Echographic features of a case of malignant intraocular medulloepithelioma

Intraocular medulloepithelioma is a rare embryonal neoplasm, usually presenting in the first decade of life as a unilateral, cystic ciliary body mass arising from the non-pigmented ciliary epithelium. Histologically, medulloepithelioma is classified as non-teratoid (a pure proliferation of medullary epithelial cells) or teratoid (containing heteroplastic elements such as hyaline cartilage, skeletal muscle, or neuroreticular tissue). Either group can be subclassified as benign or malignant, the latter showing evidence of poor differentiation, increased mitotic activity, and local invasion with or without extraocular extension. We report the progressive echographic findings in a case of malignant intraocular medulloepithelioma. Echographic examination, by B-scan (10 MHz probe) and high resolution anterior segment scan (20 MHz probe), was performed during two examinations under anaesthesia (EUA), using the 13 system.

Case report

An 18 month old girl presented with a distorted pupil apparently following a fall downstairs some 4 weeks previously. Her 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 she 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.

Figure 1 High resolution scan showing moderately reflective opacity at the pars plana, extending along a thickened looking anterior hyaloid face to the posterior lens. No ciliary body mass present.

Figure 2 High resolution scan now showing increased echoes of variable reflectivity from pars plana to ciliary body and posterior lens. Cystic structure with echolucent areas temporally (A). Very highly reflective echo inferotemporal pars plana (B). Although only very slight posterior shadowing, hyper-reflective echo persists at low gain (C), suggesting possible cartilage.
PostScript

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Drug induced autoenucleation with resultant chiasmal damage

Self enucleation or “œdipism” 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 contralateral field defect secondary to 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 picking and hair pulling) to severe forms known entity which varies from mild (skin


Figure 1 Total absence of globe.

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 with no known past medical illness. Full blood count, urea 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. 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 I4e and 13e targets, and a left hemianopia to the I2e 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.

Figure 2 Goldmann field of the left eye.

Comment

This report presents a rare case of a self enucleation of the right eye in a young man. This severe trauma resulted in loss of vision in the right eye and partial visual field loss in the other eye. Avulsion of the optic nerve was found in the right eye. The left eye showed no evidence of ischaemia or sympathetic ophthalmia. The visual field defect of the left eye resulted from traumatic severance of the right optic nerve causing secondary chiasmal damage. There were no intracranial complications.

Life threatening complications may result from self enucleation, including intracranial or subarachnoid haemorrhage, cerebrospinal fluid leakage, and bacterial meningitis. Radiographic imaging is required to exclude intracranial bleeding, optic chiasmal injury, and bone fractures.

Self enucleation or “œdipism” is a rare entity which requires operative reconstruction of the orbit, and also neurological monitoring to identify and treat any possible intracranial complications. The other eye should be observed for possible sympathetic ophthalmia which may occur because of residual uveal tissue in the socket. Visual field of the remaining eye must be performed in the acute phase, as field loss secondary to chiasmal trauma or severe traumatic optic neuropathy may occur and may respond to intravenous steroids, though this is controversial. Field loss will have implications with regard to driving and some occupations and may even require the patient to be registered as partially sighted.

Long term psychiatric therapy should be implemented to prevent further self injurious behaviour, as according to literature severe forms of self mutilation after self enucleation can occur. This includes attempted suicide.

Though cases of severe optic neuropathy and chiasmal damage are rarely reported, this may be due to lack of symptomatology and failure to perform field testing. The importance of visual field analysis cannot be overemphasised, because of the potential implications for affected patients.

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References
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. All patients were fully informed about the experimental character of the therapy and had signed an informed consent.

The ethics committee of the university had approved the study. All patients received an 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.

References


Papillary vasoproliferative changes in cat scratch disease

A 23 year old man presented with a 10 day history of general malaise and a 1 week history of floaters in both eyes, predominantly in the right. He denied a febrile illness or exposure to animals. General medical history was unremarkable. He was not taking any medications.

On examination his visual acuity was 6/4 in both eyes. The left eye subsequently developed marked papillary vasoproliferative changes at 12 months (fig 4).

Comment

Cat scratch disease (CSD) is typically a benign and self limiting illness, caused by Bartonella henselae bacteria (previously known as Rochalimaea henselae). The diagnosis of cat scratch disease was confirmed by an elevated titre for Bartonella (Rochalimaea) henselae of 1:128 using the indirect fluorescent antibody test.

The patient was treated with 150 mg oral doxycycline twice daily and 100 mg oral rifampicin twice daily. The vitritis settled with prednisone. However the aneurysmal disc changes remained unchanged. At 1 year follow up his visual acuity was 6/6 in both eyes. The left eye subsequently developed marked papillary vasoproliferative changes at 12 months (fig 4).

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Ocular manifestations of cat scratch disease (CSD) include parinaud’s oculoglandular syndrome, neuroretinitis, optic neuritis, focal choroiditis, 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 diagnostic significance and should be sought in patients with ocular CSD.

As Rochalimaea henselae lasting 6–12 weeks in the absence of antibiotic treatment. Ocular lesions occur in about 6% of cases. This case illustrates the papillary vasoproliferative changes that may suggest the diagnosis of CSD.

Figure 4 Colour fundus photograph of the left eye showing severe papillary vasoproliferative changes at 12 months.

References

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, brachytherapy, or chemotherapy. 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 transcranial approach, and 14 years after initial excision. She was found to have non-axial protrusion 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. Neurosurgical 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 with a reduction in tumour size radiologically, 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.
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 tapered, the patient's best corrected visual acuity was 20/40 right eye and light perception left eye. In 2 months following presentation, while on a prednisone taper, the patient's best corrected visual acuity was 20/40 right eye. He remained at light perception in the left eye.

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 pterygium 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 B-scan ultrasound revealed choroidal thickening right eye and haemorrhagic choroidal left eye.

Scleral cultures were taken, and the patient’s corneoscleral perforation was emergently closed with cyanoacrylate glue. An extensive laboratory evaluation for systemic aetiologies of scleral necrosis was unrevealing except for a mildly elevated rheumatoid factor of 62 IU/ml (normal <40 IU/ml). Culture results demonstrated a bilateral polymicrobial infection, with *Streptococcus pneumoniae* and *Streptococcus viridans* in the right eye, and *Streptococcus pneumoniae*, *Streptococcus viridans*, and *Haemophilus influenzae* in the left eye.

The patient was treated with topical (gatifloxacin and vancomycin), intravenous (ciprofloxacin), and subconjunctival (vancomycin) antibiotics. In addition, oral anti-collagenase therapy (tetracycline and vitamin C) was employed. Repeat cultures taken on day 3 were negative. By day 4, however, the vision in the right eye had decreased to 20/200, the area of necrosis had enlarged and the pupil began to peak nasally. Systemic immune suppression was initiated with an intravenous pulse of 1 g of both cyclophosphamide and methylprednisolone. Within 48 hours of initiating immune suppression, granulation tissue began to fill the areas of necrosis and ocular pain subsided. Two months following presentation, while on a prednisone taper, the patient’s best corrected visual acuity was 20/40 right eye. He remained at light perception in the left eye.

References

Figure 2 Axial (A) and coronal (C) MRI images before proton beam therapy. The haemangiopericytoma is evident in the left medial orbit. (B) and (D) show the same views 7 years after conservative surgery and proton beam therapy.
immune suppression supports a primary auto-immune 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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The latest (redesigned) issue of Community Eye Health (No 51) deals with the gaps between aims of Vision 2020 and how far we are still from them, especially in Africa. For further information please contact: Journal of Community Eye Health, International Resource Centre, International Centre for Eye Health, Department of Infectious and Tropical Diseases, London School of Hygiene and Tropical Medicine, Keppel Street, London WC1E 7HT, UK (tel: +44 (0)20 7612 7964; email: Anita.Shah@lshtm.ac.uk; online edition: www.jceh.co.uk). Annual subscription (4 issues) UK£28/US$45. Free to developing country applicants.

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 if 4 February 2005, and abstracts can be submitted online at www.bopss.org.