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At 34 years of age, her best-corrected visual acuity remained 20/30, but visual fields had decreased to only a central island. OCT was performed using topographical mapping and longitudinal reflectivity profile (LRP) analyses. Retinal thickness topography in the patient differed dramatically from normal (fig 1A). Especially notable was the abnormally thickened retina along the arcades (fig 1B). A difference map highlights parafoveal thinning and patches of thickened superior and inferior retina (fig 1B, inset). Laminar architecture was explored using LRP’s overlaid on cross-sectional images from the fovea into the superior retina (fig 1C,D). The patient had laminated but thinned retina in the parafovea, and, with increasing eccentricity, there was a coarsely laminated and thickened region. At further superior loci, the retina had normal thickness but was delaminated. A more detailed comparison was made of LRP’s at three eccentricities (fig 1E). At the parafoveal locus, thinning could be accounted for by missing retinal layers, specifically loss of photoreceptor wavefront components. At the more superior loci, whether increased in thickness or not, the patient’s retina had no comparable lamination with normal retina.

Comment

The OCT results in this patient with retinitis pigmentosa and PDE6B mutations are complex but interpretable. Parafoveal thinning is attributable to rod (and cone) photoreceptor layer losses. The remarkable thickening and loss of normal laminar pattern at further eccentricities is probably an OCT marker for retinal disorganisation. Thickened and dysplastic-appearing retina on OCT scans has been previously reported in two early-onset retinal degenerations with a developmental component: one, a form of Leber congenital amaurosis caused by NR2E3 mutations,9 and the other, enhanced S rod-cone dysplasia 1 (Neuromuscular disease).10 The OCT B-scan demonstrates parafoveal thinning in a patient with retinal degeneration, which can be seen in the OCT images in this patient.”

Changes in the retinal inner limiting membrane associated with Valsalva retinopathy

Valsalva retinopathy was first described in 1972 by Thomas Duane as “a particular form of retinopathy, pre-retinal and haemorrhagic in nature, secondary to a sudden increase in intraocular pressure.” Incompetent or no valves in the venous system of head and neck allow direct transmission of intraocular or intra-abdominal pressure into the head and neck. Sudden elevation of venous pressure may cause a decompression in the retinal capillary bed, with subinternal limiting membrane haemorrhages (Hg) that rarely may break through and become subhyaloid or intravitreal.1

We report histological findings of internal limiting membrane (ILM) peel in a case of Valsalva retinopathy.

Case report

A 41-year-old Caucasian male was referred to the vitreoretinal services with a spontaneous and sudden loss of vision in left eye for 3 weeks. There was no history of trauma or violent exertion but the patient had hay fever and had frequent episodes of sneezing. On examination his vision was 6/6 and hand movements in right and left eyes, respectively. Anterior segment examination (“segment exam”) was normal. Dilated fundoscopy revealed a dense vitreous haemorrhage in the left eye and normal fundus appearance in the right eye. Ultrasound echo-graphy revealed a posterior vitreous detachment, vitreous haemorrhage and a macular elevation in the left eye. Systemic examination was normal. Laboratory investigations showed normal complete blood count, prothrombin time and activated partial thromboplastin time. Blood pressure and urine analysis were normal. An MRI brain was normal.

After discussions with the patient, a decision was made to perform a 20-gauge three-port pars plana vitrectomy. Intraoperatively, after core vitrectomy and removal of the vitreous haemorrhage, a sub-ILM haemorrhage typical of Valsalva retinopathy was noted. ILM peel was performed without the assistance of dye, and the excised tissue was processed for histopathological assessment. Postoperatively, 3 months later, the patient’s vision improved from 6/6/6 unaided, with no secondary complications.

Histological examination of the excised tissue (fig 1A) revealed that it contained convoluted ILM. The vitreous (smooth) surface of ILM was free of cells, but there was a cellular component in the specimen, and this component was on the retinal side (undulated surface) of the ILM (fig 1B). The cellular component included a prominent multilayer aggregate of cells that was immunoreactive for cytokeratin 7 (fig 1C), which is a marker of transdifferentiated retinal pigment epithelial (RPE) cells. These cells were negative for glial and neural markers. Nevertheless, glial and neural elements were present in the specimen, again on the retinal rather than the vitreous surface of the ILM (fig 1D,E). CD68-positive macrophages were scattered throughout the specimen and there was also scattered pigment that was partly intracellular and partly extracellular. Perls (Prussian blue) staining confirmed that the pigment was a mix of melanin and haemosiderin (fig 1F).

Discussion

The plane of retinal Hg in Valsalva retinopathy is sometimes difficult to determine, especially in the absence of PVD. Ocular coherence tomography (OCT) has been used to determine the exact location when the vitreous medium is clear and it is generally agreed that it is sub-ILM in location. Following core and posterior vitrectomy, we could confirm that a sub-ILM haemorrhage was present. The Hg was possibly a consequence of the patient’s hay fever-related sneezing that is thought to occur from a sudden rise in the intraocular pressure caused by a forceful exhalation against a closed glottis.

Therapeutic options in Valsalva retinopathy include conservative management, surgery (vitrectomy) and laser membraneotomy. Epiretinal membrane (ERM) formation with ILM wrinkling has been reported 10 months after ND-YAG membraneotomy of Valsalva Hg. Histological examination of surgically removed ILM revealed the presence of haemosiderin within macrophages on the retinal side of the ILM and a fine glial ERM, resembling glial proliferation on the vitreous surface of the ILM.

Our case also revealed haemosiderin on the retinal surface of the ILM, again confirming the sub-ILM location of the haemorrhage, but
Sections through the excised internal limiting membrane (ILM). (A) Staining with periodic acid Schiff reagent and haematoxylin reveals convoluted ILM with adjacent cells. (B) High-power view of section in (A) to show that the cells are on the undulated (retinal) surface of the ILM (arrows) whereas no cells are seen on the smooth (vitreous) surface (arrowheads). (C) Stained with the immunoperoxidase technique for cytokeratin 7 (red-brown chromogen) and counterstained with haematoxylin; layers of transdifferentiated retinal pigment epithelial (RPE) cells are observed (arrows). Inset: higher magnification demonstrates that the RPE cells are adjacent to the undulated (retinal) surface of the ILM (arrow). (D, E) Sections stained with the immunoperoxidase technique (red-brown chromogen) for the glial marker glial fibrillary acidic protein (D) and the macrophage marker CD68 (E), respectively, and counterstained with haematoxylin. (D) Gial cells are abundant in the tissue but have a distribution different from that of RPE cells (compare with C). (E) Macrophages are more scattered throughout the tissue. (F) A section stained with Perls method reveals iron of presumed blood origin (blue deposit; arrow) as well as melanin pigment (arrowhead).

instead of an ERM there was a mixed-cell-type proliferation on the retinal surface of the ILM. The sub-ILM cells included trans-differentiated RPE cells, and hence the proliferation had the histological appearances of a proliferative vitreoretinopathy (PVR)-type membrane “beneath” the ILM. Presumably, the RPE cells had been attracted to this location by the sub-ILM blood, since it is well established that RPE cells migrate to various blood components and can move through intact retina 5.

Intraretinal pathology in PVR is a well-recognised pathological event, but usually the retinal changes resemble gliosis 6. Our case suggests that focal RPE proliferation, similar to that seen in PVR epiretinal membranes, can occur within the neuroretina, and specifically in a sub-ILM location, by transmigrated RPE cells as a response to intraretinal haemorrhage. Such proliferation might prevent complete visual recovery after reabsorption of the retinal haemorrhage and justify early surgical intervention instead of routine observation or laser membranotomy.
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