Bilateral peripapillary staphyloma: a case report

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Case Report

Keywords: Staphyloma; optic nerve; congenital

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Abstract

Background: The peripapillary staphyloma (PS) is a rare non-hereditary congenital optic disc dysplasia, which is generally unilateral and is likely to occur with poor vision. Fundus uncovered a large deeply excavated optic nerve head, retinal pigment epithelium changes at its edges and normal disc and retinal blood vessels in size and features.

Case presentation: This literature present a bilateral case of an especially congential peripaillary staphyloma (CPS), which the patient with esotropia and nystagmus was exposed to abnormality when she was only 5-month-old. Several interesting features are present in this patient.

Conclusions: Usually they have a large extent on visual loss. Thus, although extreme rare, an early detection, treatment and follow ups are necessary.

Keywords: Staphyloma; optic nerve; congenital

Background

A CPS was reported to be a rare type of posterior staphylomas, and its incidence was approximately 1.5% of all the staphyloma[1]. Fundus uncovered a large deeply excavated optic nerve head, retinal pigment epithelium changes at its edges and normal disc and retinal blood vessels in size and features. It is generally unilateral. Now, we report a bilateral case of an especially CPS, as follows:

Case Presentation

A 5-month-old infant presented for an optometric examination with esotropia and nystagmus. This girl was the healthy product of a normal gestation and was experiencing normal development general systemic examination by her pediatrician disclosed no systemic abnormalities. Anterior segment, lens, ocular motility and intraocular pressure were normal. But in her eyes, there was a deep excavation with a normal-appearing optic nerve at the base (figure 1), which the retinal pigment epithelium changes at the head of excavation. And the staphyloma in the right eye was judged to be shallower than that in the left eye. Moreover, the macular structural change was observed in the left eye because of a large range of depression. Ultrasonography showed a deep excavation at the optic nerve head with walls slanting outwards, and the left eye was large that the right eye (figure2). The diagnosis was: CPS in both eyes.

Discussion

The PS is a rare non-hereditary congenital optic disc dysplasia, which belongs to the category of congenital dysplasia. It is generally unilateral and is likely to occur with poor vision. Compared to other excavated optic disc anomalies, PS is known to be rarely accompanied by other congenital defects or systemic diseases. PS has a normal sized optic disc that is located at the bottom of a depression. On the edges of the excavation, there are extensive retinal pigment, epithelial and choroidal pigmentary atroply.

And the retinal vasculature is normal. Several interesting features are present in this patient. The patient was exposed to abnormality when she was only 5-month-old supporting the diagnostic citeria of CPS[2].

The formation of posterior scleral staphyloma is due to abnormal thinning and expansion of the sclera. The posterior pole of the eyeball is limited to outward bulging, which the curvature radius formed by the protruding part is smaller than the radius of curvature of the surrounding eyeball wall. This characteristic change is common in high myopia. Ishida [3] et al. found that 1.3% of highiy myopic eyes with posterior scleral staphyloma have choroidal atrophy at the edge of the staphyloma. Ohno-Matsui[4] and other studies found that 50.5% of high myopia showed posterior scleral staphyloma, and 45.5% of posterior scleral staphyloma involving the optic disc. They both tend to involve optic discs as well as macula region. However, the posterior scleral staphyloma caused by high myopia is relatively older, less depressed, and usually occurs in both eyes.

Usually they have a large extent on visual loss, although bilateral cases with normal visual acuity was reported. It is related to the range of the depression and whether it was associated with the accumulation of macula. Kim [5]et al described the largest case series, including 19 patients (21 eyes) with PS. Two patients (11%) had bilateral involvement and 7 eyes had severe myopia of more than 6 diopters. Most scholars agree that eyes with PS may improve eyesight to a limited extent by occulsion therapy[5]. Several reports have been published on CPS. Light stimulus to the contralateral eye might provoke contration of the PS. But no contraction was observed that reported in 2005 by Kim and colleagues in a series of patients with PS[5]. Thus, a large number of cases are needed for verification. Although extreme rare, the PS may cause severe visual impairment and an early detection, treatment and follow ups are necessary.

Abbreviations

PS: peripapillary staphyloma

CPS: congential peripapillary staphyloma

Declarations

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No funding was obtained for this study.

Availability of data and materials

The datasets during and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors’ contributions

HCG and CC were responsible for collection of date. HCG and MW performed the analysis. HCG, MW, YYS and CC were responsible for interpretation of results. HCG wrote the first draft of the manuscript. All authors reviewed and approved the final manuscript.

Competing interests

The authors declare that they have no competing interests.

Consent for publication

Written informed consent was obtained from the patient's father for publication of this Case report. I am sure it is the father of the patient.

Ethics approval and consent to participate

Not applicable.

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Figures

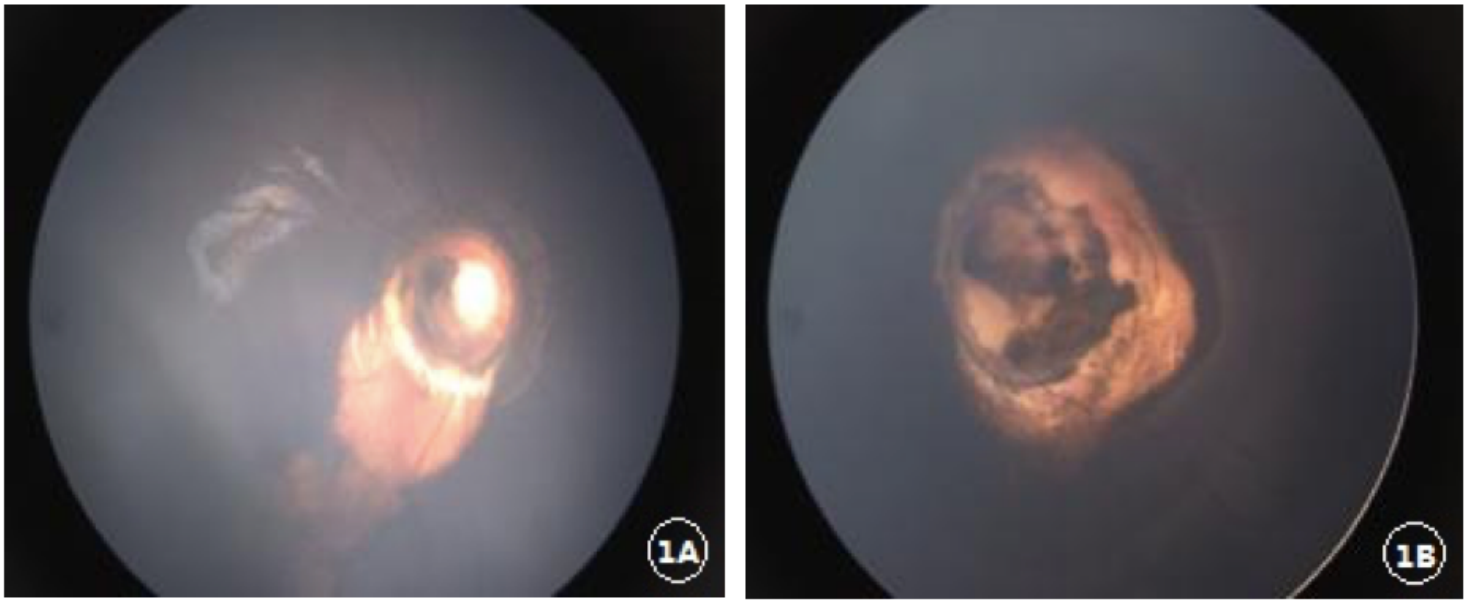

Figure 1

Fundus photograph shows a deep excavation with a normal-appearing optic nerve at the base, and the staphyloma in the right eye(1A) is shallower than that in the left eye(1B).

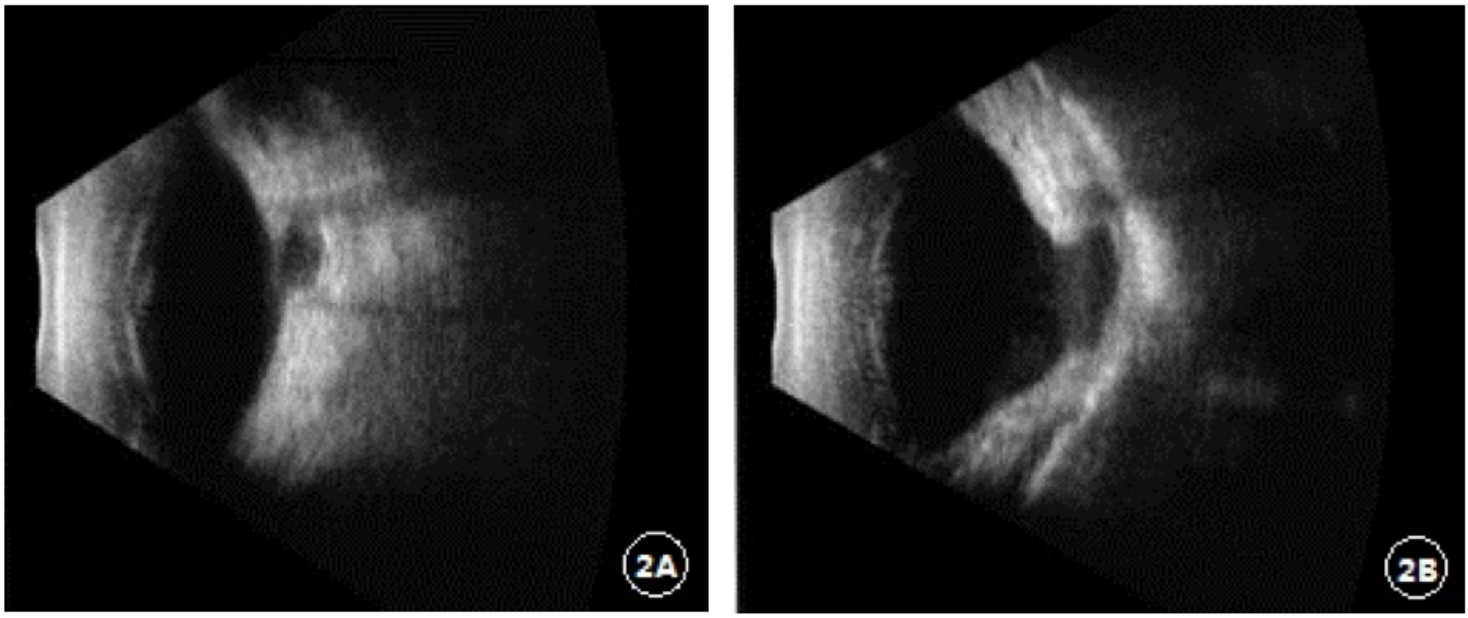

Figure 2

Ultrasonography of both eyes shows a deep excavation at the optic nerve head with walls slanting outwards, and the left eye(2B) is large that the right eye(2A).

Supplementary Files

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- supplement1.pdf