



Idiopathic choroidal effusion syndrome : A case report Effusion uvéale idiopathique : A propos d'un cas

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Abstract:

Uveal effusion corresponds to the accumulation of fluid in the supra choroidal space which is a virtual cavity located between the sclera and the choroid.

Idiopathic uveal effusion syndrome is a rare, chronic condition, most commonly in males. Its etiologies are multiple and not always diagnosed, it could rarely be linked to an overload disease (a form of mucopolysaccharidosis). The anterior segment often has no features apart from dilated episcleral vessels. The eyes are often emmetropic, the intraocular pressure is variable. Examination of the posterior segment reveals the characteristic gray choroidal detachment bubbles. Its treatment is based on systemic corticosteroid therapy and cycloplegia. The evolution is often favorable. We report the case of a 67-year-old female patient with idiopathic uveal effusion syndrome revealed by acute ocular hypertonic crisis.

Keywords : ocular hypertonia, uveal effusion, kissing sign

Résumé :

L'effusion uvéale correspond à l'accumulation de liquide dans l'espace supra choroïdien qui est une cavité virtuelle localisée entre la sclère et la choroïde.

Le syndrome d'effusion uvéale idiopathique est une entité rare, évoluant de manière chronique, le plus souvent chez un sujet de sexe masculin. Ses étiologies sont multiples et pas toujours diagnostiquées, elle pourrait rarement être liée à une maladie de surcharge (une forme de mucopolysaccharidose). Le segment antérieur ne présente souvent aucune particularité mise à part des vaisseaux épiscléraux dilatés. Les yeux sont souvent emmétropes, la pression intraoculaire est variable. L'examen du segment postérieur met en évidence les bulles de décollement choroïdien grises caractéristiques. Son traitement repose sur une corticothérapie systémique et une cycloplégie. L'évolution est souvent favorable. Nous rapportons le cas d'une patiente de 67ans qui présente un syndrome d'effusion uvéale idiopathique révélée par crise d'hypertonie oculaire aiguë.

Mots-clés : hypertonie oculaire, effusion uveale, kissing sign.

Introduction

Uveal effusion corresponds to the accumulation of fluid in the supra choroidal space, which is a virtual cavity located between the sclera and the choroid. it is most often due to ocular abnormalities where pressure and inflammatory factors are predominant.[1] This article presents a case of a patient with idiopathic uveal effusion.

Case description

A 67-year-old woman with no medical history, who complained of pain and acute digression in visual acuity in her left eye. the visual acuity after optical correction was 2/10 OS and 9/10 OR. while IOP was 40mmhg OS and 16mmhg OR. On biomicroscopic examination of the left eye, we found episcleral vessels dilation, with a circumcorneal injection. The cornea was the site of diffuse sub epithelial edema, a shallow anterior chamber with flare at 4+. The pupil was in areflexic semi-mydriasis. The lens was the site of a grade 1 nuclear cataract and incipient cortical one, the fundus examination was not accessible (Figure 1). While Examination

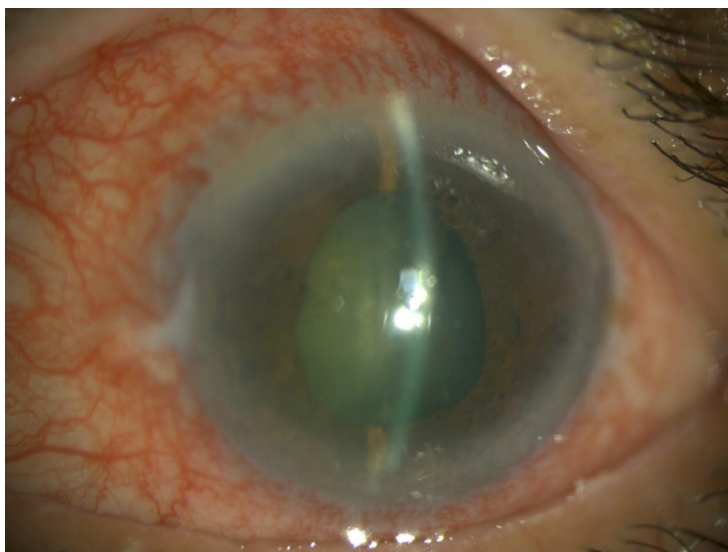


Figure 1: Photo of the left eye slit lamp showing significant inflammation of the anterior chamber as well as cataract

of the right eye found a normal anterior segment including an anterior chamber of normal depth and a gonioscopy showing an open angle on 360°. fundus examination finds a physiological the optic nerve excavation.

In view of these clinical findings, we did an ocular ultrasound which demonstrated a choroidal detachment with a “kissing sign” (figure 2). Optical coherence tomography (OCT) did not show any retinal serous detachment, the macula was normal. Thus, we decided to start a slow intravenous infusion of Mannitol as well as an oral carbonic anhydrase inhibitor.



Figure 2: Image of an ocular ultrasound of the left eye showing choroidal detachment with a «kissing sign» appearance.

Subsequently, we started an etiological investigation of her choroidal detachment: Clinically, there was no notion of trauma or previous surgery. No high blood pressure and no medication. The axial length was 22mm (OSR). Her biological assessment was normal (especially kidney function). The ocular ultrasound showed no visible tumor or scleral thickening. We also performed an orbital MRI which ruled out the possibility of a choroidal mass or posterior scleritis (Figure 3).

Ultimately, the diagnosis was an idiopathic uveal effusion which led us to start full-dose oral corticosteroid therapy as well as cycloplegic eye drops.

The evolution was marked by gradual improvement with regression of symptoms. The inflammation was clearly reduced, the IOP was normalized and her fundus became accessible (physiological excavation). Two months later, her acuity rose to 8/10 and we thus began the decrease of her corticosteroid therapy doses.



Figure 3: Photo of the MRI section showing choroidal detachment in the left eye.

Discussion

An effusion refers to a transudate (fluid only) or exudate (fluid and protein) upon the leakage of a fluid out of the vascular compartment to a nearby tissue or space. In uveal effusion, this fluid leak which comes from the choriocapillary will spread to the outermost vascular layers and finally to the suprachoroidal space which will cause cilio-choroidal detachment.[2]

It is necessary to look for recent myopia following the intake of sulfonamides as well as hyperopia giving rise to suspicion of nanophthalmos. On examination of the anterior segment, microcornea and significant keratometry are suggestive of nanophthalmos. The anterior chamber may be narrow. As for the intraocular pressure, it is variable, which can be high or, on the contrary, lowered in the event of ocular hypotonia. In fundus examination, in the event of a trabeculectomy, there may be minimal peripheral choroidal detachment. A significant uveal effusion should suggest a possible intraocular tumor.[1]

The ocular ultrasound is the first examination to be performed, looking for an internal bulge of the choroidal wall more or less circumferential with steep posterior sclera connection angles. It can also measure the axial length suggesting a closure of the angle or nanophthalmos.[3] Finally, in the event of an unclear fundus, it can find a retinal detachment and detect an associated choroidal tumor. UBM helps analyze the anterior suprachoroid space, studies details of the iris and ciliary body, and measures the anterior sclera.[4] Optical



coherence tomography (OCT) measures the thickness of the choroid and assess associated retinal abnormalities such as retinal serous detachment for example. Fluorescein and indocyanine green angiography are useful in the diagnosis of posterior uveitis and retinal affections.[1]

The etiologies to look for are ocular hypotonia due to retinal detachment, perforating trauma, post-traumatic cyclodialysis, inflammation (posterior scleritis and uveitis), choroidal tumor, vascular causes (malignant hypertension, hypoproteinemia and glomerulonephritis), angle closure, nanophthalmos, and drug causes (PG and Topiramate).[5]

Idiopathic uveal effusion syndrome is a rare and peculiar chronic condition, most often in a male. According to one author, it could be linked to an overload disease (a form of mucopolysaccharidosis)[6]. The anterior segment often has no features apart from dilated episcleral vessels. The eyes are emmetropic, the intraocular pressure is variable (normal or decreased). On examination of the posterior segment, we can find gray choroidal detachment bubbles that can stick together if there are many, as well as a non-rhegmatogenic retinal detachment varying according to the position of the head. When the latter is annular, ora serrata can be visualized.[1] Its treatment is based on systemic corticosteroid therapy and cycloplegia. The evolution is often favorable.[7]

Conclusion

Uveal effusion syndrome is a rare condition. Its etiologies are multiple and not always diagnosed[1]. Its idiopathic entity should be considered a diagnosis of exclusion[2]. Its management must be collegial and specialized.[1]

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CONFLICTS OF INTEREST:

The authors declare that they have no interest in this article.

