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Bilateral hazy vitreous in a patient with convulsions

Case

A 41-year-old gentleman presented with complaints of decreased vision and floaters in both eyes (OU) since six months with a history of convulsions for six years. Best-corrected visual acuity (BCVA) was 20/200 OU, with normal intraocular pressures. The slit-lamp examination showed quiet anterior segment OU [Fig. 1a and b]. Fundus examination after pupillary dilatation revealed dense vitreous opacities that prevented a clear fundus view [Fig. 1c]. The anterior vitreous showed strands of opacities with numerous focal attachments to the posterior lens capsule. Ultrasound B-scan revealed plenty of low reflective dot echoes with an attached retina and a normal choroid [Fig. 1d].

What is your next step?

A) Topical steroids
B) Anterior chamber tap for an infective pathology
C) Vitrectomy - diagnostic/therapeutic
D) Magnetic resonance imaging to rule out primary central nervous system lymphoma.

Findings

The slit-lamp examination showed the presence of vitreous opacities that attach to the posterior lens capsule by "foot plates" or "pseudopodia-lentis" suggestive of vitreous amyloidosis [Fig. 1a and b]. The history of convulsions further supports the diagnosis of familial amyloidotic polyneuropathy (FAP). The patient underwent pars plana vitrectomy in the right eye. The histopathology of the vitreous sample demonstrated acellular eosinophilic material on hematoxylin and eosin stain [Fig. 1e] with strongly positive Congo-red staining [Fig. 1f]. Vision improved to 20/20 in OD at 6 weeks of follow up after vitrectomy. MRI of the brain and orbit was advised at the discretion of his neurologist. The patient was advised for vitrectomy in the left eye.

Diagnosis: Vitreous amyloidosis.

Correct Answer: C

Discussion

Vitreous amyloidosis occurs in familial amyloidotic polyneuropathy (FAP), caused by mutations in the transthyretin (TTR) gene.[1] TTR is a tetrameric plasma protein (prealbumin) which polymerizes into a beta-pleated structure of amyloid fibril and deposits in the peripheral nerves (80-90%), cardiac muscle (80%), kidneys (6%), and eye (10%).[2] Leptomeningeal involvement in FAP often manifested as convulsions and was reported in 5% of cases.[3] Ocular findings include abnormal conjunctival vessels, keratoconjunctivitis sicca, pupillary abnormalities, vitreous opacities, and secondary glaucoma. The incidence of vitreous opacities varies from 5.4% to 35% and the density of the opacities determines the visual acuity and symptoms.[2] Clinical features of vitreous amyloidosis include pseudopodia lentis, glass wool vitreous, and retinal perivascular deposits. Pseudopodia lentis are whitish opacities or the foot plates, present on the posterior surface of the lens capsule which extend into the opacified vitreous.[3] Treatment includes partial vitrectomy, with residual vitreous acting as a filter that prevents the amyloid material from reaching the angles and causing or exacerbating an existing component of glaucoma.[3]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

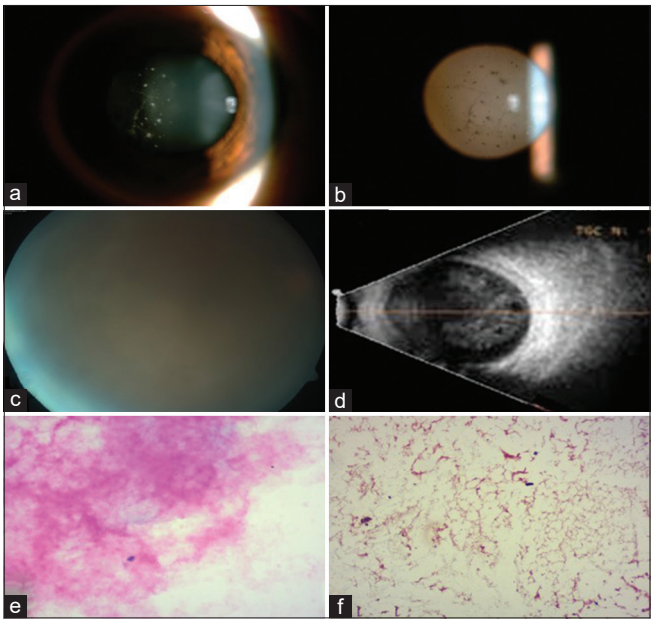

Figure 1: (a) Slit-lamp photo of the right eye showing vitreous opacities attached to the posterior surface of the lens capsule. (b) Retroillumination photograph showing the vitreous fibrils attached to the posterior lens surface. (c) Dense vitreous haze obscuring the fundus view. (d) USG B scan showing plenty of low reflective dot echoes in the vitreous with an attached retina and a normal choroid. (e) H and E staining of the vitreous sample post vitrectomy demonstrating eosinophilic material. (f) Positive Congo red staining

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Conflicts of interest

There are no conflicts of interest.

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