European Ophthalmic Pathology Society

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Case number:	pathology lab nr. 22-871
Material distributed:	1 glass slide (H&E stain) and 1 scanned slide

Phthisical Eye

Clinical History

A 57 year old male Caucasian patient was referred for increasing pain in his blind left eye. He had suffered a penetrating injury as a teenager in the 80ies with subsequent surgery the details of which he couldn't remember. Now the conjunctiva was slightly swollen, the anterior chamber was extremely shallow with iridocorneal contact, and the cornea was cloudy with superficial calcification and deep neovascularisation, and the pupil appeared yellow. On ultrasound, no worrisome structures were identified. The right eye was normal with full visual acuity and an uneventful ocular history.
As for his general health, the patient suffered from hypertension, hypercholesteremia and cardiovascular disease (1 stent) but was otherwise healthy.
Due to the patient's symptoms it was decided to remove the eye.

Ocular Pathology

Macroscopy. left eye 19 x 22 x 19mm with an encircling band; the cornea (9x11 mm) is cloudy. The optic nerve ist cut short and looks atrophic. There is no transillumination. An attempt to open the eye horizontally has to be postponed because of a massive calcification; so the globe is decalcified and then further sectioned. The cut surface appears reddich-whitish without any further details of ocular structures.

Light microscopy. Paraffin sections show a cornea with an irregular epithelium that is – with Bowman's layer almost completely missing - in immediate contact with the underlying stroma. The epithelium as well as the stroma appear somewhat oedematous, and there are numerous vessels pervading the collagen lamellae. However, no evidence of infection or some other marked inflammatory infiltrate is visible. Descemet's membrane is intact but over a long distance split with the lamellae being separated by blood. Instead of by endothelial cells, the posterior part of Descemet's is lined by fibrous tissue and some pigment-containing cells. The anterior chamber and the chamber angle are filled with blood, and also the iris is largely hemorrhagic; only a few anterior synechiae are recognizable in the periphery. On one side of the section, the angle is recessed, and here the ciliary body is detached by blood. In deeper levels, the iris root is completely separated from the chamber angle. The lens can be identigfied only as a remaining calcified nodule (in oarts with cholesterol clefts and hemorrhage), surrounded by fibrous tissue but without any identifiable lens capsule. Arising from the ciliary body/ the ora serrata there is a cyclitic membrane without significant inflammation, attached to the degenerate funnel-shaped and completely detached retina. The remaining retinal tissue is gliotic (only a few ganglion cells are still present), with hyalinised vessels, focal RPE proliferations and sub- as well

as preretinal membranes. Starting from the ora serrata towards the posterior pole, there is an almost circular bone formation at the level of the previous RPE (Bruch's membrane can only be identified focally), touching the calcified lens remnants anteriorly. Most of the vitreous cavity is filled with blood and exudate.
The optic nerve shows an advanced atrophy. Outside the sclera, the encircling band can be identified with some residual suture material but there is no major inflammation neither around this foreign material nor within any of the ocular structures themselves. Thus, there is no evidence of sympathetic ophthalmia at this stage.

Diagnosis: phthisical eye with massive ossification, lamellar splitting of Descemet's, loss of lens capsule, retinal detachment, encircling band and optic atrophy

Follow-up

The patient had an uneventful recovery with no further problems during the following years.

Discussion

Phthisical eyes with calcification and bone formation and sometimes even hematopoesis are a frequent phenomenon, especially in the context of chronic inflammation. There is still some debate on the origin of those "osteoblast" and the exact mechanism by which ossification occurs. Through inflammation-triggered promotion of various cytokines – involving especially bone morphogenic protein 7 - , transformed RPE has been suggested as the most likely source for osseous metaplasia. At the time of enucleation, no significant inflammation was present in this specimen but we don't know what has been going on in earlier years.
There are also some non-inflammatory conditions/tumour entities that tend to be associated with ossification such as teratoma, medulloepithelioma or osteoma. Here, the pathway might be somewhat different.
What is, however, quite unusual in this eye is the apparently spontaneous disappearance of the lens capsule (especially with no evidence of any major inflammation) as well the splitting of Descemet's membrane both of which shall be discussed at the meeting.

References

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