Case Report

Unusual Trigger for Contractile Movements of Optic Disc in Peripapillary Staphyloma

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Abstract

Purpose: To report a case of contractile movements of the optic disc in peripapillary staphyloma.

Methods: A 65-year-old female received a comprehensive ophthalmic examination, multimodal imaging, and computed tomography.

Results: The patient was diagnosed with peripapillary staphyloma in her right eye. Best-corrected visual acuity in her right and left eye was 20/32 and 20/40, respectively, with moderate cataracts in both eyes. Multimodal imaging revealed contractile movements of the optic disc in the right eye that were initiated by the circular rotation of the eye but by none of the triggers previously described in the literature. The patient reported no changes in her vision during contractile movements. Optical coherence tomography revealed a muscle-like structure in the optic disc during the peak of the contraction. Computed tomography did not reveal any abnormality of the optic nerve or the extraocular muscles of the right eye.

Conclusion: Contractile movements in peripapillary staphyloma may be initiated by previously unknown triggers. The pattern of the contractile movements and optical coherence tomography findings support the muscular nature of these movements.

Keywords: Contractile movements, Morning glory optic disc anomaly, Optic disc, Optical coherence tomography, Peripapillary staphyloma

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Introduction

Peripapillary staphyloma is a rare sporadic congenital unilateral condition, characterized by staphylomatous excavation of the eye fundus around the optic disc and with a relatively normal appearance of the optic nerve head at the bottom of the excavation. Despite this relatively normal appearance, peripapillary staphyloma is considered a part of the spectrum of congenital excavated optic disc anomalies which include optic disc coloboma, morning glory disc anomaly (MGDA), and optic pit.[1] In contrast to peripapillary staphyloma, in MGDA, the optic disc has an abnormal vascular pattern and central glial tuft, whereas optic disc coloboma is often associated with iris or retinochoroidal coloboma.

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Although these conditions have distinct origins, some cases of peripapillary staphyloma, MGDA, and optic disc coloboma have one phenomenon in common contractile optic disc movements.[2-5] Most descriptions of contractile optic disc movements refer to peripapillary staphyloma,[2,3,6,7] thought to be caused by the activity of the rudimentary muscle retractor bulbi.[2] Various triggers for these contractile movements have been reported suggesting a causative role of the retractor bulbi muscle. In this study, we describe contractile movements of the optic disc in peripapillary staphyloma, which are triggered by a specific eyeball movement.

Case Report

A 65-year-old female with progressive deterioration of visual acuity in her left eye was presented for ophthalmic examination.

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The medical history of the patient was unremarkable for eye diseases. Her medical records were significant for 50% stenosis of the right internal carotid artery. The best-corrected visual acuity (BCVA) in her right and left eye was 20/32 and 20/40, respectively. Axial length was 22.1 mm in both eyes. Biomicroscopy of the anterior eye segment revealed cataract LOCS 2+ in both eyes. Indirect ophthalmoscopy showed no significant changes in the left eye but an enlarged optic disc with wide excavation, circumferentially radiating vessels, and accompanied by surrounding depigmentation in the right eye. The patient was diagnosed with peripapillary staphyloma in the right eye. During indirect ophthalmoscopy, contractile movements of the optic disc were observed, including a period of progressive contraction of the excavation over approximately 3 s [Figure 1a and b]. A stable contraction with the absence of visible excavation lasted approximately 3 s, followed by a gradual opening of the excavation in a series of weakening contractions with a frequency of approximately 1 Hz. No obvious latency period was found in a series of four consecutive episodes of contractile activity. Neither the Valsalva maneuver, physical exercises, breath holding, pressure on the eye, forced eye closure, accommodative demand, or strong light stimulation to the fellow nor the affected eye allowed initiation of these contractile movements. The only stimulus which could intentionally trigger these movements was a circular rotation of the eye in a clockwise or counter-clockwise direction. To induce contractile movements, the patient was asked to quickly move their gaze over 360°. This phenomenon was registered with video scanning laser ophthalmoscopy (F-10, NIDEK, Gamagori, Japan) [Supplemental Video 1] [Figure 1c and d]. Simple eye movements in the secondary, as well as in the tertiary positions failed to cause these contractile movements. Structural optical coherence tomography during the contraction revealed complete closure of the optic disc excavation by the tissues of the excavation walls moving inward and upward. There were no transient changes in the adjacent retina. At the peak of the contraction, optical coherence tomography revealed a hyperreflective ovoid structure with muscular morphology visible at the nasal side of the optic nerve which may correspond to the muscle responsible for the contractile movements [Figure 1e and f]. During the contraction, the walls of the excavation moved centripetally. Closure of the excavation was observed with optical coherence tomography throughout the entire excavation depth. The presumed muscular structure pushed the excavation walls toward the center of the excavation raising the overlying retina toward the vitreous cavity. At the peak of the contraction, this structure occupied the space between the retina and the lamina cribrosa. Computed tomography did not find any visible abnormality in extraocular muscles or the retrobulbar part of the optic nerve in either the left or right orbit. Written informed consent was obtained from the patient after an explanation of the purpose of the study.

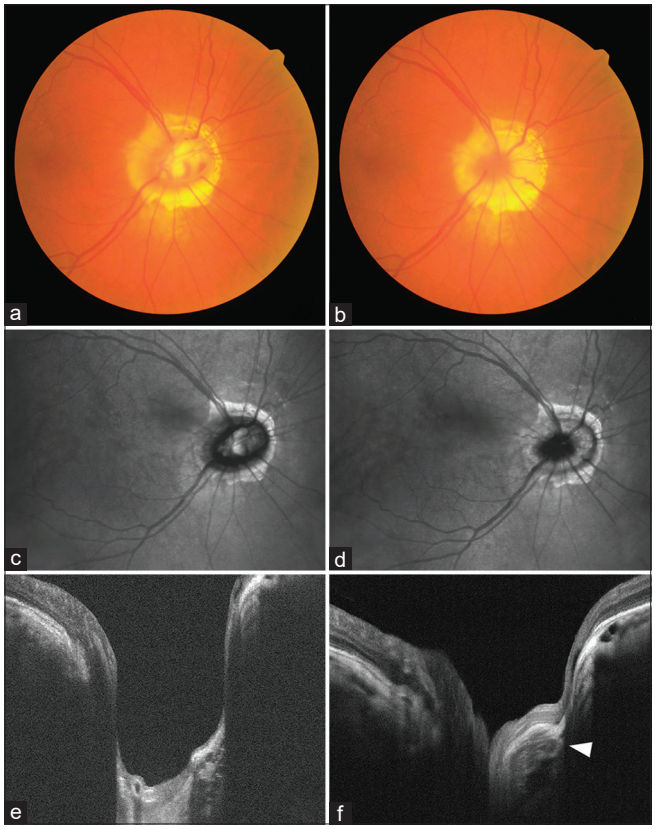

Figure 1: Multimodal imaging of the right eye. (a) Color fundus photography taken without contraction of the optic disc. (b) Color fundus photography taken during the peak of contraction of the optic disc. (c) Infrared scanning laser ophthalmoscopy image taken without contraction of the optic disc. (d) Infrared scanning laser ophthalmoscopy image taken during the peak of contraction of the optic disc. (e) Optical coherence tomography B-scan taken without contraction of the optic disc. (f) Optical coherence tomography B-scan taken during the peak of contraction of the optic disc. Arrowhead indicates structure with muscular morphology possibly responsible for the contractile movements

Discussion

Contractile movements of the optic disc in cavitary optic disc anomalies have been described in a number of cases since 1966.[2] The first cases describe this unusual finding as contractile peripapillary staphyloma.[2,3,8] After the description of MGDA as a distinct condition in 1970,[9] some cases of contractile movements of the optic disc were also referred to as MGDA. Since then, over 20 cases of contractile movements of the optic disc in cavitary optic disc anomalies have been described in the literature.

MGDA is an extremely rare sporadic congenital condition characterized by anomalous enlargement of the optic disc and/or its excavation.[9,10] Other characteristics of MGDA include an abnormal vascular branching pattern, glial remnants in the center of the optic disc, and, in some cases, retinal changes, including retinoschisis or retinal detachment.[1] MGDA has no known genetic predisposition and affects both males and females equally, with BCVA varying from mildly deteriorated (20/40 or higher) to extremely low.[1] Although the optic disc in our case demonstrated circumferentially radiating

vessels, there was no central glial tuft on the disc or choroidal coloboma, which allows us to exclude MGDA and optic disc coloboma, respectively.

Optic disc contractions can be spontaneous,[3] but in the majority of cases may be intentionally initiated with a specific trigger such as pressure on the eye,[11] forced lid closure,[2] or light stimulation to the affected[2,4] or contralateral eye.[12] The pattern of the movements described in some previous cases is similar to that observed in our patient and may reflect muscular spasm followed by depletion of the muscular activity, which results in a gradual opening of the excavation. Although some descriptions found a correlation between contractile movements and breathing,[13] these cases probably are of different nature and should not be referred to as true contractile movements. A potential mechanism for this type of movement includes imbalance of pressure between the subarachnoid space and the juxtapapillary subretinal space probably in the presence of an anomalous communication between them.

Intrascleral smooth-muscle cells were found around the bulbar segment of the optic nerve during histopathological examination in a patient with optic nerve coloboma, and this may explain the contractile movements.[14,15] In our case, a muscle-like structure that may be responsible for contractile activity was revealed with optical coherence tomography as a moderately reflective structure with an ovoid profile in cross-section. Previously, a candidate structure possibly responsible for contractile activity was noted by Yoshida *et al*. using swept-source optical coherence tomography.[7] Histopathological and optical coherence tomography data, together with the pattern of contractile movements, suggest that the mechanism of contraction is based on spastic movements of muscular fibers buried around the bulbar part of the optic nerve.

Since fundamentally different cavitary optic disc anomalies may demonstrate contractions of the optic nerve, we suggest that smooth muscle tissue in such anomalies is not a chance finding. In fact, this smooth muscle tissue results from dysgenesis of the mesodermal differentiation around the optic nerve which accompanies these conditions. However, descriptions of contractile movements remain rare. The severity of changes of the optic disc in MGDA and peripapillary staphyloma varies significantly, and this could determine the ability of the optic disc to contract. In our case, the optic disc had relatively normal anatomy, and this may reflect better preservation of the tissues and a higher density of the muscle cells. Another explanation for the rarity of this phenomenon is the diversity and complexity of the triggers which can initiate contractile movements. In our case, contractile movements were not initiated by any triggers previously described, and only the rotation of the eyeballs could initiate contractions.

Since cavitary optic disc anomalies are often associated with poor vision and lead to a request for ophthalmic examination, in a significant number of cases, contractile activity of the optic disc cavitary anomalies has been described in relatively young persons or even children. In our case, the contractile movements were found in a patient in her mid-60s who had been asymptomatic throughout her life. Since no known clinical findings pertain to optic nerve contractions, their prevalence may therefore be higher than the literature leads us to expect.

In conclusion, this case describes the contractile movements of the optic disc in a patient with peripapillary staphyloma, registered with multimodal imaging. In this case, contractile movements were initiated exclusively by circular rotation of the eye. The pattern of the contractile movements and optical coherence tomography findings support the muscular nature of these movements. Taken together, the specificity of the trigger and asymptomatic character of the contractile movements suggest that the prevalence of this phenomenon among patients with cavitary optic disc anomalies may have been underestimated.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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