Case Report

EJO European Journal of Ophthalmology

European Journal of Ophthalmology
1–4

DOI: 10.1177/1120672119864010
journals.sagepub.com/home/ejo
SAGE

Optical coherence tomography angiography in contractile morning glory syndrome

Gilda Cennamo[1], Daniela Montorio[2], Maria Angelica Breve[2], Vincenzo Brescia Morra[2] and Giovanni Cennamo[2]

Abstract

This study describes the optical coherence tomography and optical coherence tomography angiography features of three eyes of three patients affected by contractile morning glory syndrome. Optical coherence tomography angiography scans of the peripapillary retina revealed a dense microvascular network without any vascular difference between the superficial vascular plexus and the deep vascular plexus around the optic nerve. These optical coherence tomography angiography findings confirm that the contractile movement could be due to the presence of an autonomic cholinergic muscular mechanism in the posterior part of the globe. In fact in our cases, the contractile movement seemed to be induced by massage of the eyeball. Optical coherence tomography angiography is a valid, non-invasive, dyeless, and reliable method that could shed light on the pathogenesis of this rare disease of the optic disk.

Keywords

Morning glory syndrome, optical coherence tomography angiography, spectral domain optical coherence tomography

Date received: 5 January 2019; accepted: 26 June 2019

Introduction

Morning glory syndrome (MGS) is a rare congenital optic disk dysplasia characterized by a funnel-shaped excavation involving the optic disk. This excavation is filled with glial tissue and a pigment ring slightly protrudes into the peripapillary area and an increased number of straight retinal vessels radially emerging from the disk margin.[1]

MGS can be non-contractile (non-contractile morning glory syndrome (NCMGS)) or, in rare cases, it can be associated with contractile movements of the optic disk (contractile morning glory syndrome (CMGS)).[1,2]

Different mechanisms have been proposed to explain the etiology of the contractile movements.

In an attempt to shed light on this issue, we report three cases of contractile MGS evaluated by optical coherence tomography angiography (OCTA).

Cases description

We studied three eyes of three patients (two females and one male) attending the Eye Clinic of the University of Naples "Federico II" for an optic nerve anomaly.

The mean age was 19.7 ± 1.5 years (range: 18–21 years), the mean best-corrected visual acuity was 0.9 ± 0.1 logMar (range: 1–0.8 logMar), and the anterior segment did not show any alteration and strabismus was not present.

Fundus examination revealed an enlarged optic disk surrounded by annular chorioretinal atrophy with an overlying thin glial membrane in its central portion.

The retinal vessels were increased in number and appeared to arise from the disk margin from where they tended to run to the peripheral retina in a straighter course than usual.

The macula and the surrounding retina appeared normal and the fellow eye did not show any retinal alterations.

[1]Eye Clinic, Department of Public Health, University of Naples Federico II, Naples, Italy
[2]Department of Neurosciences, Reproductive Sciences and Dentistry, University of Naples Federico II, Naples, Italy

Corresponding author:
Gilda Cennamo, Eye Clinic, Department of Public Health, University of Naples Federico II, Via S. Pansini 5, Naples 80133, Italy.
Email: xgilda@hotmail.com

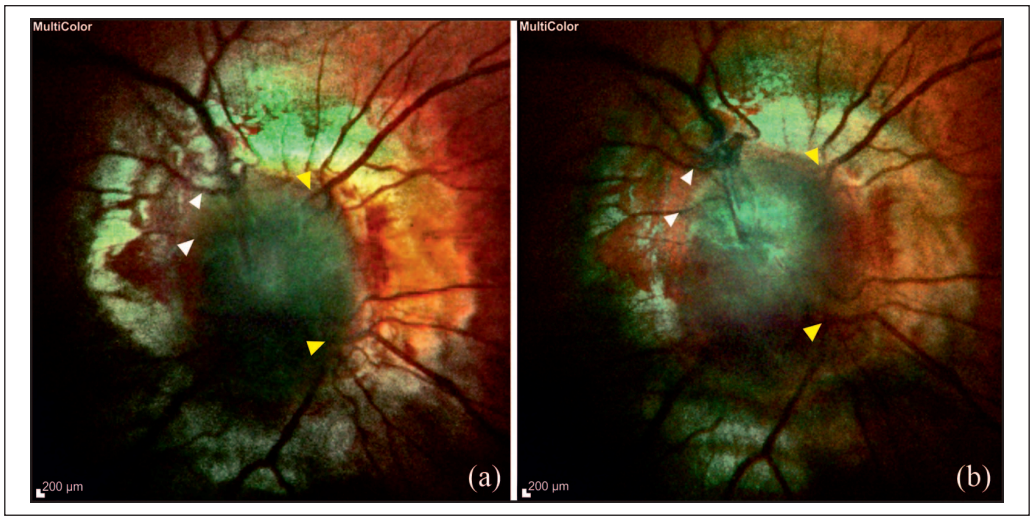

Figure 1. Case 1: multicolor image of the contractile morning glory optic disk in right eye of a female patient. (a) Non-contracted phase: the optic disk was enlarged and surrounded by annular chorioretinal atrophy with overlying thin glial membrane in its central portion. An increased number of the retinal vessel arose from the disk margin with a straight course (white arrowhead). (b) Contracted phase: during the contraction some peripapillary vessels changed slightly their direction (yellow arrowhead).

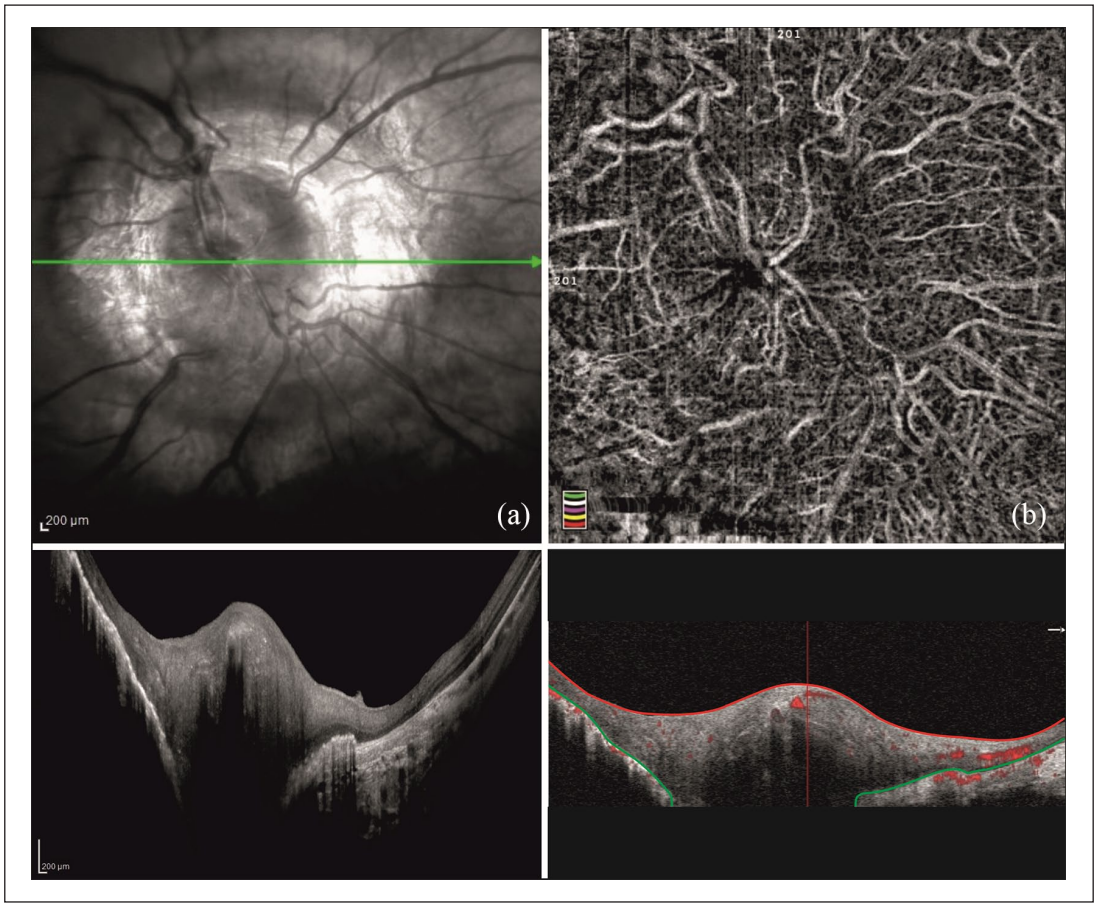

Figure 2. Case 1: (a) single-line spectral domain optical coherence tomography (SD-OCT) through the morning glory optic disk shows the deep excavation and a raised hyperreflective area corresponding to the presence of glial tissue overlying the optic disk. (b) Optical coherence tomography angiography (OCTA) of the optic disk reveals a dense microvascular network in radial peripapillary capillary (RPC) without any vascular difference between the superficial vascular plexus and the deep vascular plexus around the optic nerve.

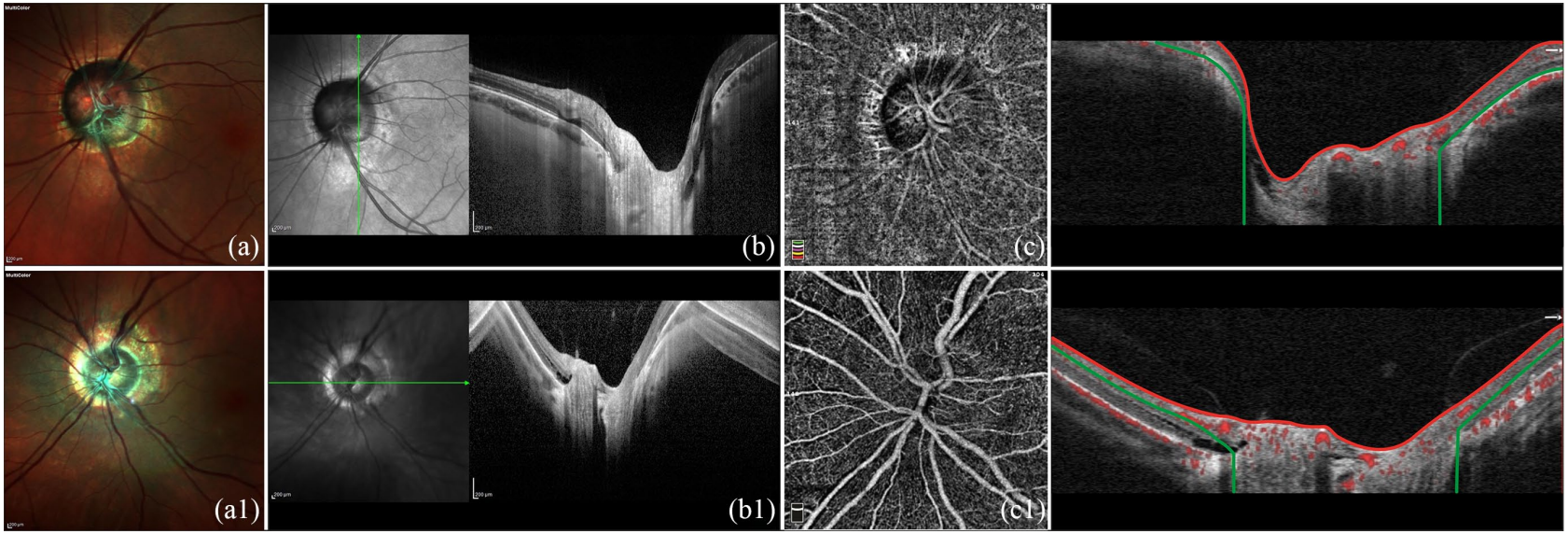

Figure 3. Multicolor images of the optic disk in left eye of a female patient ((a), case 2) and in left eye of a male patient ((a1), case 3) with contractile morning glory syndrome (CMGS) show the optic disk enlarged and surrounded by annular chorioretinal atrophy with overlying thin glial membrane in its central portion. (b, b1) (cases 2 and 3, respectively) Single-line SD-OCT through the morning glory optic disk shows the deep excavation with hyperreflective area corresponding to the presence of glial tissue overlying the optic disk and subretinal fluid. (c, c1) (cases 2 and 3, respectively) OCTA of the optic disk reveals a dense microvascular network in RPC in both cases.

During the fundus examination, the contractions of the staphyloma were shown after applying finger pressure on the eyeball and these contractions changed slightly the direction of the peripapillary vessels (Figure 1(a) and (b)).

Spectral domain optical coherence tomography (SD-OCT) (Spectralis HRA; Heidelberg Engineering, Heidelberg, Germany) over the optic disk showed the deep excavation and a raised hyperreflective area corresponding to the presence of glial tissue overlying the optic disk (Figures 2(a) and 3(b) and (b1)).

OCTA images were acquired with the Angiovue System (RTVue XR Avanti; Optovue, Inc., Freemont, CA, USA). The scans were centered on the optic disk and the radial peripapillary capillary (RPC) layer were obtained using the Angio Disc mode that analyzed the network of capillaries from the inner limiting membrane to the posterior boundary of the radial nerve fiber layer.[3]

OCTA scans of the peripapillary retina revealed a dense microvascular network in RPC without any vascular difference between the superficial vascular plexus and the deep vascular plexus around the optic nerve, confirming the findings observed also in a previous study (Figures 2(b) and 3(c) and (c1)).[4]

These OCTA findings did not support the hypothesis of a vascular malformation in the optic nerve in CMGS, as reported by Dempster et al.,[5] but confirm that the contractile movement could be due to the presence of an autonomic cholinergic muscular mechanism in the posterior part of the globe.[6]

This hypothesis is confirmed by the fact that in our cases the contractile movement seemed to be induced by massage of the eyeball, as shown in the study by Cennamo et al.[1,2]

Conclusion

We suggest that OCTA may be useful to shed light on the pathogenesis of this rare disease of the optic disk.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

References

1. Cennamo G, de Crecchio G, Iaccarino G, et al. Evaluation of morning glory syndrome with spectral optical coherence tomography and echography. *Ophthalmology* 2010; 117(6): 1269–1273.
2. Cennamo G, Sammartino A and Fioretti F. Morning glory syndrome with contractile peripapillary staphyloma. *Br J Ophthalmol* 1983; 67(6): 346–348.
3. Mase T, Ishibazawa A, Nagaoka T, et al. Radial peripapillary capillary network visualized using wide-field montage optical coherence tomography angiography. *Invest Ophthalmol Vis Sci* 2016; 57(9): OCT504–OCT510.
4. Cennamo G, Rossi C, Ruggiero P, et al. Study of the radial peripapillary capillary network in congenital optic disc anomalies with optical coherence tomography angiography. *Am J Ophthalmol* 2017; 176: 1–8.
5. Dempster AG, Lee WR, Forrester JV, et al. The 'morning glory syndrome'—a mesodermal defect. *Ophthalmologica* 1983; 187(4): 222–230.
6. Kral K and Svarc D. Contractile peripapillary staphyloma. *Am J Ophthalmol* 1971; 71: 1090–1092.