



Case report

Optical coherence tomography and video recording of a case of bilateral contractile peripapillary staphyloma

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ABSTRACT

Purpose: To report a case of bilateral contractile peripapillary staphyloma (CPS) examined by optical coherence tomography (OCT) and video recording.

Observation: We report the clinical history of a male infant with bilateral CPS. Two ophthalmic examinations under general anesthesia were performed during follow-up examinations. During the first fundus examination when the infant was 4 months old, we observed staphylomatous excavation around the optic disc, an almost normal disc appearance at the bottom of the excavation, and irregular contraction bilaterally. OCT images showed deep excavation around the optic disc bilaterally and hyperreflective tissue beneath the sclera in the right eye. During the second examination when the child was 4 years and 1 month, in addition to the previous fundus findings, the fundus examination showed white fibrous tissue on the optic disc and pigmentation around the optic disc in the right eye and a retinal cyst in the left eye. Bilateral simultaneous video recordings showed that the contractions were not synchronized between the eyes.

Conclusions: In a patient with bilateral CPS, we observed unsynchronized contraction between the eyes and the presence of sequential hyperreflective tissue under the sclera using bilateral ophthalmic approaches.

1. Introduction

Peripapillary staphyloma (PS) is a rare, congenital disorder characterized by staphylomatous excavation around the optic disc and a relatively normal appearance of the optic nerve head at the bottom of the excavation.^{1,2} Similar disorders with excavation of the optic nerve, including optic nerve coloboma, optic disc pits, and morning glory syndrome (MGS), have been differentiated with characteristic fundus features³; however, a definite molecular and embryologic basis for differentiating these entities remains ambiguous.

In rare cases with PS or MGS, contractile movements of the optic disc region have been reported.^{3–12} Historically, two main mechanisms underlying the contractility have been discussed, i.e., pressure balance^{3,5} and muscle contraction,^{4,6–11} and the latter dominates based on evidence demonstrating the presence of smooth muscle around the optic disc and its contractility.^{13–15}

In this report, we present another rare case of bilateral CPS with optic disc contraction. The patient was examined under general anesthesia during which simultaneous videos of both eyes were recorded and swept-source optical coherence tomography (SS-OCT) was performed. In addition to the previous reports, we present additional

bilateral findings to support the muscle contraction theory that were not obtained from a unilateral case of contractile PS (CPS) or contractile MGS.

2. Case report

A 4-month-old boy was referred to our hospital with suspected poor vision and nystagmus. He had no systemic diseases including the brain and no familial history of ocular diseases. His fixation and following were poor, and he had alternating esotropia. Slit-lamp examinations showed no abnormalities in the anterior and medial segments. Funduscopy showed bilateral optic disc anomalies. Comprehensive ophthalmic examinations were performed under general anesthesia including fundus examinations using the Retcam3 ophthalmic imaging system (Clarity Medical Systems, Pleasanton, CA). General anesthesia was induced using a non-depolarizing paralyzing anesthesia. Funduscopy showed staphylomatous excavation around the optic disc, an almost normal disc appearance at the bottom of the excavation, and irregular contractions in both eyes (Figs. 1 and 2). A portion of the macula was involved in the excavation in both eyes. We diagnosed the optic disc anomalies as bilateral CPS. The contractions were not

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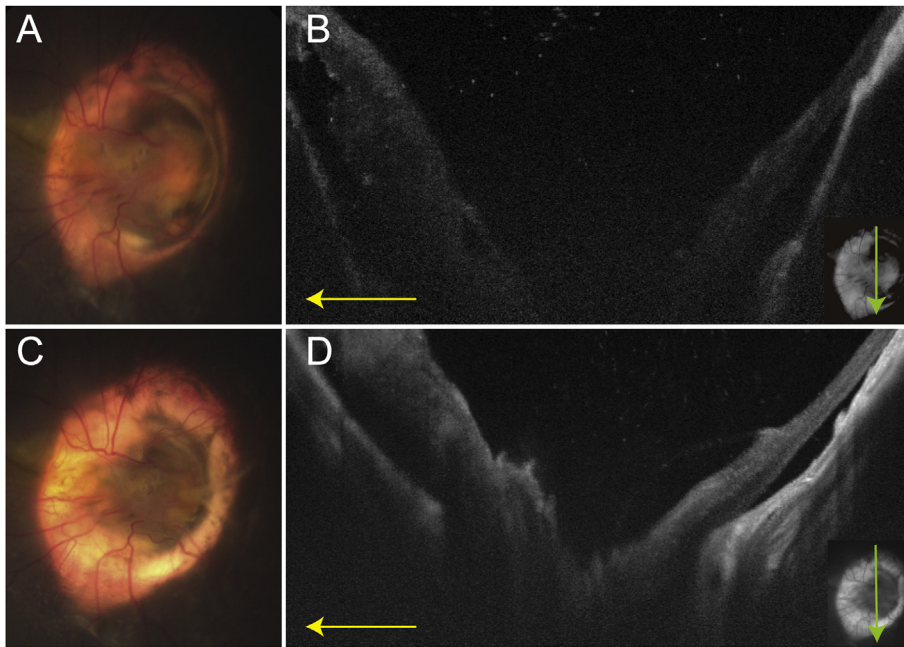


Fig. 1. Fundus photography and swept-source optical coherence tomography (SS-OCT) images of the right eye. (A) A fundus photograph of the optic nerve that is surrounded by excavation, with no contraction. A small amount of white fibrous tissue is attached to the optic nerve. (B) SS-OCT images with no contraction. Deep and wide excavation of the degenerated retina, detached from the wall of the excavation, is seen. (C) A fundus photograph of the optic nerve with contraction. The area at the bottom of the excavation becomes smaller compared with no contraction. However, the area of retinal depigmentation that surrounds the excavation enlarges compared with no contraction. (D) SS-OCT images with contraction. The diameter of the excavation becomes smaller compared with no contraction. (B) and (D) The green arrows indicate the area of the SS-OCT images. The yellow arrow indicates the direction of the images. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

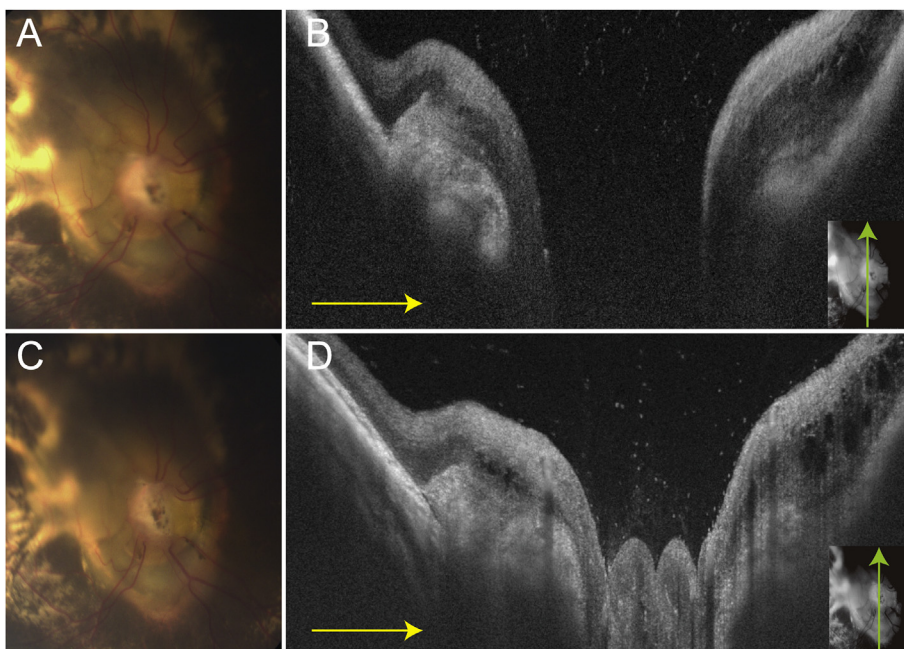


Fig. 2. Fundus photography and swept-source optical coherence tomography (SS-OCT) images of the left eye. (A) A fundus photograph of the optic nerve that is surrounded by excavation, with no contraction. Exudate around the optic nerve is prominent compared with the right eye. (B) SS-OCT images without contraction. Deep excavation is surrounded by bulging tissue under the retina. (C) A fundus photograph of the optic nerve with contraction. The change in the shape of the excavation is not apparent compared with that of the right eye. (D) SS-OCT images with contraction. The diameter of the excavation becomes smaller compared with no contraction as in the right eye. (B) and (D) The green arrows indicate the area of the SS-OCT images. The yellow arrow indicates the direction of the images. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

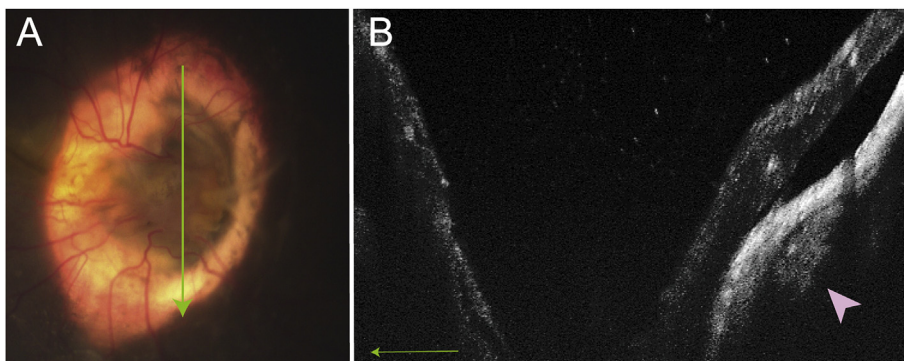


Fig. 3. Fundus photography and swept-source optical coherence tomography (SS-OCT) images of the right eye. (A) A fundus photograph with contraction. The green arrow indicates the area of the SS-OCT images. (B) SS-OCT image of the right eye. Hyperreflective tissue is seen under the sclera, which might be smooth muscle tissue (pink arrowhead). The green arrow indicates the direction of the images. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

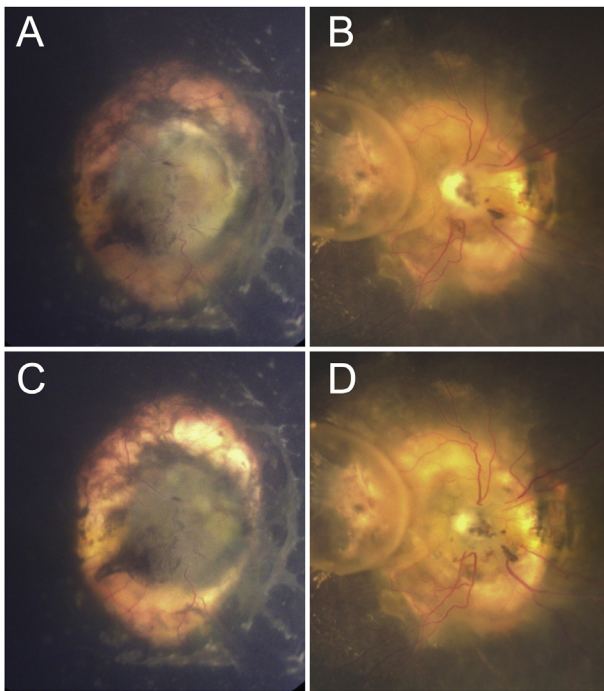


Fig. 4. Fundus photographs obtained during the second examination. (A) A fundus photograph of the right eye in which maximal contraction is seen. Compared with the first examination, the amounts of white fibrous tissue and exudates around the optic nerve have increased. (B) A fundus photograph of the right eye without contraction. The changes with contraction are not apparent compared with the first examination. The area of retinal depigmentation is smaller compared with contraction. (C) A fundus photograph of the left eye with maximal contraction. A retinal cyst is seen superonasal to the optic disc. (D) A fundus photograph of the left eye without contraction. The changes with contraction are more apparent compared with the first examination.

correlated with the respiratory cycle or pulse rate. Fluorescein angiography showed an avascular area in the peripheral retina in both eyes. Deep excavation around the optic discs in both eyes, the depth and width of which changed in association with the contractile changes (Figs. 1 and 2), was seen on the SS-OCT images. Furthermore, hyperreflective tissue was observed on the SS-OCT images beneath the sclera around the edge of the excavation in the right eye (Fig. 3, Supplemental Fig. 1).

At the age of 4 years and 1 month, the patient's best-corrected decimal visual acuity was 0.08 in the left eye but could not be measured in the right eye. Nystagmus and alternating esotropia remained. Because of developmental delays and difficulty performing examinations at the clinic, we again performed comprehensive ophthalmic examinations under general anesthesia at the age of 4 years and 8 months. Funduscopy clarified the absence of retinal detachments or breaks, but the white fibrous tissue on the optic disc and pigmentation around the optic disc became apparent in the right eye, a retinal cyst appeared superonasal to the optic disc in the left eye (Fig. 4), and contractility remained bilaterally. Bilateral simultaneous video recordings using two Retcam3 cameras were performed. The contractions did not occur synchronously in the right and left eyes, and there was no correlation between both eyes (Fig. 4, Supplementary Video).

Supplementary video related to this article can be found at <https://doi.org/10.1016/j.ajoc.2018.12.002>.

3. Discussion

In the current case report, we described the following detailed clinical findings of eyes with bilateral CPS: contractions under positive ventilation, contractions independent of light stimulus or forced eye lid

closure, unsynchronized and irregular contractions between both eyes, contractions under general anesthesia induced using a muscle relaxant, and the presence of sequential hyperreflective tissue under the sclera suggesting muscle tissue in the right eye.

Since 1966, when Wise et al.⁴ first reported CPS, several debates have ensued regarding the mechanism underlying the contractility associated with the disease. In any of the published reports, the numbers of the cases were limited, and no definite conclusions have been drawn until recently. In 1969, Sugar and Beckman⁵ initially reported the pressure balance theory and described synchronized contractions corresponding to the respiratory cycle associated with venous pressure changes. Pollock³ and other groups¹⁶ also supported the pressure balance theory in cases with CPS with a serous retinal detachment, in which the subretinal fluid flow communicated with the subarachnoid space or was between the vitreous/subretinal space and might induce contractions.^{3,16} However, as with the left eye of the current case and the case reported by Brodsky,⁸ a retinal detachment was not necessarily needed for contractions to occur and the respiration in the current case was supported by positive airway pressure. Moreover, the contractions were irregular and not synchronized with the respiratory cycle (Fig. 4, Supplementary Video). Thus, the pressure balance theory does not seem realistic.

As the evidence accumulated, the muscle contraction theory more likely explains the contractions that occur in eyes with optic disc anomalies. Font and Zimmerman¹³ and Willis et al.¹⁴ first reported circumferential smooth muscle fibers in congenital optic disc anomalies. These anomalies resulted from dysgenesis of the mesodermal differentiation around the optic nerve; extremely thin dura, hyperplastic adipose tissue and smooth muscle fibers were identified around the optic nerve, in which smooth muscle tissue was oriented in and under the sclera and choroid. Another report demonstrated the contractility and smooth muscle cells also were identified even in healthy choroid.¹⁵ In the current study, SS-OCT, which can visualize deeper tissue, enabled visualization of muscle-like hyperreflective tissue under the sclera, which was identical to the location reported by Willis et al.¹⁴

The most interesting aspect to consider is if the muscle contraction is regulated or unregulated, in other words, is the smooth muscle contraction controlled by an autonomic nerve or not? As discussed previously, smooth muscle cells were identified around an anomalous congenital optic nerve head, and no report has identified innervation to the smooth muscle cells around an anomalous optic nerve head of a congenital optic nerve head anomaly, except for one report that showed cholinergic nerve fibers co-localized with nonvascular smooth muscle cells in healthy choroid,¹⁵ which was speculated to be the origin of the contracting muscle in these optic nerve head diseases.¹² Wise et al. believed that the contraction of the muscle, which is known as the retractor bulbi muscle, was regulated through nervous system, because the contraction was induced by forced lid closure and light in the fellow eye.⁴ Kral and Svarc⁶ proposed that the contraction was regulated by an autonomic cholinergic nerve similar to the iris sphincter muscle, because contractions resulted from strong light stimulation of the fellow and affected eyes, which was supported by several reports.^{7,10,11}

Although the debate remains controversial,^{4,6–11} the muscle contraction might be unregulated based on the current findings. First, the patient was evaluated under general anesthesia, which eliminated any purposeful movements. Second, the contractions between the two eyes that were monitored simultaneously were not correlated or unsynchronized even under continuous light stimulation of the same intensity using the Retcam camera, which means light stimulation was not associated with the contractions. These facts indicated strongly that the muscle movement was involuntary and not regulated by any nerves, including the autonomic nervous system.

The current report was limited because the resolution of the SS-OCT images was low due to the deep staphyloma, and the young age of the patient made it impossible to perform some examinations. Further study is necessary to clarify the mechanism of contractions associated

with optic disc anomalies.

Patient consent and ethical approval

The Institutional Review Board of the National Center for Child Health and Development approved this retrospective observational case report, which adhered to the tenets of the Declaration of Helsinki. The mother of the current patient provided written informed consent for the ophthalmic examinations under general anesthesia and for publication of this report.

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All authors certify that they have no affiliations or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.ajoc.2018.12.002>.

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