SPONTANEOUS PRIMARY UVEAL EFFUSION SYNDROME

SÍNDROME DE EFUSIÓN UVEAL ESPONTÁNEO

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ABSTRACT

Clinical case: This was a 73 year-old male patient who developed an apparent uveal effusion syndrome in his right eye one year after cataract surgery. Once possible associated conditions were discarded, a diagnosis of spontaneous uveal effusion syndrome was reached. With appropriate systemic steroid therapy, a favourable response occurred.

Discussion: The diagnosis of uveal effusion may sometimes be difficult to establish. In order to diagnose and manage these patients, a detailed clinical examination along with fluorescein angiography, ultrasonography, ultrasound biomicroscopy (UBM) and magnetic resonance imaging (MRI) tests, must be carried out (Arch Soc Esp Oftalmol 2007; 82: 505-508).

Key words: Spontaneous uveal effusion, trans-scleral outflow, spontaneous, choroidal detachment, ultrasound biomicroscopy.

RESUMEN

Caso clínico: Presentamos el caso de un paciente varón de 73 años que desarrolló un síndrome de efusión uveal en su ojo derecho un año después de cirugía de catarata. Tras descartar posibles condiciones asociadas, se llegó al diagnóstico de síndrome de efusión uveal espontáneo. Con un tratamiento esteroide sistémico adecuado, el cuadro presentó una respuesta favorable.

Discusión: El diagnóstico de efusión uveal puede ser difícil de establecer. Para el diagnóstico y manejo de estos pacientes, se debe realizar un examen clínico detallado, junto con angiografía fluoresceínica (AFG), ultrasonografía, biomicroscopía ultrasonica (BMU) y resonancia magnética nuclear (RMN).

Palabras clave: Eefusión uveal espontánea, flujo transescleral, desprendimiento coroideo, biomicroscopía ultrasonica.
INTRODUCTION

The spontaneous primary uveal effusion syndrome, described by Schepens and Brockhurst in 1963, is defined as the abnormal accumulation of serous fluid along the outer layers of the ciliary body and choroids and is linked to a chronic and relapsing peripheral annular choroidal detachment (1). In 1982, Gass and Jallow defined the uveal effusion syndrome as the presence of idiopathic serous detachments of choroids, ciliary body and retina, stressing the fact that these may be caused by scleral abnormalities with increased resistance to the transcleral flow of intraocular proteins.

Its variable clinical presentation may lead to an erroneous diagnosis and imitate a choroidal tumor. It begins with a non-symptomatic 360° peripheral ciliary-choroidal detachment resulting from the accumulation of fluid in the choroids. The prevalence of this fluid accumulation results in the decompensation of the pigment epithelium’s pumping device. Fluid then accumulates in the subretinal space, leading in turn to an exudative retinal detachment. Treatment will vary according to the underlying cause, thus the significance of a proper diagnosis.

CASE REPORT

A seventy-three year old male patient arrives in the emergency room reporting pain and redness in the right eye (RE), associated with a decrease in visual acuity.

His personal history included high blood pressure, hypercholesterolemia, hypertensive cardiopathy and chronic renal insufficiency.

Visual acuity was .5 for both eyes. The RE biomicroscopy revealed a good anterior chamber, negative cell tyndall, pseudophakia and dilated episcleral veins (fig. 1), while the left eye was normal (LE). The values for intraocular pressure were normal. The eye fundus exam revealed elevated choroidal injuries in the RE with pseudotumoral appearance in the upper temporal quadrant (fig. 2). Ultrasound A revealed the absence of internal reflectivity and vascularization. Ultrasound B revealed a multi-lobulated choroidal mass without an associated retinal detachment, with the absence of echoes inside, posterior shadow and choroidal cupping (fig. 3). The RE’s axial length was normal. The FAG revealed an increasing hyperfluorescence without any signs of double circulation (fig. 4). The ultrasound biomicroscopy revealed a good anterior chamber, negative cell tyndall, pseudophakia and dilated episcleral veins (fig. 1), while the left eye was normal (LE). The values for intraocular pressure were normal. The eye fundus exam revealed elevated choroidal injuries in the RE with pseudotumoral appearance in the upper temporal quadrant (fig. 2). Ultrasound A revealed the absence of internal reflectivity and vascularization. Ultrasound B revealed a multi-lobulated choroidal mass without an associated retinal detachment, with the absence of echoes inside, posterior shadow and choroidal cupping (fig. 3). The RE’s axial length was normal. The FAG revealed an increasing hyperfluorescence without any signs of double circulation (fig. 4).
croscopy (UBM) revealed a 360° flat choroidal detachment as well as normal scleral thickness measurements. The NMR of the orbits and the cavernous sinus discarded the presence of an arteriovenous fistula.

A systemic treatment was prescribed with steroids at 1 mg/kg/day doses during one week, decreasing by 10 mg per week. Response was favorable during the first week, with a gradual reduction of choroidal detachments. Eight months after the onset of this condition, the RE’s visual acuity was .8 and the eye fundus revealed a decrease in choroidal detachments and the presence of a demarcation line (fig. 5).

**DISCUSSION**

Whenever accompanied by an isolated choroidal detachment, spontaneous uveal effusions pose a challenge in terms of differential diagnosis. The pathologies to be discarded include choroidal tumors, particularly choroidal melanoma (2), not to forget peripheral exudative degeneration, choroidal hemangioma, posterior scleritis, metastatic carcinomas, choroidal osteoma and degenerative retinoschisis among others.

To make an appropriate diagnosis, it is necessary to perform a full ophthalmologic exploration and ultrasound (A and B). Other complementary tests may include FAG, UBM and NMR or scan (3).

The ultrasound revealed no melanoma signs, such as posterior shadows, choroidal cupping or internal vascularization. The low internal reflectivity excluded hemangioma, osteoma and choroidal metastasis, as well as the «T sign» characteristic of posterior scleritis and scleral thickening.

Subsequently, the ultrasound suggested a localized choroidal detachment diagnosis (4). The FAG does not aid in making diagnosis unless it takes into account signs such as the double circulation characteristic of choroidal melanomas.

At this point, the most probable diagnosis was a sinus choroidal detachment resulting from a spontaneous primary uveal effusion syndrome. This finding was confirmed by the UBM, once other triggering processes had been discarded, for instance ocular hypotony, arteriovenous fistula, inflammatory or neoplastic processes or nanophthalmos.

The UBM allows for more accurate measurements of scleral thickness than the NMR (5) and in the event of uveal effusions it may suggest the type of treatment to follow.

The spontaneous primary uveal effusion syndrome has been typically associated with thickened sclerae that alter the transcleral flow and result in accumulated fluid in the suprachoroidal space. It is divided into 3 groups: Type 1 (nanophthalmic eyes) and type 2 (non-nanophthalmic) exhibit a thickened sclera due to excess of glycosaminoglycans; whereas type 3 appears in non-nanophthalmic eyes and exhibits no scleral alterations. In the present case, the multi-lobulated choroidal detachment was confirmed by the UBM as a 360° flat choroidal detachment without the associated retinal detachment and normal sclera. These findings suggest the initial
stages of a spontaneous primary uveal effusion syndrome.

Although this syndrome has been typically reported among healthy adult males, this condition has been described in individuals between 20 and 80 years of age and both genders. Bilateral, generally non-symptomatic, except in those chronic cases where the retinal pigment epithelium’s pumping device (RPE) is involved, this condition results in exudative retinal detachments and the subsequent visual alterations. Chronic cases may exhibit RPE alterations in the eye fundus and FAG (unspecified pattern).

Treatment varies depending on the type of syndrome. Sclerectomy is recommended mainly in those cases associated with altered sclera. Evolution is usually extended by remissions and exacerbations. The patient’s positive response to corticoids, unusual for this syndrome, may be explained by the beneficial action of this medication on the choroidal inflammation with vessel dilatation.

REFERENCES