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Reporte de caso

Rare ocular disorder of the optic disc resulting from a defect on the development of the neuroectoderm: *morning glory* disc anomaly

Trastorno ocular infrecuente del disco óptico resultante de un defecto en el desarrollo del neuroectodermo: anomalía de *morning glory*

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RESUMEN

Introducción: Se describen los signos clínicos de la anomalía de Morning Glory en una paciente femenina. Aunque esta anomalía es de baja prevalencia, la reportamos por la importancia del diagnóstico temprano debido a la asociación con alteraciones vasculares del sistema nervioso central que eventualmente son tratables. *Objetivo:* describir esta anomalía del disco óptico en una paciente atendida en el Hospital de San José, Bogotá. *Métodos:* búsqueda en la literatura de las manifestaciones clínicas, incidencia y patologías asociadas. *Presentación del caso:* mujer que consulta por pobre agudeza visual del ojo derecho cuya fundoscopia reveló una excavación cónica con disco óptico displásico y tejido glial alrededor de la pupila en 360 grados, pigmento retinocoroideo y aumento de los vasos retinianos que emergen del disco en distribución radial.

Palabras clave: anomalía llamada amanecer o mañana de esplendor de la papila óptica, desarrollo del neuroectodermo, vasos retinianos, distribución radial.

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ABSTRACT

Introduction: We describe the clinical findings of morning glory optic disc anomaly, in a female patient. Despite the low prevalence of this optic disc malformation and although it has no specific treatment, we report this case, for timely diagnosis is important for associated vascular and central nervous system abnormalities may eventually be treated. *Objective:* to report this case of morning glory optic disc anomaly in a patient seen at Hospital de San José, Bogotá. Study design: case report. *Methods:* A search in the literature on the incidence, ophthalmological manifestations, clinical presentation and concomitant pathologies of morning glory optic disc anomaly, was conducted. *Case report:* female patient that consulted for decreased visual acuity in the right eye. Right eye fundus examination revealed a funnel-shaped excavation, dysplastic optic disc and glial tissue covering the peripapillary region circumferentially (360°), chorioretinal pigmentary changes, and increased number of retinal vessels emerging from the optical disc in a radial pattern.

Keywords: morning glory disc anomaly, neuroectodermal development, retinal vessels, radial distribution.

INTRODUCTION

Morning glory disc anomaly (MGDA) was initially described in German literature by Handmann in 1929[1], but it was Kindler in 1970 who first termed the anomaly morning glory disc anomaly for its similarity to the morning glory flower due to sub-retinal fluid content in the disc area that creates the delusion of petals coming out of the optic nerve.[2,3] This anomaly is characterized by an enlarged optic disc and a central white glial tuft[4,5], deep excavation, lack of regularity in the contour, pigment and scleral alterations and peripapillary retinal vessels emerging radially in a straight course.[3,6] Other associated ocular abnormalities are macular capture and lutein pigment visualized in the outer segment of the defect.[1,3,7] The anomaly is typically unilateral and is not an inherited condition.[1] Visual acuity is poor, but about 30% of patients improve to a best corrected visual acuity of 20/40[3], they can also refer visual field defects including hemianopia or scotoma. Strabismus and leukocoria are other frequent reasons for consultation, especially in children, and may be related with persistent hyperplastic primary vitreous, cataracts, nystagmus, ciliary body cysts, lens coloboma, optic nerve drusen, microphthalmia and intraocular calcifications.[3,8,9] MGDA is seen more commonly in females and the incidence in the world population is very low,[10] being less common in Afro-descendants.[11]

One of the most common intraocular complications is non-rhegmatogenous retinal detachment which runs from the optic nerve head across the inner layer of the eye through the scIeral apperture causing continuous traction with axial displacement of the optic nerve.[3] There are multiple systemic alterations such as: midfacial and nervous system anomalies such as hypertelorism, cleft lip and cleft palate. Other associated findings are transphenoidal, sphenoethmoidal, sphenopharyngeal, and transethmoidal encephalocele. Agenesis of the corpus callosum, endocrine alterations due to pituitary dysfunction, hormonal alterations and diabetes insipidus have been reported in some patients.[1,3] Cerebrovascular anomalies such as moyamoya disease which features intellectual disability, cerebral stroke and convulsions, have also been demonstrated.[3] Aortic arch, subclavian and carotid aneurysms, and capillary facial hemangiomas[3] are also related with this syndrome.

Imaging studies allow identifying abnormalities related to MGDA, such as intracranial or vascular alterations which may require urgent surgical intervention. Ultrasonography B-scans allow detecting intra-ocular calcifications and overhanging retinal tissue covering a scleral posterior staphyloma.[3] Computed tomography is useful for diagnosing posterior scIeraI staphyloma, optic nerve thickening and calcifications.[1] Magnetic resonance imaging usually demonstrates a funnel-shaped optic disc with elevation of the adjacent retinal surface, and alterations of the distal intra-orbital segment of the optic nerve with effacement of the regional subarachnoid spaces and discontinuity of the uveoscleral coat. MGDA appears to be the result of a failure of normal neuroectodermal development before the seventh week of embryonic growth[3] along with primary mesenchymal abnormality, anomalies of the relative growth between mesoderm and ectoderm or an abnormal closure of the embryonic fissure. The PAX6 gene is thought to be involved[12] in the genetic changes originating MGDA.

CASE PRESENTATION

A 51-year-old female patient who presented with a history of alveolar diffuse hemorrhage associated with microscopic

polyangiitis with positive anti-neutrophil cytoplasmic antibodies treated with cyclophosphamide, severe mitral insufficiency, rapidly progressive glomerulonephritis managed with hemodialysis, pulmonary embolism treated with anticoagulant therapy, pulmonary hypertension, normocytic anemia, peptic acid disease, secondary hyperparathyroidism and depressive disorder. Patient was referred to the ophthalmology service for acute redness of the right eye of 10 hour duration, following a hemodialysis associated with marked hypertension. Ophthalmologic history was unremarkable for patient stated having no surgical or family history and no prescribed glasses. Patient underwent detailed ophthalmologic examination with the following results: Retinoscopy: right eye (OD): best corrected visual acuity: 20/150, 5.25-0.50 x 0°, left eye (OS): 20/200, 3.00-0.75 x 170°. Exophthalmometry: 15 mm/16 mm; Biomicroscopy OD: dermatochalasis, telangiectasia of the upper eyelid margin, perilimbal superior and inferior subconjunctival hemorrhage, clear cornea, anterior chamber: no cells, lens: incipient nuclear sclerosis; OS: white and quiet conjunctiva, clear cornea, formed anterior chamber, no cells, lens: incipient nuclear sclerosis; Mean intraocular pressure: 14 mm Hg in both eyes. Fundoscopic examination OD **(figure 1)**: peripapillary abundant glial tissue, cup/disc ratio (C/D) 0.3, 360° scleral crescent, peripapillary retinal pigment and increased number of retinal vessels emerging from the optical disc radially, attached retina and no alterations in the periphery; OS **(figure 2):** clear cornea, round-shaped papilla, C/D 0.2, normal neural ring, defined contour, macula: no retinal light reflex, attached retina. Diagnosis: subconjunctival hemorrhage in the right eye, morning glory disc anomaly in the right eye, and refractive amblyopia in the left eye.

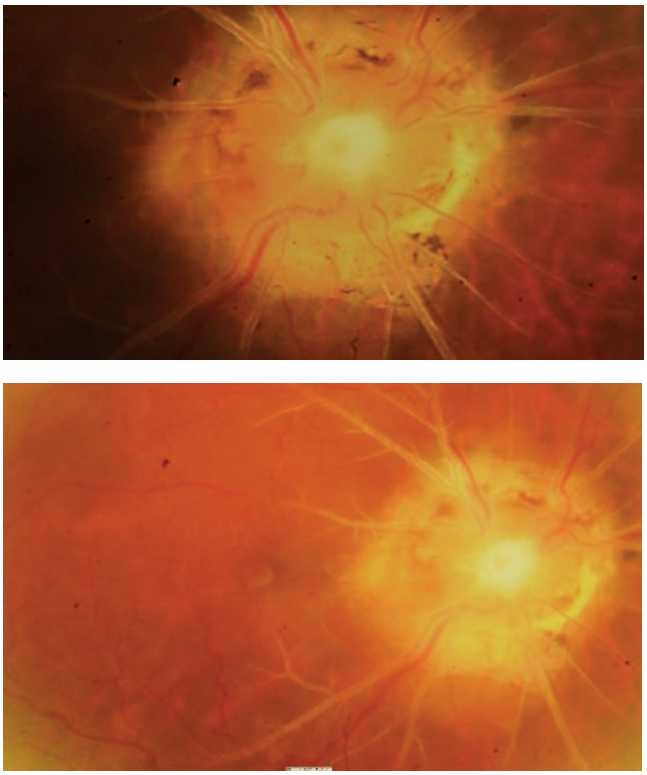

Figure 1 A y B. Optic nerve of the right eye. Source: the authors.

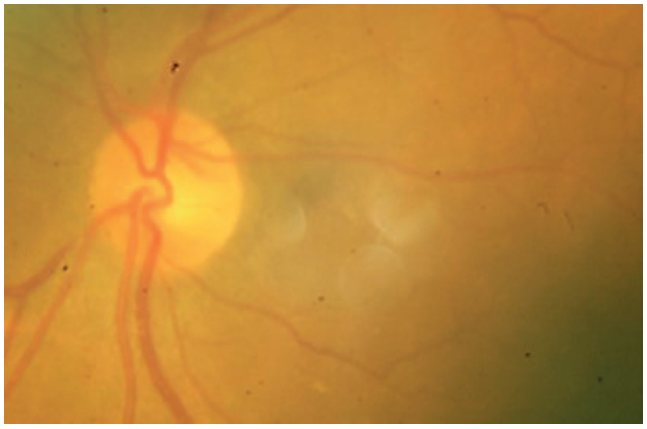

Figure 2. Optic nerve of the left eye. Source: the authors.

DISCUSSION

Anatomic alterations of the optic nerve and of the retina similar to those described in the world literature, were found in our patient, the optic disc was enlarged with a funnel-shaped optic nerve covered by glial tissue and lack of regularity in the contour, peripapillary pigmentary changes and straight retinal vessels emerging in a radial spoke-like pattern.[1,3] Morning glory disc anomaly is usually associated with low vision[3], which coincides with our case report, since the patient had a best corrected vision of 20/200 in the affected eye. Our case was a female patient and agreed with the highest prevalence of gender distribution reported in the literature, and unilateral presentation is more frequent as in our patient.

CONCLUSION

Morning glory disc anomaly can be found during a regular eye check-up. It´s low incidence prompts appropriate recognition of its signs and symptoms amongst physicians of various specialties in order to achieve an accurate diagnosis and provide timely treatment to these patients, taking into account that some cases may require urgent intervention.

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