Short communication

Uveal effusion associated with an IgM lambda subtype monoclonal gammapathy

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ABSTRACT

Case report: A 68-year-old man was referred to the hospital with progressive decreased vision in the right eye over the past year. A moderate cataract and annular choroidal thickening were found. The diagnosis of uveal effusion was confirmed by ultrasound and fluorescein and indocyanine green angiography. Laboratory studies showed an IgM lambda subtype monoclonal gammapathy of undetermined significance. The patient underwent cataract surgery, and a sub-Tenon’s triamcinolone injection with a satisfactory short-term outcome.

Conclusion: This association has not been previously reported, and it shows that IgM lambda subtype monoclonal gammapathy of undetermined significance should be added to the list of disorders associated with uveal effusion.

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EFUSIÓN UVEAL ASOCIADA A GAMMAPATÍA MONOCLONAL DE SIGNIFICADO INDETERMINADO TIPO IG M LAMBDA

RESUMEN

Caso clínico: Varón de 68 años que consulta por disminución de agudeza visual progresiva en ojo derecho en el último año. En la exploración destaca una catarata moderada y un engrosamiento anular coroideo. Mediante ecografía ultrasónica, angiografía fluoresceínica y verde de indocianina, se confirma el diagnóstico de efusión uveal. El estudio hematológico diagnostica una gammapatía monoclonal de significado indeterminado tipo IgM lambda. Se decide tratamiento quirúrgico de la catarata e inyección subtenoniana de triamcinolona, consiguiendo un resultado visual muy satisfactorio a corto plazo.

Conclusion: Esta asociación no ha sido descrita previamente en la literatura, por lo que la gammapatía monoclonal de significado indeterminado IgM lambda podría incluirse en la lista de enfermedades asociadas a efusión uveal.

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Clinic case

A patient, aged 68, visited owing to progressive VA reduction in RE with one year evolution. Relevant personal history includes controlled AHT.

Corrected VA of 0.5 in RE and 1.0 in LE. IOP was of 22 and 17 mmHg respectively. The only finding in biomicroscopy was moderate corticonuclear cataract in RE, the fundus of which exhibited serous retina detachment associated to diffuse annular thickening of the choroids (Fig. 1). Left eye fundus was normal (Fig. 2).

Ecograph (Ultrascan Imaging System, Alcon) evidenced choroidal thickening and hyper-ecogenic membrane adhered to the optic nerve, suggesting retina detachment with subretinal liquid (Fig. 3). RE fluorescein angiography (Trc 50-ix, Topcon) revealed multiple choroidal thickening without loss of contrast, peripapillary hyperfluorescence points and temporal juxtamacular serous elevation (Fig. 4). Indocyanine green angiography showed perivascular hyperfluorescence in superior and inferior temporal arches (Fig. 5). RE macular OCT (Cirrus HD-OCT, Zeiss) confirmed said subretinal elevation (Fig. 6).

After approaching the case as uveal effusion syndrome and with suspected primary tumor versus metastasis, a methodological study was carried out to reach the diagnostic of IgM lambda-type monoclonal gammopathy of undetermined significance (MGIS).

The patient was intervened for RE cataract and reached a corrected VA of 0.7. One month after the operation, a subtenon triamcinolone injection was administered and the patient reached a VA of 1.0 after 2 months. Due to satisfactory vision, additional invasive treatments at the ocular level were discarded and evolution controls were decided.

Discussion

Uveal effusion is caused by alterations in the sclera which produce obstruction of the vorticose veins with the ensuing choroidal congestion and difficulty in ocular venous return. This involves the accumulation of sub-choroidal liquid and annular cilia-choroidal detachment which, in the long-term, will cause imbalances in the retina pigment epithelium and deteriorate its pumping mechanism, and may be associated to exudative retina detachment.

Even though the most frequent cause of uveal effusion is idiopathic, it is important to discard other etiologies due to the clinical impact these would involve for
the patient. Differential diagnostic includes iatrogenic causes (post-surgery, panphotocoagulation, ocular hypotony), inflammatory causes (posterior scleritis, orbital pseudotumor), vascular causes (carotid-cavernous fistula), tumors (choroidal melanoma, choroidal hemangioma, choroidal osteoma and metastasis)\(^1\) and hematological causes (lymphoma, multiple myeloma\(^2\)).

Imaging tests will assist in discarding said etiologies. Ocular echography evidenced choroidal and retinal detachment and discard the characteristic “T-sign” of posterior scleritis, without showing the typical posterior shadow of melanoma on the internal hyperecogenicity of osteoma or metastasis\(^2,5\). Fluorescein angiography revealed signs of RPE imbalance with the typical “leopard spots” appearance.\(^2\) Indocyanine green angiography assisted in the differential diagnostic with choroidal hemangioma.\(^6\) Nuclear magnetic resonance of orbits and cavernous sinuses served to discard arteriovenous fistula\(^1\) and solid ocular tumors, while ultrason sound biomicroscopy was a precise method for measuring scleral thickness.

All the said ecographic and angiographic signs were found in the patient reported herein. In addition, optical coherence tomography contributed to the diagnostic confirmation.

The clinic course is usually prolonged with remissions and exacerbations, with poor response to medical treatment.\(^6\) If the cause is inflammatory, one option is the use of systemic corticoids although results are not generally very satisfactory.\(^1\) Subtenon triamcinolone injections have been tried recently and exhibited prolonged effect.\(^7\) This technique has been applied in the present case with successful results in the short term.

In what concerns surgical treatment, scleral resection techniques around vorticose veins have been described, although these are difficult to perform and very risky.\(^8\) Accordingly, the most widely applied technique is superficial sclerotomy followed by deep sclerectomy, a simple technique with very good results.\(^2\)

MGIS is characterized by the presence of genetically identical cells which produce immunoglobulin detected in blood in the form of a monoclonal strip.\(^9\) MGIS is the monoclonal gam mopathy which most frequently presents said strip, followed by multiple myeloma, which is an entity broadly described in the literature in association to uveal effusion.

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**Fig. 4** – Right eye fluorescein angiography: choroidal hyperfluorescence in patches and peripapillary hyperfluorescence dots, both in early and late angiogram times.

**Fig. 5** – Right eye indocyanine green angiography: hyperfluorescence in posterior pole, perivascular in superior and inferior arches and in nasal retina.
The prevalence is approximately 3.2% at age 50 and higher in patients over 70. Patients are typically asymptomatic and diagnosed casually in the course of routine examinations.

Prognosis and evolution are uncertain because 20–30% of MGIS can evolve to malign diseases such as multiple myeloma, macroglobulinemia, amyloidosis or lymphoproliferative syndromes.

Due to the absence of symptoms and clinically significant signs, no treatment is required other than close clinical follow-up.

The association of multiple myeloma with uveal effusion and other ocular expressions is well-known. However, the present clinic case could be the first case described in the literature of uveal effusion associated to MGIS, a benign expression but a potential precursor of multiple myeloma.

Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES


