

# GIANT NODULAR POSTERIOR SCLERITIS COMPATIBLE WITH OCULAR SARCOIDOSIS SIMULATING CHOROIDAL MELANOMA

## ESCLERITIS NODULAR POSTERIOR GIGANTE COMPATIBLE CON SARCOIDOSIS OCULAR SIMULANDO UN MELANOMA DE LA COROIDES

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### ABSTRACT

**Case report:** A 30-year-old man was referred to our ocular oncology service with a diagnosis of amelanotic choroidal melanoma of the left eye. The following tests were performed: ophthalmoscopy, fluorescein angiography, indocyanine green angiography, ultrasonography, magnetic resonance imaging and biopsy.

**Discussion:** The diagnosis of giant nodular posterior scleritis, as suggested by ultrasonography, was confirmed by biopsy. A comprehensive medical evaluation was performed, but no etiology was found. The histology revealed a granuloma compatible with ocular sarcoidosis. A rapid response was obtained by systemic steroid administration (1 mg/kg). Sarcoidosis continues to be a challenge in diagnosis. It is important to distinguish nodular posterior scleritis from choroidal melanoma (*Arch Soc Esp Ophthalmol* 2007; 82: 563-566).

**Key words:** Scleritis, eye neoplasms, uveal neoplasms, choroid neoplasms, sarcoidosis, choroidal melanoma.

### RESUMEN

**Caso clínico:** Un paciente de 30 años fue enviado a nuestro servicio oncológico de referencia con el diagnóstico de melanoma amelanótico de la coroides del ojo izquierdo. Se hicieron los siguientes exámenes: oftalmoscopia, angiografía fluoresceínica, angiografía con verde de indocianina, ecografía, resonancia magnética y biopsia.

**Discusión:** El diagnóstico de escleritis nodular posterior gigante basado en la ecografía se confirmó mediante una biopsia. Se realizó un examen médico completo buscándose el posible agente causal; no se encontró la etiología. La histología reveló un granuloma que era compatible con una sarcoidosis ocular. Mediante la administración de esteroides por vía oral (1 mg/kg), se obtuvo una evolución rápidamente favorable del cuadro. El diagnóstico de esta afección sigue siendo un desafío. Es importante diferenciar una escleritis posterior nodular de un melanoma de la coroides.

**Palabras clave:** Escleritis, tumores del ojo, tumores de la úvea, tumores de la coroides, sarcoidosis, melanoma de la coroides.

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## INTRODUCTION

Giant nodular posterior scleritis is a severe inflammatory pathology involving the posterior sclera and may cause a choroidal detachment up to the ora serrata. This is the case in 2 to 12 percent of scleritis cases. It affects mainly females (66-83 percent). It may appear as a granulomatous disease, but in most cases it is idiopathic and unilateral (1). Due to its shape and volume, it may be mistaken for a uveal tumor (2,3). The case described herein was referred to the ophthalmology department as a choroidal melanoma.

## CASE REPORT

In November 2005, a 30-year-old Caucasian male, without relevant medical history, was referred to the ophthalmology unit reporting a choroidal melanoma in his left eye (LE).

Since August 2005, the patient had noticed in his RE a unilateral reduction in visual acuity (VA). He had been complaining for a month of ocular pain in the said eye.

The right eye's (RE) VA was 10/10 with  $-0.5$  D spherical correction, while the LE was  $<1/10$  without correction and 8/10 with an added  $-5.00$  D spherical correction.

The RE ophthalmologic examination showed no relevant signs.

The slit lamp examination of the LE revealed a marked ciliar injection, a slight inflammatory reaction in the anterior chamber and a convex iris; ocular pressure (OP) was 17 mmHg. The LE fundus revealed the presence of a circumscribed amelanotic mass in the nasal sector (fig. 1A). Ultrasound B showed thickening of the eye wall exceeding 5 mm. In the extrascleral space, there was a hypoechogenic area (fig. 1B). The fluorescein angiography and the Indocyanine green revealed that the injury was surrounded by well-visualized choroidal flaps; no intratumoral vascular network was appreciated (fig. 2).

An orbital-cerebral nuclear magnetic resonance (NMR) confirmed the scleral thickening of nodular appearance capturing the contrast in the posterior-internal area of the LE, but revealed no other injuries (fig. 3).

A detailed study in search of a rheumatic etiology (rheumatoid factor, antinuclear antibodies, cANCA, antiphospholipid antibodies) and an infectious etio-

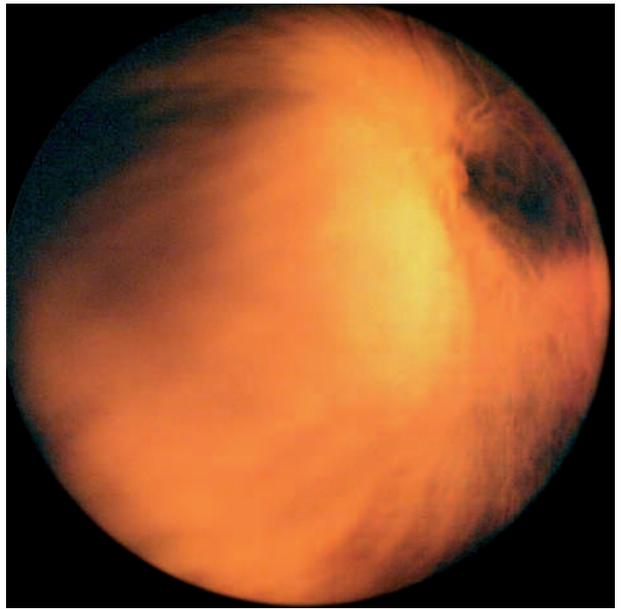


Fig. 1A: LE fundus. Presence of a circumscribed amelanotic mass in the nasal sector.

logy (TPHA, toxocariasis) was negative. Levels of calcium in the blood were normal: 2.39 mmol/l. Chest X-rays were normal. The angiotensin-converting enzyme was slightly increased: 73U/L (normal, 10-55), being compatible with a granulomatous

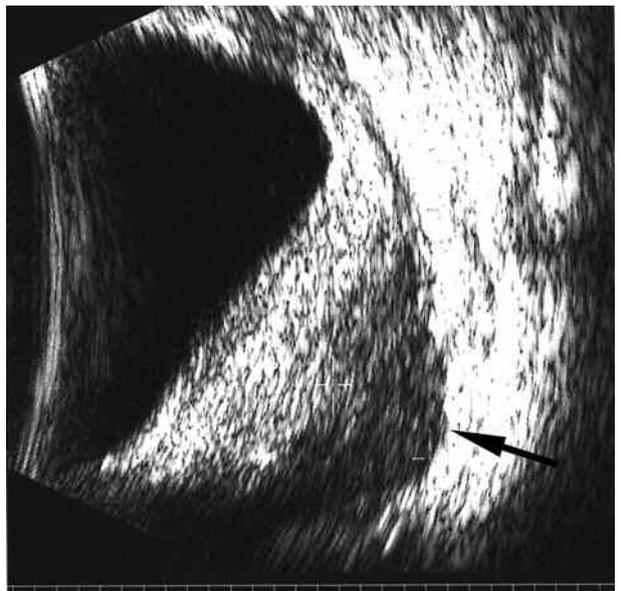


Fig. 1B: Ultrasound B. Retinochoroidal and scleral thickening exceeding 5 mm. In the extrascleral space (arrow), there is a hypoechogenic area corresponding to the inflammation of the subretinal space.

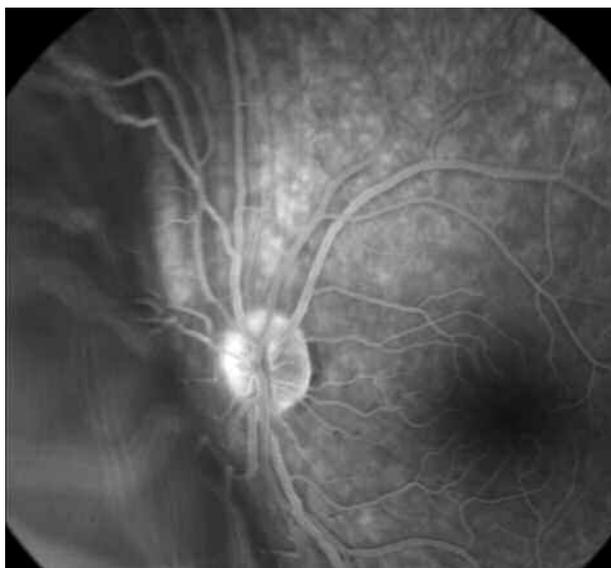


Fig. 2: Fluorescein angiography (A) and Indocyanine green (B). Presence of choroidal flaps along the mass contour. No intratumoral vascular network could be visualized.

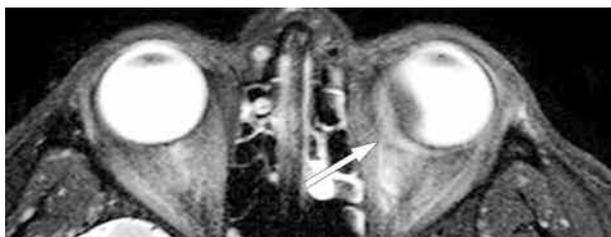


Fig. 3: NMR. Axial T2-weighted image with fat suppression. The thickening of the ocular wall is hyperintense with respect to the vitreous; the hyperintense extra-scleral layer corresponds to the subretinal fluid (arrow).

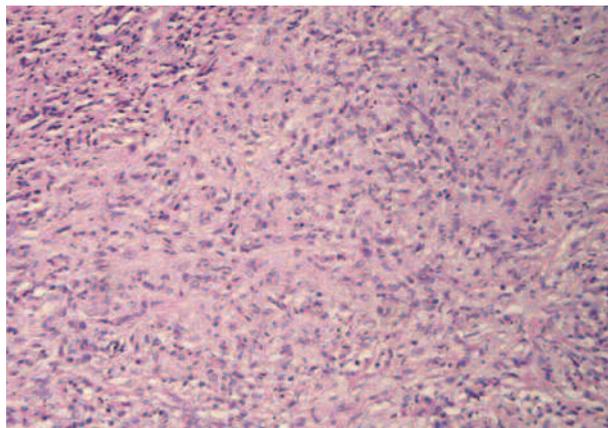


Fig. 4: Pathologic anatomy. Hematoxylin-eosine, x200. Granuloma made of epithelioid cells, surrounded by some lymphocytes.

disorder, while the remaining examinations did not provide arguments in favor of either sarcoidosis or tuberculosis.

Taking into account the anamnesis, clinical examination, angiography, ultrasound and NMR, the patient was diagnosed with giant posterior scleritis. The extra-scleral biopsy and the results of the histopathologic examination confirmed the presence of a granulomatous scleritis compatible with a sarcoidosis (absence of pathogenic agents visible with Ziehl-Neelsen stain, Gram, methenamine silver and PAS stain) (fig. 4).

The patient was treated with oral corticoids at 1 mg/kg/day doses, gradually reducing these doses due to the good therapeutic response down to 0.5 mg/kg/day after six months. On top of this, a local pattern was added in the LE with 1 drop 3 times a day

of 0.1% dexamethasone, aimed at treating the inflammation in the anterior chamber, and 1 drop 3 times a day of 1% atropine to reduce the ciliary spasm.

Check-up of the LE six months later showed 10/10 VA with +0.50 D spheric correction. The ciliary injection had disappeared, together with the inflammatory reaction in the anterior chamber, while the lens was transparent; OP was 15 mmHg. In the eye fundus, the marked elevation of the nasal sector flattened (fig. 5A). Ultrasound B revealed that total thickness of the ocular wall in the inferior nasal sector measured approximately 2 mm (fig. 5B).

## DISCUSSION

This case illustrates the challenging differential diagnosis posed by choroidal melanoma and giant posterior nodular scleritis.

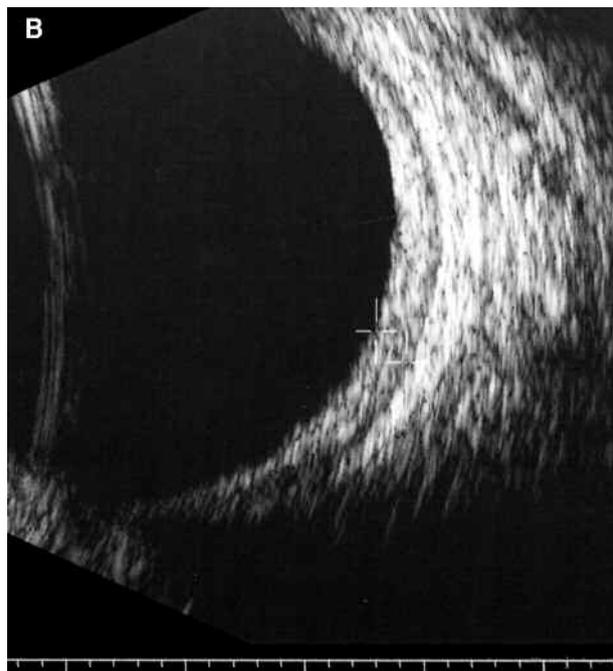
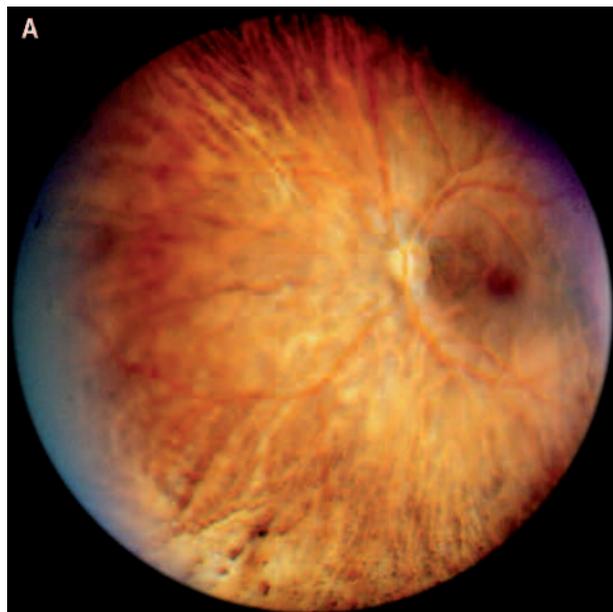


Fig. 5: After six months of corticoid treatment. A: LE fundus. Disappearance of the mass in the nasal sector. B: Ultrasound B. Reduction of injury's thickness down to approximately 2 mm.

In the available literature from 1956 to 1973, different studies included statistics regarding histopathologic diagnoses of enucleated eye globes, the result of mistakenly diagnosing an uveal melanoma; the percentage of uveitis or posterior sclerosis ranges from 0.5 to 6.6 percent. There is a decrease in diagnostic errors since the 1970s with the coming of modern diagnostic means (4).

Several factors led us to diagnose this male patient with posterior scleritis. Both the ocular pain and ciliary injection were compatible with an inflammatory pathology responsible for the ciliary spasm and temporary myopia. The significant amelanotic mass in the eye fundus associated with choroidal flaps without its own vascular network seemed to suggest the presence of a posterior scleritis. The ultrasound revealed the thickening of the ocular wall associated with the presence of extra-scleral fluid corresponding to the inflammation of the subretinal space; this, in turn, reinforced our hypothesis. The NRM aided in discarding an eventual expansion process, but did not allow for the exclusion of a lymphoma. These elements were the basis for performing a scleral biopsy that confirmed the diagnosed giant posterior nodular scleritis compatible with sarcoidosis. In ocular sarcoidosis, barely 25 to 50 percent present systematic involvement (5). Typically, corticoid treatments have an excellent response.

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