DIFFUSE UVEAL MELANOMA. A CLINICAL CASE

MELANOMA UVEAL DIFUSO. A PROPÓSITO DE UN CASO

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ABSTRACT

Case report: We report the case of a 58-year-old man who presented at our emergency department with ocular pain and progressive loss of vision in his left eye. Ophthalmic examination detected a pigmented mass in the iridocorneal angle, involving the ciliary body. Glaucoma had also developed secondary to the melanotic lesion. Ophthalmoscopy showed a large pigmented choroidal tumor. After enucleation, pathologic examination confirmed the diagnosis of a diffuse ocular melanoma.

Discussion: Diffuse uveal melanoma is a malignant tumor that metastasises early. Enucleation is the treatment of choice (*Arch Soc Esp Ophthalmol* 2006; 81: 545-548).

Key words: Uveal melanoma, ciliary body, melanotic lesion, choroidal tumor, enucleation.

RESUMEN

Caso clínico: Se presenta el caso de un varón de 58 años que acudió a urgencias con dolor ocular y pérdida progresiva de visión del ojo izquierdo. La exploración oftalmológica detectó una masa pigmentada en el ángulo iridocorneal afectando cuerpo ciliar. El paciente presentaba glaucoma secundario a la lesión melánica. La oftalmoscopia mostró una tumoración coroidea pigmentada de gran tamaño. Tras la enucleación ocular, el estudio anatomopatológico confirmó el diagnóstico de melanoma ocular difuso.

Discusión: El melanoma uveal difuso es un tumor maligno que produce metástasis en los primeros años tras su diagnóstico, siendo la enucleación el tratamiento indicado.

Palabras clave: Melanoma uveal, cuerpo ciliar, lesión melánica, tumor coroideo, enucleación.

INTRODUCTION

Uveal melanoma constitutes the most frequent primary malign intraocular tumor in adults. Its prevalence ranges between 6-7 cases per million inhabitants/year (1). Diffuse uveal melanoma accounts for 4-6% of all uvea melanomae and it grows infil-

trating and thickening the choroid in a diffused manner with hardly any protrusion (2). The edges are irregular with tongue-shaped extensions, giving it the appearance of a geographic chart. Its extension is greater than the nodular melanoma and it may comprise the entirety of the choroid, with Bruch's membrane remaining intact (2).

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Approximately 59% of patients may exhibit glaucoma and inflammatory symptoms due to areas of necrosis, with the mortality rate rising to 73%, exceeding that of the nodular melanoma (2). The extraocular extension is more frequent than in the circumscribed melanoma (it appears in 53% of cases) and is present in some patients at enucleation (2). The differential diagnosis of this melanoma can be complicated due to its non-typical clinical presentation, with the main doubts being choroidal metastasis and the “Bilateral melanocytic uveal proliferation associated to systemic malignancy” (2,3).

CASE REPORT

We present the case of a 58-year old man who attended the urgency ward due to intense ocular pain with progressive loss of vision in his left eye. The ophthalmological exploration revealed a pigmented mass in the iridocorneal angle which protruded to the anterior chamber, involving the iris (fig. 1). The clinical assessment of the patient was compatible with acute glaucoma secondary to said mass in the anterior chamber. Ophthalmoscopy revealed a large irregular pigmented mass with plateaus (figs. 2, 3). The characteristics revealed by the eye echography were compatible with a large ocular melanoma with diffused extension (fig. 4). The Magnetic Nuclear Resonance (MNR) revealed a 17-mm mass discretely hypertense in T1 and hypoten-

Fig. 1: Presence of a pigmented tumoration in the superior temporal quadrant of the anterior chamber.

Fig. 2: Eye fundus image showing the diffuse extension of the melanoma and the presence of an associated retina detachment extending to the macular area.

se in T2, compatible with the diagnostic of choroidal melanoma (1).

The Oncology service carried out an extension study which discarded metastatic dissemination. Due to the extension of the tumor, the patient symptoms and the absence of any possibility of visual recovery, enucleation treatment was considered (1,3).

After extraction, the macroscopic assessment of the ocular globe evidenced the presence of

Fig. 3: Eye fundus showing a large pigmented tumoration occupying a large interior area of the ocular globe.

Fig. 4: Echographic vision of the tumor showing its height.

scleral invasion. The patient evolved positively, maintaining ocular motility and a satisfactory appearance with the external eye prostheses as well as a good quality of life, with the only drawback being the difficulty to perform daily tasks which required good binocular vision. The macroscopic assessment of serialised sections of the ocular globe revealed an endophytic growth circumferential mass comprising virtually the entire choroid, extending up to the ciliar body and the iris (fig. 5), histologically corresponding to a proliferation of fusiform and epithelioid cells

Fig. 5: Macroscopic section of the ocular globe evidencing the extension of the tumor throughout the choroid circumference, reaching the ciliar body and the iridocorneal angle, protruding into the anterior chamber.

with a cytoplasm that was large, pigmented with oval-shaped nuclei with prominent nucleolus, all significantly atypical.

The number of mitosis was high, evidencing hemorrhage and necrotic areas. These anatomic and pathological data confirmed the diffuse uveal melanoma diagnostic, with perforation of the scleral wall reaching the external surface thereof.

DISCUSSION

The diffuse ocular melanoma tumor is more aggressive and difficult to prognosticate than the circumscribed melanoma (2). Its greater extension, together with the involvement of the ciliar body, determine the negative prognosis of this tumor (1). It is frequent to find scleral invasion at diagnostic, as could be observed in our patient. The melanoma exhibits metastatic dissemination in the first five years after being diagnosed. The factors which enhance this possibility include the tumoral size, the involvement of the ciliar body and the scleral invasion among others (1-3). Our patient comprised all said factors and therefore the treatment of choice was enucleation vis-à-vis conservative treatments for the eye globe such as brachitherapy, proton beam or a combination thereof with transpupillary thermal therapy (1,3). The latest studies supplied by the Collaborative Ocular Melanoma Study Group (COMS), published in the COMS report n.º 10 (4) as well as in the COMS report n.º 18 (5), concluded with similar survival rates between the patients treated with enucleation and those treated with brachitherapy with I^{125} , contradicting the theories proposed by Zimmerman (4).

In what concerns quality of life, an increasingly important concept for Medicine, the latest studies comparing quality of life in patients treated for choroidal melanoma concluded that it is similar for both treatments (brachitherapy and enucleation) (1).

The histopathological study definitively confirmed the diagnostic suspicion. In addition to the diffuse growth and the scleral extension, the high number of mitosis and the aspect of the nucleolus (considered to be important prognostic factors) confirmed the negative prognosis of the tumor affecting our patient (2).

In less than a year after diagnostic, the patient suffered hepatic metastasis which led to an important and progressive deterioration of his health up to his demise.

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