Short communication

Retinal detachment associated with Morning Glory syndrome

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\textbf{Article Info} & \textbf{Abstract} \\
Article history: & Case report: A twenty-three-year-old woman was diagnosed of a morning glory papillary anomaly, then with normal visual acuity (VA). Nine years later, the VA decreased to 0.4, secondary to a serous macular detachment, confirmed by optical coherence tomography (OCT). After treatment with C2F6 gas injection, positioning, and peripapillary laser, the VA improved to 0.7 and the foveolar area reattached.
Received 10 December 2010 & Discussion: The Morning Glory syndrome usually has an early diagnosis due to poor visual acuity. Thirty-eight percent of the cases have retinal detachment. We show an unusual case of Morning Glory syndrome with a serous detachment, successfully treated with gas and laser.
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\textbf{Resumen}

Caso clínico: Mujer de 23 años con anomalía papilar de morning glory con agudeza visual (AV) de 1. Nueve años después presenta disminución de AV (0,4) por desprendimiento seroso macular confirmado por tomografía de coherencia óptica (OCT). Tratado con inyección de gas intraocular C2F6, posicionamiento y láser, conseguimos la desaparición del líquido subretiniano y una AV final de 0,7.

Discusión: El síndrome de morning glory suele diagnosticarse precozmente debido a la mala AV. Un 38% de los casos presentan desprendimiento de retina. Mostramos un caso inusual de síndrome de morning glory con desprendimiento seroso tratado con éxito mediante gas y láser.

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Introduction

The Morning Glory syndrome is a generally unilateral congenital dysplasia of the optic nerve, characterized by a bell-shaped excavation, central fibroglyal tissue and radial retinal vessels reminiscent of the morning glory flower.\(^1\) Up to 38% of cases exhibit retinal detachment, the origin of which is controversial. It is believed that the subretinal liquid could be due to complications with the vitreous humor or with the subarachnoid space. Diagnostic is usually early due to anisometropia, poor visual acuity (VA) and concomitant strabismus.\(^2\)

This paper presents an unusual case of Morning Glory syndrome with macular serous detachment and a 10-year evolution.

Clinic case

Female, age 23, with a personal history of epilepsy who visited the practice due to astenopia, exhibiting a corrected visual acuity of 1 in both eyes with slight anisometropy. The ocular fundus revealed peripapillary atrophy with increased excavation, with vessels radiating outwards surrounded by fibroglyal tissue, compatible with Morning Glory papillary anomaly in LE (Fig. 1). The RE posterior pole was normal. The visual field (VF) exhibited an increased blind spot in the LE (Fig. 1). The evoked potentials were normal, although the LE latencies were at the higher limits of normal ranges.

Magnetic resonance revealed agenesia of the callous body and communication of the frontal handles due to the absence of the interventricular septum in addition to a dermoid cyst at the base of the frontal lobes with signs of rupture to the subarachnoid space and the ventricular system and hydrocephalus (Fig. 2).

Nine years after the diagnostic, the patient exhibited diminished VA in LE (0.4) due to serous macular detachment. Optic coherence tomography (OCT) confirmed the existence of subretinal liquid with macular retinoschisis, without detecting the presence of tears in the neurosensory retina (Fig. 3). Angiography showed a peripapillary serous detachment with foveolar involvement (Fig. 4). After three months of observation without resolution, an intravitreal injection of gas C2F6 was carried out in prone position and subsequent peripapillary laser photocoagulation after retina planation. Progressively we observed a reduction in the subretinal liquid and displacement of the central macular area with a progressive VA improvement. One year later, a follow-up with OCT confirmed the disappearance of the foveal subretinal liquid and retinoschisis resolution, with a final VA of 0.7 (Fig. 5).

Discussion

The Morning Glory syndrome is a non-hereditary and usually unilateral optic nerve disease exhibiting increased excavation, neuroretinal ring hypopigmentation, radial retinal vessels with gylial tissue and a funnel-shaped arrangement. Occasionally, it is associated to pigment deviation from the macula to the temporal side and to traction membranes.\(^2,3\)

Although the etiology of this syndrome is unknown, a theory states that it is due to an incomplete closure of the foetal fissure as in the case of optic nerve coloboma. Another theory suggests a faulty closure of the scleral wall and the cribriform plate due to poor mesenchymal differentiation. Recently, mutations in gene PAX6 had been discovered.

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Fig. 1 – (A) RE fundus: normal. Normal visual fields in gray scale. (B) LE fundus: Morning Glory anomaly. Visual fields: increased blind spot.
Fig. 2 – Nuclear magnetic resonance: callous body agenesis with absence of the intraventricular septum and communication of the frontal handles, in addition to frontal cortical dysplasia areas and signs of rupture towards the subarachnoid space and the ventricular system with the ensuing hydrocephalus.

Fig. 3 – Retinograph at 50°, photograph with green filter at 50° and linear HD OCT (composition). Morning Glory anomaly with associated macular serous detachment. OCT: papillary malformation (glyal tissue), subneurosensorry retina liquid and retinoschisis in nasal area.
Fig. 4 – AGF: superior peripapillary serous detachment and nasal to papilla foveal involvement.

Overall, 38% of cases develop retina detachment. The pathogenesis of the Morning Glory syndrome is controversial:

(1) Regmatogenous.

(2) Exudation due to abnormal retinal vessels.

(3) Communication of the vitreous humor through the optic disk to the subretinal space.

(4) Escape of cerebrospinal fluid through a communication of the subretinal space with the subarachnoid space.

Frequently the syndrome is associated to alterations in the endocrine gland, the kidney, the midline and the encephalocele.

The mean diagnostic age is of 24 months due to the appearance of anisometropic amblyopia associated to strabismus in 50% of cases, and it affects both sexes equally.

When the papillary anomaly is accompanied by retina detachment and intraocular calcifications, a differential diagnostic must be made with retinoblastoma. A thickened and winding optic nerve must be differentiated from glioma. Another disease which, even being rare, must be taken into account is the Aicardi syndrome, characterized by callous body agenesis, epilepsy and colobomatous alteration of the optic nerve head dotted chorioretinal lesions.

The treatment for retina detachment associated to the Morning Glory syndrome is controversial. Some authors

Fig. 5 – Ocular fundus evolution (retinograph at 50°) and OCT (HD foveolar scan 6 mm). (A) Macular serous detachment diagnostic with retinoschisis in nasal area. (B) After 3 months of treatment: temporal liquid displacement. Absence of liquid in foveal area. Slight persistence of retinoschisis. (C) After 12 months follow-up: subretinal liquid and retinoschisis disappeared.
recommend waiting 3 months for the spontaneous reabsorption of the subretinal liquid, while others recommend vitrectomy with posterior hyaloids peeling to release traction.\textsuperscript{1,3,4}

In our case, we opted for a simple gas injection, peripapillary positioning and laser with good tomographic and visual evolution.

In conclusion, this paper presents an exceptional case of \textit{Morning Glory} syndrome with late diagnostic due to good initial VA. The high prevalence of retinal detachment makes it necessary to carry out regular checkups. When the \textit{Morning Glory} syndrome exhibits macular serous detachment, the injection of gas and laser could be a valid therapeutic option.

\textbf{Conflict of interests}

The authors have no conflict of interests to declare.

\textbf{REFERENCES}