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 neural 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 both normal on fundoscopy 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 retrolental 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 medullopithelioma 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.
Self enucleation or ‘oedipism’ is a rare form of ocular mutilation. It is a well described 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 severance traumatic optic neuropathy may occur 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 overemphasized, because of the potential implications for affected patients.

Acknowledgements
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References
Repeted 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 1.0–10.5 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 −0.79 (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 about 20 mg triamcinolone acetonide in 0.2 ml Ringer’s solution as previously described in detail.

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 5 eyes, respectively, intraocular pressure increased to values higher than 21 mm Hg in 5 eyes, respectively, intraocular pressure increased to values higher than 21 mm Hg in 5 eyes, respectively, intraocular pressure increased to values higher than 21 mm Hg in 5 eyes, respectively, intraocular pressure increased to values higher than 21 mm Hg.

Intraocular pressure ranged between 21 and 24 mm Hg. In all cases, the intraocular pressure decreased within 2 days after the injection.

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, predominately in the right. He denied a febrile illness and exposure to animals. General medical history was unremarkable. He was not taking any medication.

On examination his visual acuity was 6/4-1 in the right eye and 6/4 in the left eye. Anterior chambers were quiet in both eyes. The left eye subsequently developed marked papillary vasoproliferative changes at 12 months (fig 4).
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 externtion, 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 orbital medial haemangiopericytoma 2 years following radical excision of the tumour via a transfrontal approach, and 14 years after initial excision. She was found to have non-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. 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 oculomotor function. 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).

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. 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 neovascular glaucoma, and severe vision loss.

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, 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. 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. 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. 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 and perhaps its use should therefore be considered as an adjuvant in the treatment of recurrent disease, particularly when the tumour is in a superficial location.

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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.1,2 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. 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 (gatifloxicin 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.

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 B irradiation, thiotepa, and mitomycin C.3–6 The time course of events, as well as the response to
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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References

NOTICES

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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.