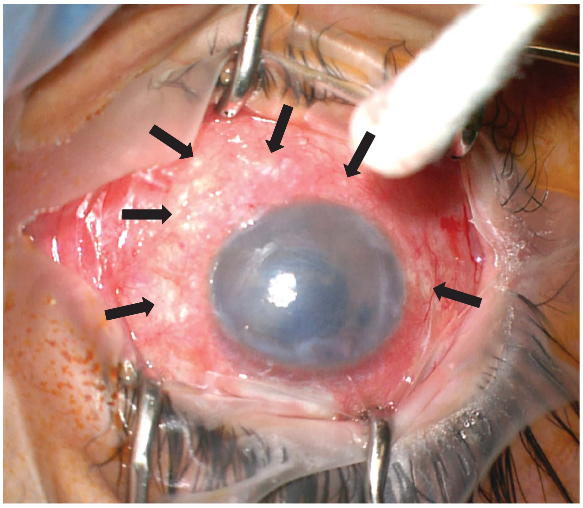

Figure 1 An intraoperative photograph shows extensive subconjunctival silicone oil accumulation (arrows).

the underlying Tenon's capsule and sclera in all quadrants secondary to oil and scarring. It was not felt that removal of the oil was feasible, as it was invested in the scarred conjunctiva. The previous sclerotomy sites could not be visualized. At postoperative month 2, the extensive subconjunctival silicone oil was still present.

Comment

Our case confirms that unsutured sclerotomies may be associated with diffuse subconjunctival oil spillage. Mild subconjunctival silicone oil leakage occurs commonly after vitrectomy.[1] Histopathological studies have found that small subconjunctival silicone oil deposits occur 30% of the time and often cannot be detected on slit lamp exam.[2] Using a similar wound construction technique as in our patient, case series with 23-gauge systems have detected small subconjunctival silicone oil bubbles 8–10% of the time, sometimes with mild postoperative discomfort.[3,4] We recommend a lower threshold to suture sclerotomies in silicone oil cases.

Acknowledgements

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Sir,

Delayed diagnosis of occult ocular juvenile xanthogranuloma mimicking non-accidental injury

We present a case of juvenile choroidal xanthogranuloma (JXG) in a child, mimicking non-accidental injury.

Case report

A 3-year-old boy presented with a 2-week history of drowsiness and left eye redness. His father reported that the child fell from a height of about 20 cm. The fact that a doubt had arisen for the cause of the eye problem raised the suspicion of non-accidental injury. No abnormality was detected on physical examination and magnetic resonance imaging of the brain. His past medical history was unremarkable. On ophthalmological examination, visual acuity was 0.2 LogMAR in the right eye and perception of light in the left eye. Slit-lamp examination revealed a total hyphaema in the left eye, while the right eye showed no ocular abnormality. The intraocular pressure was 63 mm Hg in the left eye and dropped to 10 mm Hg after washing-out of the blood from the anterior chamber. No iris lesions were evident. Additionally, there was no fundal view of the left eye due to a dense vitreous haemorrhage. Ocular ultrasound examination showed a thicker choroid temporally, but no evidence of retinal detachment or calcification.

Following a left eye vitrectomy and lens aspiration, the retina demonstrated retinal infiltrates and a pale optic disc (Figure 1). A vitreous biopsy was taken, and immunocytochemistry analysis showed the cells to stain strongly with CD68 and weakly with S-100. PAS was found to be positive within the cytoplasm. A diagnosis of choroidal JXG was made.

The family had been assessed by social services and no concerns had been raised. Fourteen months after the initial presentation, there is no involvement of the fellow eye.

Comment

Our case is the first to use a vitreous biopsy to diagnose this condition and the fourth in the literature of JXG with ocular involvement and absence of cutaneous manifestations.[1–3] If spontaneous hyphaema and vitreous haemorrhage are present, one should include in the differential diagnosis the possibility of JXG, along with non-accidental injury and malignancy, even in the

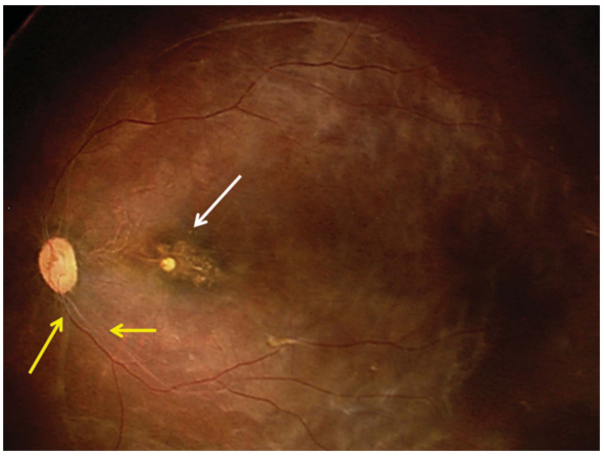

Figure 1 Fundus photograph of the left eye showing posterior segment involvement with sclerosed blood vessels and a pale optic disc (yellow arrows) and retinal infiltrates (white arrow).

absence of skin or systemic lesions. Consideration of this condition may prevent needless investigations for child protection.

Conflict of interest

The authors declare no conflict of interest.

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Sir,

Medulloepithelioma in DICER1 syndrome treated with resection

Medulloepithelioma is usually unilateral and arises from the nonpigmented epithelium of the ciliary body and rarely from the optic nerve. They generally occur in the first decade of life and present as a fleshy pink tan mass. We report here a familial cancer predisposition syndrome, which is not well documented in the ophthalmology literature.

Case report

A 16-year-old Caucasian female presented with blurred vision in her right eye. Her past medical history included ovarian Sertoli–Leydig tumor treated with resection, thyroid papillary tumor treated with thyroidectomy, pinealoblastoma treated with chemo-radiation, chronic renal insufficiency, and cystic disease of the kidneys and lungs. Her left eye was prephthisical secondary to anterior segment dysgenesis and had no light perception since early childhood. Visual acuity in the right eye was 20/60 and anterior segment examination showed posterior subcapsular and cortical cataractous changes. Fundus examination showed a white mass arising from the ciliary body (Figure 1a) with partial retinal detachment (Figure 1b), subretinal fibrous bands, and a giant retinal tear inferotemporally. Ultrasonography showed a densely hyper-reflective lesion of 8-mm thickness. Incisional biopsy via a lamellar scleral flap revealed cartilage suggestive of teratoid medulloepithelioma. The patient underwent transcleral excision of the residual ciliary body tumor with pars plana lensectomy, pars plana vitrectomy, partial retinectomy, silicone oil injection, and scleral buckling. Pathology showed scarce neuroepithelial cells with no malignant features and abundance of cartilaginous tissue consistent with teratoid cartilaginous medulloepithelioma of the ciliary body. At the second postoperative month, she had a visual acuity of 20/80 with aphakic glasses and her retina was attached. There was no evidence of recurrence 6 months postoperatively. Initial genetic testing was inconclusive for the *DICER1* gene mutation and the sample is being preserved for future study.

Comment

DICER1 gene (OMIM 601200) is a member of the ribonuclease III family and is inherited as an autosomal dominant trait with cytogenetic location 14q32.13.[1] The reported tumors with DICER1 syndrome are described in Table 1.[2] Priest *et al*[3] reported four patients with medulloepithelioma and pleuropulmonary blastoma, which possibly indicated DICER1 syndrome. Previous reports of local resection for medulloepithelioma have shown poor response with recurrence necessitating enucleation.[4,5] The role of chemotherapy and radiation therapy in patients with medulloepithelioma has not been well documented. Our patient showed no evidence of recurrent disease at last