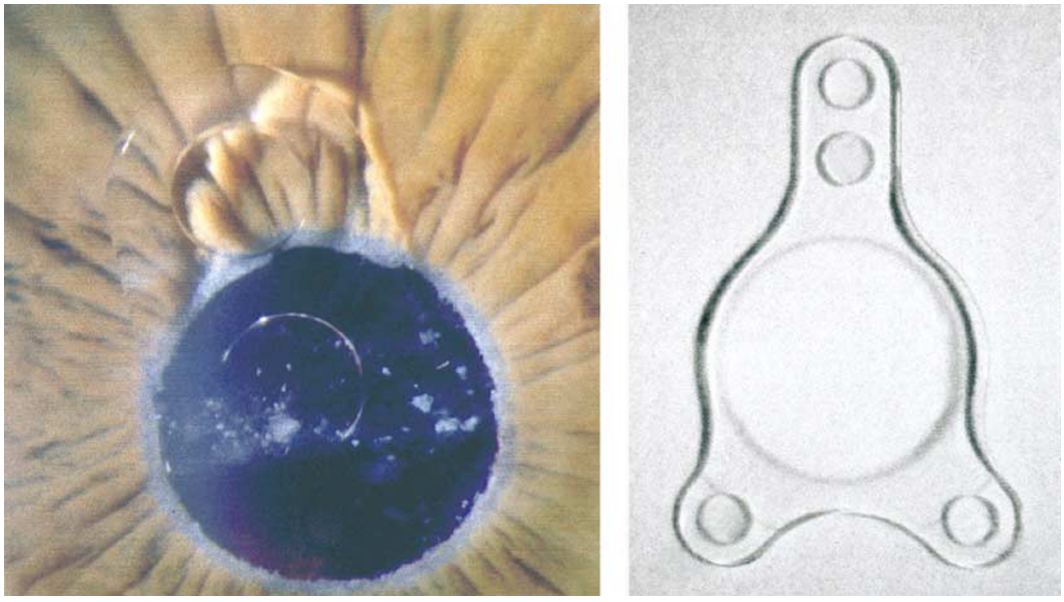

Figure 1 Clinical photograph of the displaced IOL haptic, accompanied by a photo of the Pearce tripod lens.

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Sir,

Bilateral microphthalmos and orbital cyst

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Microphthalmos with orbital cyst is a rare congenital cystic abnormality of the globe and orbit that is caused by faulty closure of the posterior part of the embryonic fissure. The cysts project through a congenital defect (coloboma) in the wall of a microphthalmic eye and are lined by a neuroectoderm.[1,2]

We present the clinical and histopathological findings of microphthalmos with orbital cyst in a 21-year-old woman who was followed for bilateral microphthalmos since birth and had recent onset of bilateral angle closure glaucoma.

Case report

A 21-year-old white female had been followed with the diagnosis of bilateral microphthalmos and no vision since birth. She had a long history of glaucoma and used many topical and oral medications. She was referred to the Ocular Oncology Service for evaluation of severe pain that developed in both eyes about 3 weeks earlier. She was otherwise healthy and had no family history of similar developmental anomaly.

Her visual acuity was no light perception in each eye. There was bilateral enophthalmos with microphthalmic globes, microcornea, corneal oedema, iris bombé with pupillary seclusion, flat anterior chamber and dense cataracts in both eyes (Figure 1a). There was no view of the fundus. Both globes were hard to palpation.

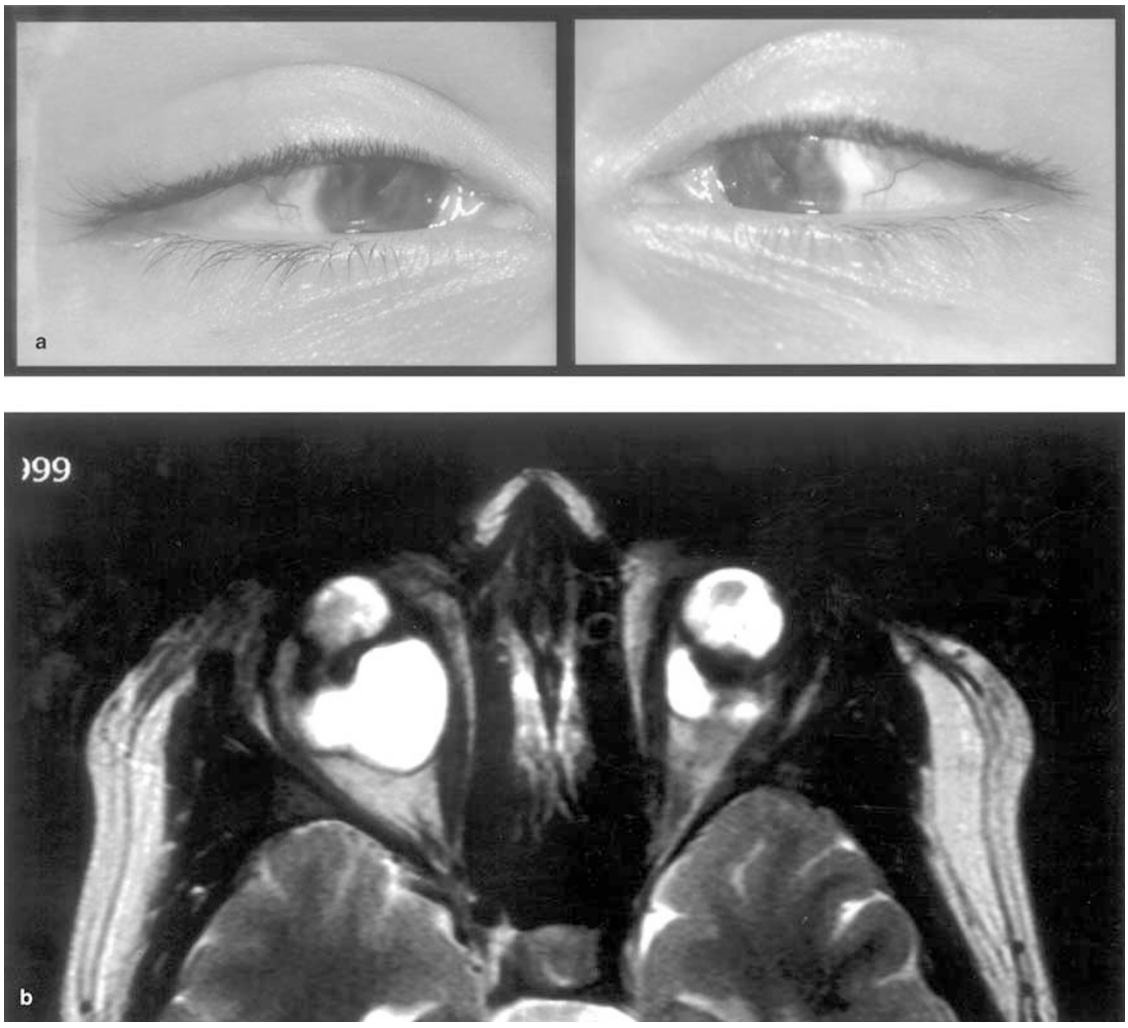

Figure 1 (a) External photograph shows bilateral microphthalmic globes with bilateral esotropia. (b) Magnetic resonance imaging displays bilateral microphthalmos with orbital cyst that is hyperintense to vitreous on T2-weighted images.

Microphthalmos precluded accurate tonometry. Ultrasonography showed microphthalmos and an acoustically hollow lesion measuring 20 mm in diameter, consistent with an associated cyst in the inferior quadrant of both orbits. Magnetic resonance imaging (MRI) confirmed bilateral lobulated nonenhancing intraconal cysts, which appeared hypointense to vitreous on T1-weighted images and hyperintense to vitreous on T2-weighted images (Figure 1b). Based on clinical and imaging findings, the diagnosis of bilateral microphthalmos with orbital cyst complicated by bilateral angle closure galucoma was made. Since her eyes were blind and painful, and had a poor cosmetic appearance, the patient requested bilateral enucleation. Both eyes and the attached orbital cysts were removed intact. The patient was relieved of pain and had improved appearance.

Gross examination disclosed bilateral microphthalmos with orbital cyst (Figure 2a). Histopathologic examination of both globes showed similar findings including a thickened and oedematous cornea, shallow anterior chamber, total posterior synechiae causing pupillary seclusion and occlusion of the anterior chamber angle by peripheral anterior synechiae (Figure 2b). A mass of glial tissue filled the vitreous cavity, and there was extensive fibrous and osseous metaplasia of the retinal pigment epithelium. In both globes, the retina was totally detached (Figure 2a). The orbital cyst was lined by gliotic neural tissue with foci of chronic inflammatory cells and contained proteinaceous fluid.[3]

Comment

Microphthalmos with orbital cyst is a congenital anomaly of the globe caused by a defect in the closure of the

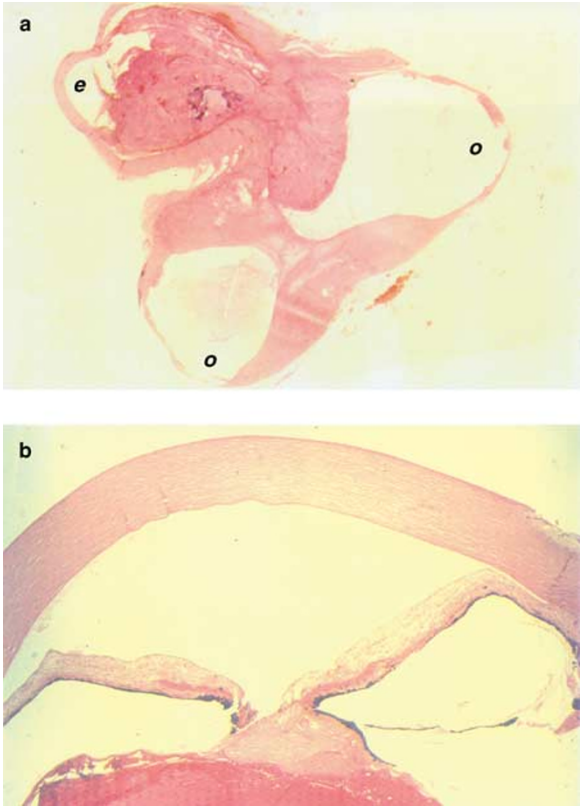

Figure 2 (a) Histopathology shows microphthalmos (e) with lobulated orbital cyst (o) on the right eye. A mass of glial tissue fills the vitreous cavity and extends into the cyst (haematoxylin–eosin, original magnification $\times 2$). (b) Pupillary seclusion, iris bombé and peripherial anterior synechia (haematoxylin–eosin, original magnification $\times 5$).

embryonic fissure at the 7 to 20-mm stage of development during the 6–7 weeks of gestation.[1] It is usually unilateral with bilateral cases observed rarely.[2,4,5] The vision is usually poor. Although the exact frequency is unknown, bilateral microphthalmos with orbital cyst can be associated with systemic abnormalities involving the central nervous system, renal, cardiovascular systems or chromosomal deletions, whereas unilateral cases usually are not associated with systemic abnormalities.[2,6] Our patient was unusual because she had bilateral disease but no associated systemic abnormalities.

There is limited information about glaucoma in bilateral microphthalmos with orbital cyst. Glaucoma associated with microphthalmos is usually a result of chronic angle closure. It results from pupillary block owing to the presence of a disproportionately large lens in a small anterior segment and other associated anterior segment developmental abnormalities.[7,8] Our case is unusual in that the patient had bilateral microphthalmos with orbital cyst that was complicated by bilateral angle closure glaucoma at age 21 years.

The patient with microphthalmos and orbital cyst may present in a variety of ways including pseudoanophthalmos, microphthalmos without detectable cyst or a protruding orbital mass. Imaging techniques such as computed tomography or MRI could be helpful in the diagnosis of microphthalmos with orbital cyst. In some instances, as in our case, the cyst is not recognized clinically in early years of life but was seen on orbital imaging studies in later years.[9] It is possible that in these cases, delayed diagnosis of glaucoma may contribute to blindness. Late-onset proptosis has also been observed due to massive gliosis in the cyst.[10] The management consists of observation in the majority of cases. Aspiration of the cyst or surgical iridectomy may also be considered depending upon the clinical findings. When the cyst is large and cosmetically unacceptable, or when the glaucoma develops in the blind microphthalmic eye, surgical removal is often the treatment of choice.

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Sir,

Should we discontinue tamoxifen in a patient with vision-threatening ocular toxicity related to low-dose tamoxifen therapy?

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Tamoxifen, a triphenylethylene nonsteroid oestrogen antagonist, has been widely used as an adjuvant postoperative therapy of oestrogen receptor-positive breast cancer. Its ocular toxicities, such as retinopathy, keratopathy, optic neuritis and cataract, have been reported since 1978, and tend to occur in patients who have a higher total dose and longer treatment.[1,2] These complications seldom cause significant visual impairment and, except for crystalline retinopathy, are reversible upon discontinuation of tamoxifen. We report a breast cancer patient who, despite the presence of vision-threatening ocular toxicity of low-dose tamoxifen therapy, made a full visual recovery of the left eye after cataract surgery without tamoxifen discontinuation.

Case report

A 45-year-old woman, a breast cancer patient with a history of receiving modified radical mastectomy in 1999, complained of progressive visual loss in her left eye for 5 weeks. Before her first presentation, she had received a cumulative dose of 1.86 g tamoxifen (20 mg/day) as postoperative adjuvant therapy for 3 months. Examination revealed a best-corrected visual acuity (BCVA) of RE 6/8.6 and LE 6/60. Anterior segment examination disclosed asymmetrical central posterior subcapsular opacity of both eyes, which was more severe in the left eye. No corneal opacity existed, and intraocular pressure and colour sensation were normal in both eyes. Funduscopic examination was bilaterally unremarkable. Electroculography (EOG) showed a subnormal Arden ratio: 180% in the right eye and 151% in the left eye. Flash electroretinography (ERG) for the maximal combined response disclosed slightly decreased amplitudes of both the cone and rod response in both eyes. Except 4-dioptre myopia, she did not have any ocular disease in the past. Her vision was 6/6 with myopic glasses before the initiation of tamoxifen treatment. A family history of ocular disease also did not exist. Although these ocular abnormalities were considered to be related to tamoxifen, the treatment persisted and the dosage was still 20 mg/day for fear of recurrence of breast cancer.

After 8 months, the BCVA deteriorated to RE 6/12 and LE 4/60. She decided to receive phacoemulsification and posterior chamber intraocular lens implantation of the left eye. Her BCVA of LE returned to 6/6 after cataract surgery. Then 4 months later, the acuity of LE went back to 6/15. After Nd:YAG capsulotomy, her vision returned to 6/6 again. However, the right eye acuity continued to get worse. A year after the cataract surgery, the BCVA of RE deteriorated to 6/30 while LE remained 6/6. The central posterior subcapsular opacity of the right eye became denser (Figure 1). The specific whirling corneal opacity and crystalline retinal deposit still did not exist (Figure 2), but the Arden ratio decreased to 119% in the right eye and 124% in the left eye. Amplitudes of ERG did not worsen any further in both eyes. We inferred that the impaired vision of the right eye resulted from the progressively denser posterior subcapsular lens opacity. The low-dose tamoxifen treatment was kept on to meet its standard regimen.

Comment

Low-dose tamoxifen ocular toxicity is well documented and its incidence was reported to be 6.3 and 12% in two