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Contractile peripapillary staphyloma: OCTA documentation of increased peripapillary vessel density during transient visual loss episodes

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ABSTRACT

Purpose: to describe a patient with a contractile peripapillary staphyloma and transient visual loss (TVL) that underwent repeated OCTA examination documenting disc contraction and increased peripapillary vessel density as the mechanism of TVL.

Observations: a 28-year-old male presented multiple daily episodes of TVL for the last 5 years. Fundoscopic examination revealed a peripapillary staphyloma. The fundus photographs and SS-OCT demonstrated flattening of the posterior polo and crowding of the contracted optic disk, which became hyperemic with tortuous and dilated veins during visual loss episodes. OCTA showed temporary increased peripapillary vessel density, presumably from severe venous congestion leading to TVL during the contraction.

Conclusion and Importance: increased peripapillary vessel density can be demonstrated by OCTA during TVL in contractile peripapillary staphyloma. These findings indicate that severe venous stasis during disc contraction is the cause of TVL.

1. Introduction

Peripapillary staphyloma is a rare congenital ocular malformation in which the optic disc is located at the base of a funnel-shaped scleral deformation and thinning at the posterior pole of the eye.[1] While in most cases the condition is stable, contractile movements of the optic disc region may occur, similar to what has been described in optic disc coloboma and morning glory anomaly.[2,3] While a pressure imbalance between the intraocular and the cerebrospinal fluid has been suggested as an explanation for the peripapillary movements,[1] previous reports favor a muscle contraction as the responsible mechanism based on sequential disc photography or video recording[2,4] and a single report documenting optical coherence tomography (OCT) evidence of episodic disc deformity.[5] Optical coherence tomography angiography (OCTA) of contractile peripapillary staphylomas however, has not been previously reported.

Transient visual loss (TVL) may occur in peripapillary contractile staphylomas and other excavated disc anomalies,[2,3] but the exact mechanism of temporary monocular blindness is unclear. We have examined a patient that had repeated episodes of monocular visual loss as the sole presenting symptom of a peripapillary staphyloma. Sequential evaluations using high-resolution sweep-source (SS) OCT and OCTA at different examinations with or without visual loss were performed and indicated that disc deformity, peripapillary contraction, increased vessel density, and retinal venous congestion are the most likely mechanisms to explain TVL in such condition.

2. Case report

A 28-year-old previously healthy man complained of ten to twenty daily episodes over the last five years of acute TVL in the left eye (OS) lasting around 30–40 seconds followed by complete visual recovery. The TVL episodes were described as acute events of blurred vision in the whole field of vision with slight color vision desaturation, but without any localized scotomas or positive visual phenomena. The patient was a physician and had carefully observed that episodes of monocular blindness did not have any specific trigger; in particular, he denied any relationship with breathing movements, the Valsalva maneuver, accommodation, illumination of the other eye, exercise, head tilt movement, or change in body position. There was no history of neurological diseases or use of medications. Several previous ophthalmologic, clinical, and neurologic evaluations failed to establish a cause for his TVL

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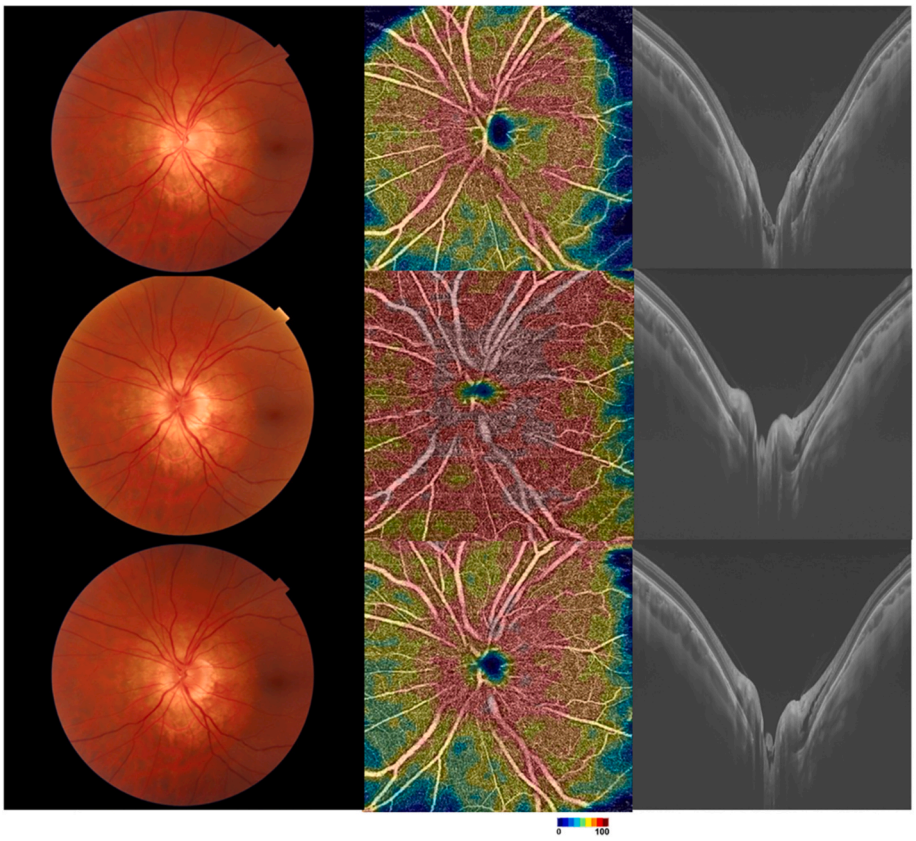

Fig. 1. Fundus photographs (left), OCTA (middle), and line B-scan of the optic disc (right) in an eye with peripapillary staphyloma. Upper row: at a resting period without visual complaint. Middle: at a contracted period, during a visual loss episode. Notice the reduction of the disc diameter with cupping, hyperemia of the disc, increased peripapillary vessel density and modification of the disc contour. Bottom: intermediate stage of contraction without the visual loss.

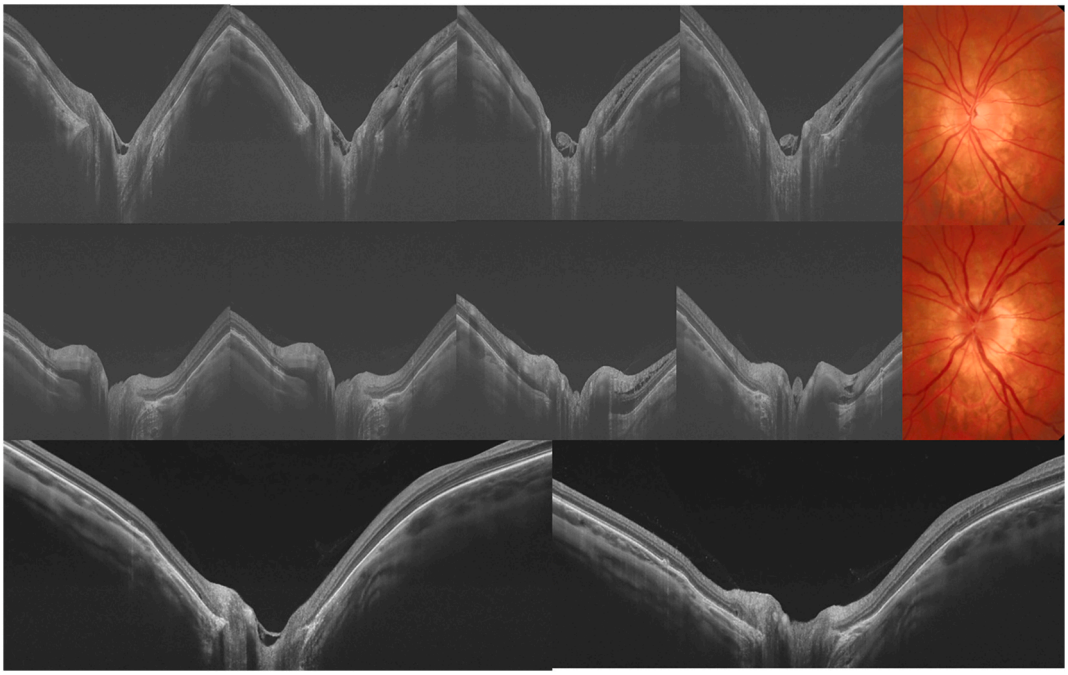

Fig. 2. Radial line B-scan and fundus photographs of the optic disk in an eye with peripapillary staphyloma. Upper row: radial line scans of the non-contracted optic disc. Middle row: contracted peripapillary staphyloma showing anterior displacement of the posterior sclera on OCT line scans and reduced optic nerve diameter with venous dilation in the fundus photograph. Notice also the enhanced cystic changes in the inner retina. Bottom row: OCT images including the optic disc and fovea in the same linear scan without contraction (left) and with contraction (right) showing cystic changes in the outer plexiform layer in the fovea.

episodes. Magnetic resonance imaging of the brain and orbit were normal except for a posterior bulging of sclera in OS.

On examination, the best corrected visual acuity was 20/20 with a small astigmatism (-.50 cylinder) correction in both eyes. Extra ocular movements, pupillary reflexes, biomicroscopy, intraocular pressure measurements, and standard automated perimetry were unremarkable. Funduscopic examination was completely normal in the right eye (OD) and showed only a 360-degree peripapillary pigmentary abnormality in OS (Fig. 1), with posterior bulging of the disc compatible with a peripapillary staphyloma.

Fundus photographs, SS-OCT, and OCTA were performed on different occasions, with or without TVL episodes. Three patterns could be observed and are exemplified in Fig. 1. First, a baseline, "resting" state when the optic disc had a central cup and a peripapillary pigmented abnormality, normal peripapillary vessel density on OCTA and a funnel-shaped aspect of the posterior pole on OCT with the disc at its tip (Fig. 1 upper row). Second, a fully contracted status associated with TVL when the optic disc became small and hyperemic with some vein engorgement, with increased vessel density on OCTA and clear modification of the posterior pole deformation with relative flattening of the peripapillary area on OCT (Fig. 1, middle row). Third, an intermediate state of some disc contraction and scleral deformity but without significant modification of vessel density compared to the baseline (Fig. 1, lower row). There was no fluid shift in the subretinal space in any examining condition.

SS-OCT radial scans passing through the optic disk demonstrated detailed morphological changes during the contraction depicted in Fig. 2. Compared with the "resting" state (Fig. 2, upper row) the SS-OCT showed crowding and flattening of the optic disk, which made some cystic spaces in the inner retina more visible while contracted (Fig. 2, middle row). In a wide scan including the optic disk and macula it is possible to note that, during the contraction, the cystic changes also occur in the fovea (Fig. 2, lower row), probably due to the reduction of the axial diameter which caused loosening of the retina and enhancement of the cystic spaces.

3. Discussion

Initially, our case is interesting because it documents TVL as an unusual presentation of peripapillary staphyloma in a 28-year patient without any history of previous ocular disease. Although TVL has been reported in several cases of excavated disc anomalies (particularly, optic disc coloboma and the morning glory anomaly), usually the subject is aware of the anomaly in the affected eye. Since our patient had normal visual acuity and was unaware of the anomaly, he underwent an extensive investigation of the causes of TVL. Despite the presence of abnormal pigmentation around the optic disc, the diagnosis was initially missed by several physicians, which is partially explained by the fact that the posterior optic disc displacement was not as severe as in other cases of the anomaly. High-resolution SS-OCT was helpful in confirming the suspected clinical diagnosis of posterior staphyloma by documenting a deformation of the eye around the optic disc, which was located at the bottom of a funnel-shaped posterior sclera position. Our findings are in agreement with previous reports that showed, on OCT examination, a deep excavation involving the optic disk with preserved peripapillary retinal layers around the posterior staphyloma and a steep slope to the bottom of the optic disc.[5–7]

Our case is also interesting because repeated examinations using disc photographs, SS-OCT, and OCTA were possible both during episodes of visual loss as well as when the patient was asymptomatic. Such repeated testing made it possible to analyze disc appearance, peripapillary vessel density, and the contour of the posterior pole (using photographs, OCTA, and OCT) at different times (resting or contracted) with or without visual loss. In the contracted stage, SS-OCT demonstrated a crowding of the optic nerve, which became hyperemic with reduced cupping and dilated retinal veins, particularly during TVL episodes. Also, in the contracted state, OCT showed flattening of the posterior pole and anterior displacement of the distal optic disc when compared to the non-contracted state (Figs. 1 and 2), similar to what has recently been reported by Yoshida et al.[5] Previous studies have suggested that such optic nerve anomaly is associated with intrascleral and choroidal smooth fibers oriented concentrically around the distal optic disk,[8] possibly a primitive ciliary body located on the posterior sclera that is somehow responsible for the episodic contraction of the optic disc. In our case, however, despite multiple examinations and careful questioning of the patient, we could not identify any factor that might induce contraction or TVL. More specifically, we were unable to document disc contraction synchronous with the respiratory cycle or induced by light as described in other studies.[4,9] The reasons why in our case none of these triggering factors were present are unclear.

Finally, our case is unique because we were able to perform repeated high-quality OCTA and simultaneous photographic recording of the optic disc and peripapillary, both in the contracted and non-contracted disc positions with or without TVL. Disc size reduction, associated with hyperemia and vein engorgement (presumably by heterotopic muscle fiber contraction) could be graded and were most intense when TVL was present (Fig. 1). OCTA showed clear increases in peripapillary vessel density in the contracted positions when associated with visual loss, likely pointing to venous congestion and blood stasis with slowing of the retinal circulation as the cause of the TVL, which was reversible when the contraction ceased. Therefore, the mechanism of TVL in our case seems similar to what has been found in patients with impending central retinal vein occlusion from hematologic disorders when isolated retinal venous engorgement was the sole finding in patients with TVL.[10,11] It is important to emphasize that high-quality OCTA images were possible in our case probably because the disc displacement was not as severe as in other cases of peripapillary staphylomas, when adequate imaging of the disc is not feasible at least with the current technology.

4. Conclusion

SS-OCT is useful to confirm the presence of posterior staphyloma of the optic disk excluding other congenital abnormalities and to demonstrate the structural changes that occur during the contraction of the staphyloma. OCTA helped to detect increased vessel density as the staphyloma contracted, throwing light on the pathophysiology of TVL and suggesting temporary venous stasis of the optic disc.

Patient consent

The patient consented to publication of the case in writing.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The following authors have no financial disclosures: K. H., L.D.C., M. L.R.M.

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References

1. Wise JB, MacLean AL, Gass JD. Contractile peripapillary staphyloma. *Arch Ophthalmol*. 1966;75(5):626–630.
2. Brodsky MC. Contractile morning glory disc causing transient monocular blindness in a child. *Arch Ophthalmol*. 2006;124(8):1199–1201.

3. Graether JM. Transient amaurosis in one eye with simultaneous dilatation of retinal veins. In association with a congenital anomaly of the optic nerve head. *Arch Ophthalmol*. 1963;70:342–345.
4. Farah ME, Uno F, Bonomo PP, Nobrega M, Hofling-Lima AL. Contractile peripapillary staphyloma with light stimulus to the contralateral eye. *Arch Ophthalmol*. 2001;119(8):1216–1217.
5. Yoshida T, Katagiri S, Yokoi T, Nishina S, Azuma N. Optical coherence tomography and video recording of a case of bilateral contractile peripapillary staphyloma. *Am J Ophthalmol Case Rep*. 2019;13:66–69.
6. Lee KM, Woo SJ, Hwang JM. Evaluation of congenital excavated optic disc anomalies with spectral-domain and swept-source optical coherence tomography. *Graefes Arch Clin Exp Ophthalmol*. 2014;252(11):1853–1860.
7. Woo SJ, Hwang JM. Spectral-domain optical coherence tomography of peripapillary staphyloma. *Graefes Arch Clin Exp Ophthalmol*. 2009;247(11):1573–1574.
8. Poukens V, Glasgow BJ, Demer JL. Nonvascular contractile cells in sclera and choroid of humans and monkeys. *Invest Ophthalmol Vis Sci*. 1998;39(10):1765–1774.
9. Sugar HS, Beckman H. Peripapillary staphyloma with respiratory pulsation. *Am J Ophthalmol*. 1969;68(5):895–897.
10. Biousse V, Newman NJ, Sternberg Jr P. Retinal vein occlusion and transient monocular visual loss associated with hyperhomocystinemia. *Am J Ophthalmol*. 1997;124(2):257–260.
11. Shuler Jr RK, Biousse V, Newman NJ. Transient monocular visual loss in two patients with impending central retinal vein occlusion. *J Neuro Ophthalmol*. 2005;25(2):152–154.