CASE REPORT

A Case of Atypical Idiopathic Choroidal Effusion Syndrome
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ABSTRACT

Uveal effusion syndrome is characterized by annular ciliochoroidal detachment, shifting non-rhegmatogenous retinal detachment, unremarkable inflammation in the anterior eye segment and normal intraocular pressure. A 36-year-old Caucasian hypermetropic male presented in the eye casualty with a week history of curtain like effect in front of his left eye associated with worsening of vision and flashing lights. Left fundus examination revealed retinal detachment with smooth shaped elevation superonasaly and detached retina inferiorly confirmed on ultrasound B-scan with no tobacco dust in the anterior vitreous. Diagnosis of uveal effusion syndrome was made. Lamellar sclerectomies in the two quadrants were performed. Fourteen months postoperatively, the left eye choroidal effusion gradually settled down and at 18 months the retina flattened. Early diagnosis, close follow-up, and appropriate management are mandatory to improve or maintain visual function in such patients.

Key words: Uveal effusion syndrome. Lamellar sclerectomy. Non-rhegmatogenous retinal detachment.

INTRODUCTION

Uveal effusion syndrome typically affects otherwise healthy middle-aged men. It is characterized by annular ciliochoroidal detachment, shifting non-rhegmatogenous retinal detachment, unremarkable inflammation in the anterior eye segment and normal intraocular pressure.1

In uveal effusion an abnormal collection of fluid expands the suprachoroidal space, producing internal elevation of the choroidal layer. There are various inflammatory and hydrostatic conditions that can cause uveal effusion, but in some cases no obvious cause exists.2

It may be idiopathic, inflammatory (due to a trauma, ophthalmic surgery, choroiditis, sympathetic inflammation, Harada's disease, scleritis, on pan photocoagulation) or hydrostatic (due to an arteriovenous shunt located in the dura mater, hypotonia, haemorrhagic leak or microphthalmos) and escitalopram-induced uveal effusions.3

Secondary uveal effusion syndrome may develop after argon laser therapy,4 or accompany anterior ischaemic optic neuropathy.5 Single cases of the syndrome have been described in the course of primary pulmonary hypertension or congenital heart defects. Secondary uveal effusion syndrome is treated with furosemide, steroids, haematocrit reduction by elimination of fibrinogen and high-molecular proteins.6

Differential diagnosis of uveal effusion syndrome should take into consideration primary uveal tumours like malignant melanoma, metastases and haemorrhages.7

CASE REPORT

A 36-year-old Caucasian hypermetropic male presented in the eye casualty with a week history of curtain like effect in front of his left eye associated with worsening of vision and flashing lights. Presenting visual acuity was 6/9 and 6/24 in the right and left eye respectively.

Anterior segment of both eyes did not reveal any abnormality and presenting IOP was 18 mmHg and 14 mmHg in right and left eye respectively. Left fundus examination revealed retinal detachment with smooth shaped elevation superonasaly and detached retina inferiorly confirmed on ultrasound B-scan with no tobacco dust in the anterior vitreous (Figures 1 and 2).

Provisional diagnosis of rhegmatogenous retinal detachment with secondary choroidal effusion was made and listed for left eye retinal detachment repair. Pre-operative examination of the left fundus revealed 360 degree choroidal effusion with no evidence of retinal breach or shifting fluid.

Axial length of the right and the left eye was recorded as 19.40 mm and 19.24 mm respectively.

Patient was sent for expert opinion to tertiary referral unit and diagnosis of uveal effusion syndrome was made. Lamellar sclerectomies in the two quadrants were performed and sclera was sent for histology which showed one small focus of acute inflammation near to the edge of the section with no evidence of malignancy.

Over a period of 14 months postoperatively, the left sided eye choroidal effusion gradually settled down, with residual choroidal effusion in the inferonasal quadrant (Figure 3). Eighteen months postoperatively, the retina was found to be completely flat. However, the visual acuity reduced markedly over the course of disease in the left eye to 1/60.

He is under yearly review and has flat retina with extensive scarring and stable visual acuity.
DISCUSSION

The uveal effusion syndrome is a rare disease, which may either affect a normal eye with unaffected or pathologic sclera, or accompany microphthalmos. The sclera may be thickened, but contains no inflammatory cells. The disease usually affects collagen fiber bundles in the sclera with marked accumulations of proteoglycans in the scleral extracellular matrix. Thickening of the sclera is usually associated with hypoplasia or partial absence of the vortex veins.

The sclera becomes less permeable to fluid and colloid. Maintenance of scleral hydrostatic and osmotic pressure gradient is the prerequisite of choroid adhesion to the sclera. The colloid diffusing from choroid vessels to the extracellular space can be largely eliminated by the sclera. The retinal pigmented epithelium is equipped with a mechanism, which pumps aqueous humor only and not colloid, i.e. sub-retinal fluid with high protein content.

Excessive leakage from choroidal capillaries, associated with the insufficiency of fluid-eliminating mechanisms, leads to choroidal and retinal detachment associated with effusion. Stretching of the pigmented epithelium by sub-retinal fluid accumulation weakens the bonds between the epithelial cells, leading to rupture, which, however, is rare. Ciliochoroidal detachment is manifested by intrachoroidal, and especially suprachoroidal, fluid accumulation, visible on the circumference. This phase is usually asymptomatic, corresponding to the sub-clinical stage of uveal effusion syndrome and may be detected by MRI and ICG.

During the further course of the disease, the fluid accumulated in the choroid causes decompensation of retinal pigmented epithelium and inhibits the outflow through the epithelium. Sub-retinal fluid accumulates without intrachoroidal fluid outflow into the sub-retinal space, with progressing effusive retinal detachment.

This stage is characterized by the onset of clinical symptoms in one or both eyes. The natural course is fluctuating, with recurrences. The prognosis is unfavourable in cases of bullous retinal detachment. Visual acuity impairment may persist despite restoring retinal adhesion as a result of treatment. Uveal effusion syndrome is a serious condition that is difficult to treat and can lead to severe and permanent visual loss in both eyes.

The treatment of idiopathic uveal effusion syndrome includes: sclerotomy, sclerectomy, scleral decompression, scleral windows with topical mitomycin and vitrectomy. Early diagnosis, close follow-up, and appropriate management are mandatory to improve or maintain visual function in such patients.

REFERENCES


