may be precipitated by external factors like iron overload, alcohol, and oestrogens.

Ocular manifestations are caused by deposition of photoactive porphyrins in ocular tissues with lid scarring, ectropion, lacrimal scarring, scleromalacia, and corneal thinning described.³⁻⁷

Treatment involves avoidance of precipitating factors and UV exposure, while success has been achieved with phlebotomy and iron chelating agents.

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Sir,
Segmental neurofibromatosis

Neurofibromatosis (NF) is classified under phakomatoses, which are a group of disorders where

neurological abnormalities are combined with congenital defects of skin, retina, and other organs. Localised orbital neurofibromas are discrete, space-occupying lesions within the orbit and may be difficult to differentiate from other orbital tumours; multiple tumours can occur. We report here a case of a patient with orbital neurofibromas in a segmental pattern unassociated with systemic von Recklinghausen's disease.

Case report

A 52-year-old Caucasian woman was referred to the orbital eye clinic for management of a 10-year history of exophthalmos of her left eye (Figure 1). This was not associated with any diplopia in primary position or painful eye movements.

Examination revealed a systemically healthy woman, with no signs of thyroid status abnormality. The best-corrected visual acuities were right 6/6 and left 6/5. Palpation revealed a soft tender mass in the superior nasal region of the left orbit producing a 5 mm proptosis and 10 mm downward displacement. Extraocular movement examination showed a restriction of up-gaze of the left eye especially in the adducted position. Slit-lamp biomicroscopy showed normal anterior segments; no signs of optic nerve compromise were seen and fundus examination was normal.

Orbital magnetic resonance imaging demonstrated three discrete tumours in the left orbit (Figure 2). This prompted the diagnosis of neurofibromas and the patient was examined and found to have no stigmata or family history of NF 1 or NF 2.

The two more anterior masses were successfully removed, while removal of the more deeply seated tumour was abandoned owing to the close proximity to the neurovascular bundle in the inferior orbital

Figure 1 Clinical photograph showing exophthalmos and downward displacement of the left eye.

Figure 2 (a) (MRI showing separate tumours in the superior orbit and temporal fossa, sagittal section. (b) MRI showing associated subcutaneous lump in the left cheek, most probably a neurofibroma.

fissure and the fact that there was no threat to visual functions.

Histological examination of the two lesions disclosed a rather myxoid-looking lesions set in fibrous tissue. The cells had elongated nuclei and brightly eosinophilic cytoplasm. Immunocytochemistry showed S100 positive areas in the lesion and scattered axons were demonstrated with immunocytochemistry to neurofilaments.

Comment

NF is a heterogeneous disease and is classified into eight different types for prognostic significance and genetic implications by Riccardi.¹

The term segmental neurofibromatosis has been mainly used in dermatology to describe restricted distribution of café-au-lait macules and freckling, often in a unilateral, dermatomal distribution.^{2,3} This represents a

postzygotic, somatic mutation leading to mosaicism. Although in theory, offspring and other first-degree relatives of patients with segmental NF should be free of stigmata, there have been reports suggesting the possibility of familial transmission, especially in patients with bilateral Lisch nodules.^{4,5}

Along these lines, one could describe multiple orbital neurofibromas appearing also in a segmental fashion, affecting the intraorbital branches of the first division of the trigeminal nerve. In a similar manner, this would represent a postzygotic mutation with only the affected segment actually having the NF mutation. We believe that there are no clear diagnostic criteria for consistent reporting of such cases of segmental neurofibromatosis on the basis of a few case reports.⁶

The diagnosis of multiple neurofibromata is an important differential for discrete lesions in the orbit.⁷ This may represent a mild form of neurofibromatosis or a case of segmental NF. We recommend that the patient in these circumstances should be referred to a genetics specialist for a thorough family examination and possibly molecular investigations. The literature that has been reviewed here on the subject of segmental NF comes from an era where molecular biology testing was not available; therefore, the figures quoted are solely based on clinical and pathological specimens. In our case, the patient did not have any children and was of postmenopausal age; therefore, genetic testing was declined. It is important, however, to know the genetic implications for other family members, especially in cases of family planning. We feel that molecular studies are required in the field of segmental neurofibromatosis to define the incidence of the condition and the risk of transmission of the mutation to offspring.

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Sir,
Pars plana vitrectomy for traumatic cyclodialysis with persistent hypotony

Cyclodialysis is a disinsertion of the ciliary body from the scleral spur. It may occur accidentally by trauma, iatrogenically during intraocular surgery, or deliberately as a planned procedure for the treatment of glaucoma. Cyclodialysis clefts may result in hypotony, shallow anterior chamber, hypotony maculopathy, and possibly loss of vision. Different treatment modalities have been reported to repair traumatic cyclodialysis.

We describe a patient with traumatic cyclodialysis that was treated successfully with pars plana vitrectomy, gas tamponade, and cyclohexy with trans-scleral diathermy following the unsuccessful use of trans-scleral ciliary body sutures. Ultrasound biomicroscopy (UBM) proved helpful to identify precisely the location and extent of the cyclodialysis cleft and to observe the regression of the ciliochoroidal space postoperatively with the closure of the clefts.

Case report

A 27-year-old man was referred to us for decreased vision with persistent hypotony in the right eye (RE) after colliding with motorboat while jet skiing 3 months earlier. The patient had undergone reconstructive surgery for maxillofacial fractures. On examination, best-corrected visual acuity was 20/200 RE and 20/10 left eye (LE). The intraocular pressure (IOP) was 2 mmHg RE and 14 mmHg LE. The anterior chamber was shallow, and a cyclodialysis cleft was found superotemporally. Ophthalmoscopy revealed swollen optic disc and oedema of the retina with macular folds. The LE was unremarkable. The axial length was 19.21 mm in the RE and 23.39 mm in the LE.

Figure 1 Nasal cyclodialysis cleft (arrow) is found behind closed angle and supraciliary fluid (asterisk). Imaging was limited because of patient discomfort.

We performed a surgical cyclohexy by directly suturing the ciliary body to the scleral spur in the superotemporal quadrant, according to the technique reported by Kuchle and Naumann.¹ Following this treatment, no initial improvement was observed the IOP in the RE remained 2 mmHg. A posterior subcapsular cataract subsequently developed, and visual acuity decreased to 20/400. UBM (Humphrey UBM840, Humphrey Instruments, San Leandro, Ca, USA) examination of the ciliary body disclosed another cyclodialysis cleft nasally (Figure 1), that opened toward the anterior chamber and 360° of ciliochoroidal fluid.

At 2 months after the initial operation, we performed phacoemulsification of the lens, intraocular lens implantation, three-port pars plana vitrectomy, peeling of the posterior hyaloid membrane and fluid–gas exchange with 20% SF₆. At the end of the surgery, trans-scleral diathermy was applied posterior to the sites of the cyclodialysis clefts to anchor the ciliary body to the sclera firmly.

On the next day, the IOP showed a transient rise to 33 mmHg. The IOP decreased to normal range after the second postoperative day, and the ciliochoroidal detachment regressed. At 1 year after surgery, the best-corrected visual acuity improved to 20/20, and the IOP was 17 mmHg. UBM revealed closure of the nasal cyclodialysis cleft (Figure 2). The axial length of the right eye was elongated to 21.94 mm. Optic disc oedema and macular oedema had completely resolved.

Discussion

UBM provides high-resolution images of the anterior ocular segment and enables examinations focused on