Morning Glory Syndrome: The Optic Disc Flower in Two Patients

Síndrome de Morning Glory: A Flor do Disco Óptico em Dois Pacientes

Beatriz Soares [1], Elsa Góis[2], Carla Ferreira [2], Marco Vieira [2], Nuno Gomes [1]*

***Corresponding Author**/Autor Correspondente:
Beatriz Cabral Soares [beatrizclsoares@gmail.com]
Praceta Maria Luísa Canavarro 52 - 2º Dt., 4445-506 Ermesinde, Portugal

10.48687/lsj.v2i4.83

Abstract

The morning glory syndrome (MGS) was first described by Handmann in 1929. This syndrome is a rare congenital malformation of the optic disc (OD), usually unilateral. Fundus findings include a large OD with indistinct borders surrounded by depigmented areas with a central funnel shaped excavation, glial tissues and straight vessels radiating out. The main purpose of this study is to report 2 cases with this rare syndrome seen at our Ophthalmology department in 2021. We have included 2 patients, one male and one female who presented with complains of vison loss. On examination a diagnosis of MGS was established. Although rare, clinicians must keep in mind the diagnosis of MGS to avoid unnecessary exams and to be able to advise the patients in the best possible way.

Keywords: Optic Disc/abnormalities; Optic Nerve Diseases/congenital

Resumo

Handmann foi o primeiro a descrever a síndrome de *morning glory* (SMG) em 1929. Esta síndrome é uma malformação congénita rara do disco óptico, normalmente unilateral. Achados fundoscópicos incluem um disco aumentado com os bordos pouco definidos, rodeada por áreas despigmentadas, uma escavação em forma de funil, presença de tecido glial e vasos finos. O objetivo deste estudo é reportar 2 casos desta síndrome rara observados no nosso departamento de Oftalmologia em 2021. Foram incluídos dois pacientes, um masculino e um feminino que apresentavam queixas de perda de visão. Ao exame objetivo foi confirmado o diagnóstico de MG em ambos. Apesar de rara, deve-se ter em conta o diagnóstico de SMG de modo a evitar exames desnecessários e ser capaz de aconselhar os pacientes da melhor maneira possível

Palavras-chave: Disco Óptico/anomalias congénitas; Doenças do Nervo Óptico/congénito

1. Serviço de Oftalmologia, Hospital Lusíadas Porto, Porto, Portugal. **2.** Serviço de Oftalmologia, Hospital Lusíadas Braga, Braga, Portugal.
Received/Recebido: 04/12/ 2021 **- Accepted/Aceite:** 12/12/2021 **- Published/Publicado:** 31/12/2021

Introduction

In 1929 a group of optic disc (OD) anomalies was observed by Handmann. Later in 1970, these anomalies were named by Kindler as morning glory (MG), the origin of this name is linked with the appearance of the papillary region that resembles a blooming tropical flower with the same name.[1]

MG is a rare congenital anomaly that involves the OD, and is associated with a peripapillary scleral defect, absence of lamina cribosa and a recess formed by an axial retrodisplacement.[1,2]

In this pathology the disc appears enlarged and presents a funnel-shaped excavation, presence of glial tissue and a chorioretinal pigmented ring.[1,2]

This anomaly is typically unilateral, but there have been bilateral cases reported in the literature. MGS is also more common in females with a ratio of 2:1. Recent reports indicate a prevalence of 2.6/100 000.[3,4]

We present two cases of MGS, one on a female patient and one on a male patient.

Case Reports

Case 1

A 20-years-old man was referred to our ophthalmology service for a differential diagnosis of an OD anomaly. His medical and ocular history were unremarkable.

Ocular examination revealed a best corrected visual acuity of 20/20 on both eyes. The refractive error of the right eye was +0.50 (-1.00) 170º and of the left eye was -2.00. An afferent pupillary defect was not noticed during examination. Biomicroscopic evaluation of the eye was normal.

Fundus examination of the left eye (Fig. 1) was normal; however, the right eye presented an enlarged OD, with funnel shaped excavation. The edges of the disc were slightly blurred, and the disc was surrounded by a chorioretinal pigmented ring. A collection of white glial tissues was present at the bottom of the disc, anomalous blood vessels emerged from the centre of the disc in a radial pattern, they were more numerous and thinner. There was a serous retinal detachment of the inferior retina.

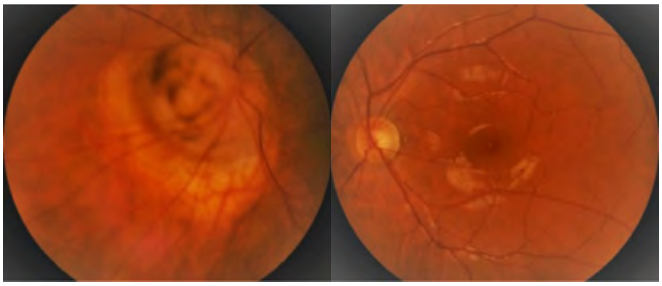

Figure 1. Male, 20 years. Disc changes visible on the right eye.

Case 2

A 22-year-old woman presented at our ophthalmology service for a routine appointment. She had been previously diagnosed on her right eye with microphthalmia and endotropia. On the appointment the patient presented a visual acuity of light perception in her right eye and 20/25 in her left eye. The refractive error on the right eye was emmetropic and on the left eye -3.00 40º. Krimsky test the right eye confirmed the endotropia. It also presented nystagmus and microphthalmia. An afferent pupillary defect was not noticed during examination.

The fundus of the right (Fig. 2) eye showed an enlarged OD with a funnel-shaped excavation, white glial tissue was visible at the centre of the disc. The edges of the optic disc were uneven and was also possible to observe a chorioretinal pigmented ring. Anomalous blood vessels emerged from the optic disc borders, they appeared thinner and more numerous and presented a radial pattern. It was also possible to observe remains of the hyaloid. There was no signal of retinal detachment. Examination of the left eye was normal.

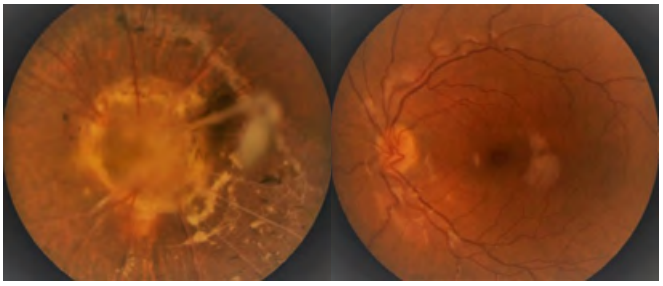

Figure 2. Female, 22 years. MGS in the right eye.

Discussion

MGS is a congenital disc anomaly first described by Handman and then named by Kindler in 1970. This pathology can also be associated with some systemic malformations.[3,5,6]

This syndrome is rare with a prevalence of 2.6/100 000, it is predominant on females with a sex ratio of 2:1. In most cases MG is unilateral however bilateral cases have been reported. In this case report we present two patients one female and one male both with a unilateral form of this pathology.[3,4]

Morning glory pathogenesis is controversial, Schneider thought that this pathology was a form of coloboma, where the defect was central, but later Mauschot showed us that there is no neuroectodermal defect. Instead, he suggested that the morning glory was a primary mesodermal congenital lesion, caused by an incomplete closure of the scleral posterior wall and lamina cribosa during the 6th week of intrauterine life. The genes linked with this pathology are not well known, but the *PAX6* gene could be linked to the anomaly.[2]

MG has some specific characteristic as an enlarged, excavated and funnel shaped OD; a chorioretinal pigmented ring

that can be found on the peripapillary area; glial proliferation that is present on the centre of the disc or at the bottom; and anomalous retinal vessels that emerge radially from the centre of the disc - these vessels are normally more numerous and thinner.[1,2,4]

Patient with MG may have other complications in the affected eye or less frequently in the contralateral eye. Some of these findings include strabismus, amblyopia, congenital cataracts, coloboma of the lens, nystagmus, optic nerve drusen's, glaucoma, persistent hyaloid remnants, microphthalmia and serous retinal detachment. This last one occurs in approximately 30% of the cases. Some of these anomalies were present in our patients, such as serous retinal detachment, endotropia, microphthalmia and persistent hyaloid remnants.[1,3,7,8]

Visual acuity is normally severely compromised being typically worse than 20/200. In our case report we had a patient with a visual acuity reduced to light perception. However visual acuity of 20/20 has been reported as was described with the other patient.[3]

Conclusion

We present two different cases of MGS observed in our hospital. These cases showed the degree of variability of this disease. One was a male patient, and the other was a female patient. One of our cases presented many complications associated with this pathology while the other one had just one complication. Visual acuity and fundoscopy presentations were also different. Although rare, clinicians must keep in mind the diagnosis of MGS to avoid unnecessary exams and to be able to advise the patients in the best possible way.

Responsabilidades Éticas

Conflitos de Interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.

Fontes de Financiamento: Não existiram fontes externas de financiamento para a realização deste artigo.

Confidencialidade dos Dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.

Consentimento: Consentimento do doente para publicação obtido.

Proveniência e Revisão por Pares: Não comissionado; revisão externa por pares.

Ethical Disclosures

Conflicts of Interest: The authors have no conflicts of interest to declare.

Financing Support: This work has not received any contribution, grant or scholarship.

Confidentiality of Data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Patient Consent: Consent for publication was obtained.

Provenance and Peer Review: Not commissioned; externally peer reviewed.

ORCID iD: Beatriz Cabral Soares https://orcid.org/0000-0003-3378-1846

References

1. Bakayoko S, Coulibaly B, Sidibe MK, Gyrr E, Sissoko M, Saye G,et al. Morning Glory syndrome in two malians: case report from IOTA-University Hospital. Open J Ophthalmol. 2020;10.
2. Osaguona VB, Momoh RO. Morning glory syndrome in a Nigerian: a case report. J West African Coll Surg. 2017;7:128-34.
3. Grondalski S. Morning glory syndrome with good visual acuity. Orlando: American Academy of Optometry; 1996. [accessed Aug 2021] Available from: https://www.aaopt.org/detail/knowledge-base-article/morning-glory-syndrome-good-visual-acuity.
4. Jiang H, Liang Y, Long K, Luo J. Postoperative follow-up of a case of atypical morning glory syndrome associated with persistent fetal vasculature. BMC Ophthalmol. 2019;19:150. doi: 10.1186/s12886-019-1154-6.
5. Araissi AB, Fayed A, Helmy Y. Bilateral morning glory syndrome with bilateral persistent fetal vasculature in a patient with Joubert's syndrome. Case Rep Med. 2020;2020:5872758. doi: 10.1155/2020/5872758.
6. Dedhia CJ, Gogri PY, Rani PK. Rare bilateral presentation of morning glory disc anomaly. BMJ Case Rep. 2016;2016:bcr2016215846. doi: 10.1136/bcr-2016-215846.
7. Hodgkins P, Lees M, Lawson J, Reardon W, Leitch J, Thorogood P, et al. Optic disc anomalies and frontonasal dysplasia. Br J Ophthalmol. 1998;82:290-3. doi: 10.1136/bjo.82.3.290.
8. Shami M, McCartney D, Benedict W, Barnes C. Spontaneous retinal reattachment in a patient with persistent hyperplastic primary vitreous and an optic nerve coloboma. Am J Ophthalmol. 1992;114:769-71. doi: 10.1016/s0002-9394(14)74061-9.