

EUROPEAN OPHTHALMIC PATHOLOGY SOCIETY

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Case number: pathology lab nr. 22-055
Material distributed: 1 glass slide (H&E stain)

Painful “Phthisis”

Clinical History

In 3/2021, the 60 year old male patient was referred to our Department for further treatment of his painful phthisical left eye. In 1983, he had suffered a penetrating injury from a metal foreign body to this eye and was treated using various surgical procedures including those for retinal detachment. Nevertheless, the eye became blind shortly afterwards.

At presentation, his right eye showed no significant pathology while his left (aphakic) eye displayed a fairly clear cornea with some small keratic precipitates, a mildly rubeotic iris with anterior synechiae and an irregular pupil. The retina appeared somewhat pale with areas of detachment and scars, and IOP was 10 mmHg. A conservative treatment with topical steroids, atropin and lubricants was effective for half a year but then he came back with recurrent pain and insisted on enucleation. Clinical findings were unchanged compared to his presentation 6 months earlier.

Ocular Pathology

Macroscopy.

left globe 21.5 x 20 x 19 mm with 13 mm of optic nerve attached. The cornea (11 x 10.5 mm) is concave but fairly clear. Transillumination is – apart from some shadowing in the lower half - unremarkable. The most intriguing finding is a collection of various types of mostly flat artificial material sewed to the upper posterior sclera. The eye is opened horizontally. Now extensive retinal/choroidal scarring is evident with atrophy of the RPE and a partially detached retina. In addition, some focal retinal hemorrhages are seen. In the anterior segment, whitish remnants of the lens capsule are present, but further details cannot be evaluated due to the cornea bent backwards.

Light microscopy.

The cornea exhibits a flat epithelium overlying a mostly intact Bowman's layer, early band keratopathy near the limbus and some peripheral neovascularization. Descemet's membrane is grossly thickened with occasional guttae, and the endothelium is somewhat vacuolated but still fairly cellular. The chamber angle is occluded due to anterior synechiae on one side and fibrous tissue on the other. The trabecular meshwork itself appears scarred. The iris contains focal chronic inflammatory cell infiltrates especially near the sphincter, and the iris pigment epithelium reveals a partial schisis-like splitting. Of particular interest, however, is a complete endothelialisation of the anterior chamber with endothelium covering the iris surface and some lens remnants and extending backwards across the ciliary body onto the peripheral retina where it fuses with an epiretinal membrane. Around the posterior half of the globe, the extraocular material attached to the sclera shows only a mild tissue reaction but occasional round cell infiltrates in particular near one ciliary

nerve and at presumably trauma-associated perforation sites in the pars plana/ora serrata region. The retina is partially detached (and atrophic) and in other areas attached to the choroid through scar tissue. Where the RPE is still in situ, it reveals focal proliferations but also calcification and early bone formation especially where some of the extraocular material has migrated into/through the sclera. The optic nerve is atrophic.

There is no unequivocal evidence for sympathetic ophthalmia.

Immunohistochemistry.

The endothelial membrane reacts strongly with anti-vimentin and somewhat with S100, and occasional cells react with CKpan, CK7 and CK18

Diagnosis: Endothelialisation after perforating trauma and indenting retinal surgery

Discussion

From the macroscopic appearance, our first interest were the various “retinal implants” sutured to the sclera near the optic nerve since their shape and number were quite unusual to us. However, in retrospect, those could be found in the older ophthalmic surgery catalogues and had been used for a certain time until the area of vitrectomy came up. Certain sponges are still used nowadays, but indenting surgery seems slowly to become less relevant within the surgical repertoire. In this eye, some of the problems caused by these implants are evident; on the other hand, they lasted for almost 40 years in this particular instance.

Thus, from a histopathological point of view, the advanced endothelialisation fusing with an epiretinal membrane is much more impressive. For quite some time there must have been a balance between obstructing the outflow and hindering the production of aqueous by overgrowing the ciliary processes. Also it is still intriguing to speculate on the factors that might stimulate the proliferation and migration of endothelial cells. Apparently, a damaged or occluded chamber angle (which is also present in our specimen) seems a precondition for an endothelial outgrowth, perhaps due to the lack of a functioning contact inhibition by the trabecular meshwork cells. On the other hand, abnormal endothelial cell function – despite a clear cornea! - is also reflected in the abnormal thickening of Descemet's membrane (clinically the other cornea was completely normal). Furthermore, in the cases described in the literature so far the endothelial spread usually stops on the iris. In our specimen, however, it extends far back over the pars plana and seems to fuse with an epiretinal membrane derived from mostly RPE cells.

References

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