportunity of examining a case of malignant medulloepithelioma by B-scan and high resolution scan at both early and later stages of development, showing significant progression of the condition. Our case presented with a pupil abnormality, localised segmental cataract and probable early cyclitic membrane arising from the pars plana but not with a typical ciliary body mass. Development of a cyclitic membrane is a recognised feature associated with medulloepithelioma.** We have demonstrated the value of high resolution echography in such cases particularly demonstrating early changes when little was clinically evident. Our case demonstrates that, even in the absence of a ciliary body mass, the suggestion of early or unusual retrolental membrane should alert one to the possible diagnosis of intraocular medulloepithelioma.
Drug induced enucleation with resultant chiasmal damage

Self enucleation or “oedipism” is a rare form of self mutilation, occurring with paranoid delusions, either as a result of a drug related toxic psychosis or in functional psychoses, such as schizophrenia. It is most often described in acutely psychotic patients, who have religious or sexual delusions. We report a case of self enucleation of a right eye in a 19 year old male—induced by drug psychosis. The force of autoenucleation traumatised the chiasm resulting in a left temporal field defect. Only two cases of chiasmal damage have been reported in the literature. Many drugs are known to cause hallucination leading to self mutilation. Deliberate self harm (DSH) is a well known entity which varies from mild (skin picking and hair pulling) to severe forms of self mutilation like self amputation and self enucleation. DSH is known to result from delusions and command hallucinations occurring in psychotic and mood disorders, dementia, personality disorder, drug misuse, and mental retardation. In adults, attempts at ocular damage are associated with acute psychosis, self enucleation being the extreme form of ocular mutilation.

Case report

A 19 year old man was admitted following attempted enucleation of his right eye during an acute psychotic episode after taking ecstasy, LSD, and excess alcohol. He described, “seeing an army of police officers attacking him.” He attempted to remove a “bomb” which had gone into his eye using a nail clipper and pliers. He was still in a state of psychosis running around aimlessly and had to be held to the ground by six people to prevent further self mutilation.

He was admitted to a general medical hospital in a state of psychosis and was treated with haloperidol. He had no known past psychiatric disorders. He is healthy, fit and well with no known physical illness. Full blood count, ura and electrolytes, and blood gases were normal.

Toxicology of urine and blood were positive for amphetamine; no test for LSD was performed.

He was transferred to the Royal Victoria Eye and Ear Hospital on the same day in a stable condition. Ocular examination revealed no perception of light in his right eye with severely chemosed eyelids and the globe could not be visualised. Visual acuity in the left eye was 6/6 with normal anterior segment and fundus examination.

Exploration of the right socket under anaesthesia revealed no identifiable structures except for orbital fat, inferior rectus muscle, conjunctiva, and Tenon’s capsule (fig 1). The optic nerve stump was not visible. A porous polyethylene orbital implant was inserted, the Tenon’s and conjunctiva sutured over it in separate layers. Considering the instruments and force used to extract the eye, surprisingly there was no apparent loss of conjunctival tissue, leaving adequate fornices for a prosthesis, which was fitted 6 weeks later.

Goldmann visual field showed left upper temporal quadrantanopia to the 14e and 13e targets, and a left hemianopia to the 12e target (fig 2), indicating trauma to the chiasm. The patient has been advised not to drive at present. A magnetic resonance image showed no visible abnormality of the chiasm or left visual pathway. The patient is being closely monitored by the psychological team and is attending a drug rehabilitation clinic.

References


Figure 1 Total absence of globe.

Figure 2 Goldmann field of the left eye.
Repeated intravitreal injection of triamcinolone acetonide for diffuse diabetic macular oedema

Recent studies have suggested that intravitreal triamcinolone acetonide may be a treatment option for diffuse diabetic macular oedema. Since the duration of the effect of an intravitreal application of triamcinolone acetonide lasts between 4 weeks and up to 9 months, the purpose of this study was to evaluate the effect of a repeated intravitreal injection of triamcinolone acetonide.

Case reports

The clinical interventional case series study included four patients with diffuse diabetic macular oedema who consecutively received a second intravitreal injection of about 20 mg triamcinolone acetonide 7.6 (SD 3.9) months (median, 6.7 months; range, 4.1–13.1 months) after the first injection, and for whom follow up was longer than 3 months. The second injection was carried out, since visual acuity had decreased again after an initial increase following the first intravitreal injection. Mean follow up after the second injection was 5.6 (SD 4.2) months (median 5.6 months; range 1–10.5 months). Mean age of the patients was 62.0 (5.0) years (range 56.8–67.8 years; median 61.7 years), refractive error ranged between a mean of −0.75 (SD 1.19) dioptres and +2.50 (1.38) dioptres.

After the first injection, visual acuity increased from 0.12 (SD 0.05) (range 0.08–0.20) to a maximum of 0.23 (SD 0.14) (range 0.10–0.40). Converting visual acuity measurements into the logarithm of the minimum angle of resolution (logMAR) showed a change in the minimum angle of resolution from 0.95 (SD 0.17) logMAR units to 0.71 (SD 0.29) logMAR units. After the second injection, visual acuity increased from 0.12 (SD 0.06) (range 0.08–0.20) to a mean maximal visual acuity of 0.18 (SD 0.06) (range 0.10–0.25). The minimum angle of resolution changed from 0.97 (SD 0.19) logMAR units to 0.77 (SD 0.17) logMAR units.

All eyes increased in visual acuity. After the first injection, and after the second injection, respectively, intraocular pressure increased to values higher than 21 mm Hg in three eyes. For all of these eyes, intraocular pressure could be normalised by topical antiglaucomatous treatment.

Comment

The data of this study may suggest that the repeated intravitreal injection of about 20 mg of triamcinolone acetonide as treatment of diffuse diabetic macular oedema can be associated with an increase in visual acuity again in those patients who as “triamcinolone responders” showed an improvement in visual acuity after a preceding intravitreal injection of triamcinolone acetonide. It is in agreement with a previous study on repeated intravitreal injections of triamcinolone acetonide for exudative age related macular degeneration.
Figure 4 Colour fundus photograph of the left eye showing severe papillary vasoproliferative changes at 12 months.

as Rochalimaea henselae lasting 6–12 weeks in the absence of antibiotic treatment. Orbital lesions occur in about 6% of cases. This case illustrates the papillary vasoproliferative changes that may occur with CSD. Retinal vascular proliferative changes may be of diagnostic significance and should be sought in patients with ocular CSD. Ocular manifestations of cat scratch disease include Parinaud’s oculoglandular syndrome, neuroretinitis, optic neuritis, focal choroidoretinitis, retinitis, exudative maculopathy, serous retinal detachment, vitreous inflammation, and in this report, retinal vasoproliferative lesions. An angiomatous lesion has previously been described in cat scratch disease. It has also been known to be associated with central retinal artery and vein occlusion, as well as neovascular glaucoma.

Doxycycline and rifampicin have been used to treat neuroretinitis in CSD, as they have superior penetration into the central nervous system and eye. Long-term prognosis is usually good, although some patients may acquire a mild post infectious optic neuropathy. The prevalence of CSD requires further study but it may be more common than expected in the general population or in patients with idiopathic uveitis. Therefore, it is important to look for papillary vasoproliferative changes that may suggest the diagnosis of CSD.

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Treatment of recurrent orbital haemangiopericytoma with surgery and proton beam therapy

Orbital haemangiopericytoma is a rare, potentially malignant vascular tumour, which can affect any part of the orbit. The treatment of choice is complete surgical excision but because of its vascular nature and tissue friability during surgery this is frequently not achieved. Difficulties in correctly diagnosing this tumour preoperatively compound this problem. Incomplete excision is associated with increased risk of local recurrence and metastatic disease, which are reported to occur in up to 43% and 15% of cases respectively, and can take many years to become manifest. Treatment options for orbital recurrence include orbital exenteration, further attempts at complete excision, or local excision and adjuvant therapy with radiotherapy, chemotherapy, or chemotheraphy. To our knowledge the use of proton beam therapy in controlling recurrent orbital haemangiopericytoma has never been described. We therefore report such a case.

Case report
A 61 year old woman presented with a second recurrence of a left medial orbital haemangiopericytoma 2 years following radical excision of the tumour via a transfrontal approach, and 14 years after initial excision. She was found to have an axial proptosis of the globe associated with a soft tissue swelling at the medial canthus (fig 1). Her visual acuities were unchanged from initial presentation at 6/6 in the right eye and 6/9 in the left eye. She also had restriction of the left globe on dextroversion resulting in diplopia, which, although longstanding, had deteriorated considerably as shown by the field of binocular single vision. Neuroradiological review was arranged and it was thought that further attempts at complete excision would be unsuccessful. This, together with the patient’s desire not to undergo further major surgery, led us to explore other treatment options. She therefore underwent further local excision via a medial orbital approach to debulk the tumour mass followed by proton beam therapy with a total irradiation dose of 50 Gy equivalent in 20 fractions over 28 days. The protocol for this management plan employed standard parameters, with treatment margins of 2.5 mm on either side of the tumour, and 2.0 mm at its posterior limit. She responded well to this treatment, and a subjective improvement in ocular motility. In the 7 years following her proton beam treatment serial magnetic resonance imaging (MRI) has revealed no further growth of the residual tumour mass (fig 2). She maintained a visual acuity of 6/9 in the left eye for 3 years following treatment but subsequently developed ischaemic optic neuropathy reducing her acuity to 5/60.

Comment
This patient, having already undergone extensive orbital surgery on two occasions, was reluctant to consider a further procedure. Although there is evidence to support the use of adjuvant external beam radiotherapy after conservative surgery,1 it was felt that because of the size and location of our patient’s tumour, the dose required was likely to cause significant damage to surrounding structures. A decision was therefore taken to explore proton beam therapy as a potential adjuvant treatment for our patient, particularly as our department has gained experience with its use in the management of uveal melanomas. Following discussion with our colleagues at the Douglas Cyclotron, Clatterbridge Centre for Oncology, it was decided that owing to the relatively superficial location of the tumour, proton beam therapy would be a feasible option.

High energy heavy charged particle irradiation with proton beams offers several advantages over external radiotherapy. The particles have minimal scatter and can be collimated into small beams delivering maximal ionisation as they stop. This results in the phenomenon of the Bragg peak which is “modulated” with depth to produce a precise uniform dose volume.4 As the depth of penetration can be controlled, the radiation can be targeted almost exclusively at the tumour mass with minimal irradiation of surrounding tissues.5 The radiation dose of 50 Gy equivalent was chosen empirically and based on the normal dose used for the treatment of choroidal melanomas as there is no precedent in using this treatment for orbital haemangiopericytoma.

Unfortunately, this patient developed ischaemic optic neuropathy which is a recognised complication of proton beam therapy, particularly when more than 2 mm of optic nerve is irradiated at 30 Gy equivalent.6 Because of the size and location of our patient’s tumour, there was a high risk of this complication occurring, as there would have been had external beam radiotherapy been used instead. Nevertheless, this report demonstrates that proton beam therapy can be effective in controlling recurrent orbital haemangiopericytoma.

Figure 1 (A) Before further local excision and proton beam therapy the left medial canthal swelling and lateral globe displacement is obvious. (B) 7 years after treatment the reduction in tumour mass is still clearly visible.
Bilateral surgically induced necrotising scleritis with secondary superinfection

Surgically induced necrotising scleritis (SINS) is a rare complication of ocular surgery that has been described after pterygium excision, cataract extraction, trabeculectomy, penetrating keratoplasty, strabismus surgery, and retinal detachment repair. We describe a rare case of bilateral necrotising scleritis complicated by a secondary polymicrobial infection following uncomplicated phacoemulsification and pterygia excision.

Case report

A 66 year old Samoan male, with type II diabetes, end stage renal disease, coronary artery disease, and gout underwent uncomplicated combined phacoemulsification and bare sclera pterygium excision (without antimetabolites) in the right eye, followed 1 month later by the same combined procedure in the left eye. Three weeks later, the patient developed severe right sided eye pain. An erythrocyte sedimentation rate was 98 mm in the first hour, and oral prednisone (80 mg/day) was initiated. A temporal artery biopsy was negative and prednisone was discontinued after 4 days of treatment. One week later, the patient developed left sided eye pain as well as a productive cough. Corneoscleral necrosis developed in the left eye and rapidly progressed to perforation.

On examination, the visual acuity was 20/40 right eye and light perception left eye. In the left eye (fig 1), there was ischaemic scleral necrosis with thinning, perforation, and iris prolapse at the nasal limbus.

Comment

We believe this complicated case of bilateral, rapidly progressing, necrotising scleritis in a post-surgical patient represents a case of bilateral surgically induced necrotising scleritis (SINS) complicated by secondary polymicrobial infection. SINS has been reported following bare sclera pterygia excision, though it is more commonly associated with adjunctive β irradiation, thiopeta, and mitomycin C. The time course of events, as well as the response to

References

immune suppression supports a primary autoimmune aetiology complicated, secondly, by a polymicrobial infection. Immune suppression, however, was initiated only after aggressive antimicrobial therapy.

The necrotising nature of the scleritis in our patient is consistent with the series by O’Donoghue et al in which a fourfold greater rate of necrosis occurred in post-surgical patients compared to a non-surgically induced scleritis population. In their series, the majority of patients (75%) had two or more surgical procedures before the onset of scleritis, and systemic immune suppression was necessary in 93% of patients. Our case similarly illustrates the need to consider immunosuppressive therapy in patients with postoperative necrosis.

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NOTICES

Worldwide clinical trials for new technique for early detection of eye disease
A unique new non-invasive technique for high resolution optical imaging of the eye is receiving global acclaim. By combining two high-resolution imaging technologies, the new technique provides doctors with 3-D images of the retina, macula and the optic nerve.

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British Oculoplastic Surgery Society
Call for papers for the 5th annual meeting of the BOPSS to be held on 15 and 16 May 2005 at The Belfry, Birmingham. The abstract submission deadline is 4 February 2005, and abstracts can be submitted online at www.bopss.org.