Short communication

Giant optic disk melanocytoma complicated with massive intraocular seeding

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ABSTRACT

Case report: A 33-year-old female with a large, unilateral, dark tumor of the optic disk with a presumed diagnosis of melanocytoma. After a follow-up of nineteen years, the tumor produced massive intraocular seeding with cataract, secondary glaucoma (pigmentary, melanocytomalitic, inflammatory and pupillary seclusion glaucoma) and amaurosis. The eye is enucleated and the pathological examination confirmed the diagnosis.

Discussion: Optic disk melanocytoma is a benign stationary tumor. However, sometimes there are complications, including visual loss. Our case confirms that vitreous seeding is due to tumor necrosis with dissemination of tumoral debris and melanin, which are phagocytized by macrophages and produce an inflammatory reaction, trabecular plugging and ocular hypertension.

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Melanocitoma gigante de la papila óptica complicado con siembra intraocular masiva

RESUMEN

Caso clínico: Mujer de 33 años con gran tumouración oscura sobre papila óptica unilateral diagnosticada presuntamente como melanocitoma. Tras seguimiento de 19 años, la tumouración produce siembra intraocular masiva con catarata, glaucoma secundario (pigmentario, melanocytomalítico, inflamatorio y seclusión pupilar) y amaurosis. Tras enucleación se confirma anatomopatológicamente el diagnóstico.

Discusión: El melanocitoma de papila óptica es un tumor benigno estacionario, pero raras veces ocasiona complicaciones incluida la pérdida visual. Nuestro caso corrobora que la siembra vitrea está causada por necrosis tumoral con disseminación de detritos tumurales y melanina, que fagocitados por macrófagos, producen reacción inflamatoria, ocupación trabecular e hipertensión ocular.

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Introduction

Melanocytoma is considered to be a type of melanocytic nevus. It is usually located in the optic disk although it can also be found in any other part of the uvea. Clinically, it is characterized by exhibiting a dark brown or black color. At the histological level, it comprises by intensely pigmented round and oval cells with abundant cytoplasm and small, rounded and uniform nuclei.\(^1\)

The term optic disk melanocytoma was coined by Zimmerman in 1962.\(^2\) Cogan\(^3\) defined it as optic nerve magnocellular nevus. Shields et al.\(^1\) have proposed the denomination of optic nerve hyper pigmented magnocellular nevus because they consider this definition more in accordance with the nature of the tumor.

Clinical case

A Hispanic female, aged 33, without relevant history had complained of progressive vision reduction in the left eye (LE). The exploration revealed a visual acuity (VA) of 0.05 in LE. The ocular fundus of said eye showed an intensely pigmented polypoid raised papillary mass as well as rows of mobile pigment in the vitreous cavity (Fig. 1). The rest of the ophthalmological exploration gave normal results.

An echography revealed a solid lesion. The A mode revealed high internal reflectiveness without spontaneous vascular movement. At that time, several experts were consulted; some expressed their preference for a malign melanoma diagnostic while others preferred melanocytoma. The decision was taken to maintain the patient in observation. Throughout 19 years, the lesion progressively increased in size without rapid changes in its configuration and accordingly the melanocytoma diagnostic was proved to be appropriate. The patient lost vision in the LE and funduscopy was no longer possible, initially due to the vitreous pigment and later due to cataracts.

Nineteen years after the first visit, the patient began to complain of intense pain due to acute glaucoma with inflammation in the anterior chamber and pupilar seclusion without rubeosis. For this reason enucleation was decided. The anatomo-pathologic ocular globe assessment (Figs. 2–4) revealed that the chamber angle was open and contained abundant macrophages full of pigment which infiltrated the trabecular mesh, surrounding Schlemm’s canal, covering the anterior iris surface and appearing over the lens posterior capsule between the ciliary processes and in the anterior vitreous. The lens was affected by cataracts. An intensely pigmented tumor was observed emerging from the optic nerve head and extending along the juxta-papillary retina, made up by large polyhedron-shaped cells with abundant cytoplasm and small picnotic nuclei. The majority of these cells were anucleolated...
Fig. 4 – Cross-section of the enucleated eye showing the dark brown globular mass emerging from the optic papilla and invading the optic nerve, including the posterior line of the surgical excision. Pigmentary dispersion in the anterior vitreous and lens anterior and posterior capsule.

even though a few had small nucleoli (Fig. 5). The tumor exhibited surface necrotic spots, with dotted pigmented macrophages over the internal limiting membrane and in the vitreous. Intensely pigmented spindle cells could be seen in some areas. In many areas, the pigmented cells extended along the internal limiting membrane of the retina reaching the ora serrata. Stained preparations of the tumor showed benign cytologic characteristics typical of melanocytoma cells (Fig. 6). The tumor did not reveal mitotic activity and malign melanoma or extraocular extension evidences could not be observed.

Discussion

The majority of melanocytomas do not produce important visual deterioration. However, slight loss of vision arises in about 26% of the cases, generally due to foveal exudative retinal detachment or neuroretinitis caused by tumor necrosis. Severe visual loss may arise only rarely, secondary to central retinal vein occlusion, spontaneous tumor necrosis or malignant transformation.

In 1987, Yamaguchi et al. described the first case of an optic disk melanocytoma causing pigmentary dispersion in the vitreous and 15 years later in the anterior chamber. Until then, this was attributed exclusively to malign melanoma. These authors postulated that the posture necrosis of the tumor accounts for the release and deposition of melanine granules.

In 1989, Mansour et al. described a case similar to the present one in which an ocular globe was enucleated after exhibiting exaggerated growth, observing that next to the typical melanocytoma settings melanophages and small spindle cells appeared with irregular, hyperchromatic and vesicular nuclei, similar to A spindle cells of iris composite melanoma. The said authors describe infiltration of tumor cells in the papillary area, of the internal limiting membrane, of the retina vessels and of the large vessels of the optic nerve head. As described above, in this case we also found spindle cells in the tumor and tumoral cells over the internal limiting membrane around the papilla.

Traditionally, melanocytoma has been considered as a stable tumor without a tendency to grow. However, the follow-up of many cases has documented that 10–15% exhibit slow long-term growth. This growth need not be considered as a sign of malignant transformation.

In the present case, slow growth has occurred throughout the period of at least 19 years which has led to tumor necrosis with massive dispersion of tumor material in the vitreous and anterior chamber, to the point of causing amaurosis and pigmentary and inflammation glaucoma due to pupil seclusion.

Fig. 5 – The histopathology of the pigmented optic nerve tumoration exhibits abundant large cells with polyhedron-shaped central or paracentral small nuclei (lighter areas) and abundant melanine pigment. Three cystoid spaces can be appreciated with melanine-filled macrophages (hematoxiline–eosine, 40x).

Fig. 6 – Decolored preparation of the tumoration showing rounded and polyhedron-shaped cells with small central or paracentral nuclei and low nucleocytoplasmatic quotient. In some areas, aggregates of more densely pigmented cells can be seen dispersed within the tumor (40x).
Conflict of interests

None of the authors have declared any conflict of interests.

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