American Journal of Ophthalmology Case Reports 26 (2022) 101468

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

Retinal detachment and retinoschisis associated with optic disc pit in peripapillary staphyloma

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ARTICLE INFO

Keywords:
Peripapillary staphyloma
Optic disc pit
Laminar cribrosa defect
Subarachinoid space
Retinal detachment

ABSTRACT

Purpose: To determine the characteristics and clinical course of an eye with a peripapillary staphyloma and an optic disc pit associated with a retinal detachment (RD) and retinoschisis.

Observations: A 44-year-old woman had a peripapillary staphyloma with a shallow RD and retinoschisis in her right eye. The optic disc was located in the peripapillary staphyloma with steep walls, and an optic disc pit was located on the temporal area of the optic disc cup. Optical coherence tomography (OCT) revealed a separation of the lamina cribrosa, herniated retinal tissue, and a subarachnoid space corresponding to the optic disc pit. Pars plana vitrectomy with laser photocoagulation around the temporal margin of the peripapillary staphyloma was performed to treat the RD, and the treatment was successful.

Conclusion and importance: Clinicians should be aware that eyes with a RD and retinoschisis associated with a deep peripapillary staphyloma and an optic disc pit exist. The RD and retinoschisis can be successfully treated by vitrectomy.

1. Introduction

There are rare cases of congenital excavated optic disc anomalies which are recognized as belonging to the same spectrum of optic disc disorders. A peripapillary staphyloma, optic disc pit, morning glory disc anomaly, and optic disc colobomas are included in this spectrum of optic disc disorders.[1] The excavated disc anomalies occasionally occur together in the same eye, e.g., an optic disc pit and a coloboma,[2] but to the best of our knowledge, there has not been a report on the co-existence of a peripapillary staphyloma and an optic disc pit in the same eye. A peripapillary staphyloma is a unilateral deep excavation around a normal optic disc.[1,3] A retinal detachment (RD) around the optic disc is occasionally observed in eyes with a peripapillary staphyloma.[4,5] Another laboratory group and our group have described the OCT images of a retinal tear in a peripapillary staphyloma which caused a RD around it.[4,5]

An optic disc pit typically appears as single grayish round depression on the optic disc.[3] Histopathologically, the optic disc pit is seen as a herniation of retinal tissue into an excavation that can stretch into the subarachnoid space through a defect in the lamina cribrosa.[1,3] Eyes with an optic disc pit occasionally also have a serous RD and/or retinoschisis in the macular area, and it is then called an optic disc pit maculopathy.[6,7]

We present our findings in an eye with a peripapillary staphyloma and an optic disc pit associated with a RD and retinoschisis. The eye was treated successfully by vitrectomy.

2. Case report

A 44-year-old woman noticed a slight reduction in the vision in her right eye five years earlier and went to a local clinic. Macular retinoschisis was detected, and it was connected to a peripapillary staphyloma. The decimal best-corrected visual acuity (BCVA) of her right eye was 0.5, and the retinoschisis was followed without treatment at the local clinic. Her right visual acuity had decreased over the past two months due to the development of a foveal retinal detachment (RD), and she was referred to our hospital for treatment.

At our initial examination, her decimal BCVA was 0.2 in her right eye and 1.2 in her left eye. The intraocular pressure was 17 mmHg in both eyes, and the ocular axial length was 24.3 mm in both eyes. The anterior segment was completely normal in both eyes. She had no family history of systemic medical problems.

Our fundus examination revealed a peripapillary staphyloma, and the optic disc was large with a whitish round abnormality which

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https://doi.org/10.1016/j.ajoc.2022.101468
Received 30 October 2021; Received in revised form 1 March 2022; Accepted 2 March 2022
Available online 4 March 2022

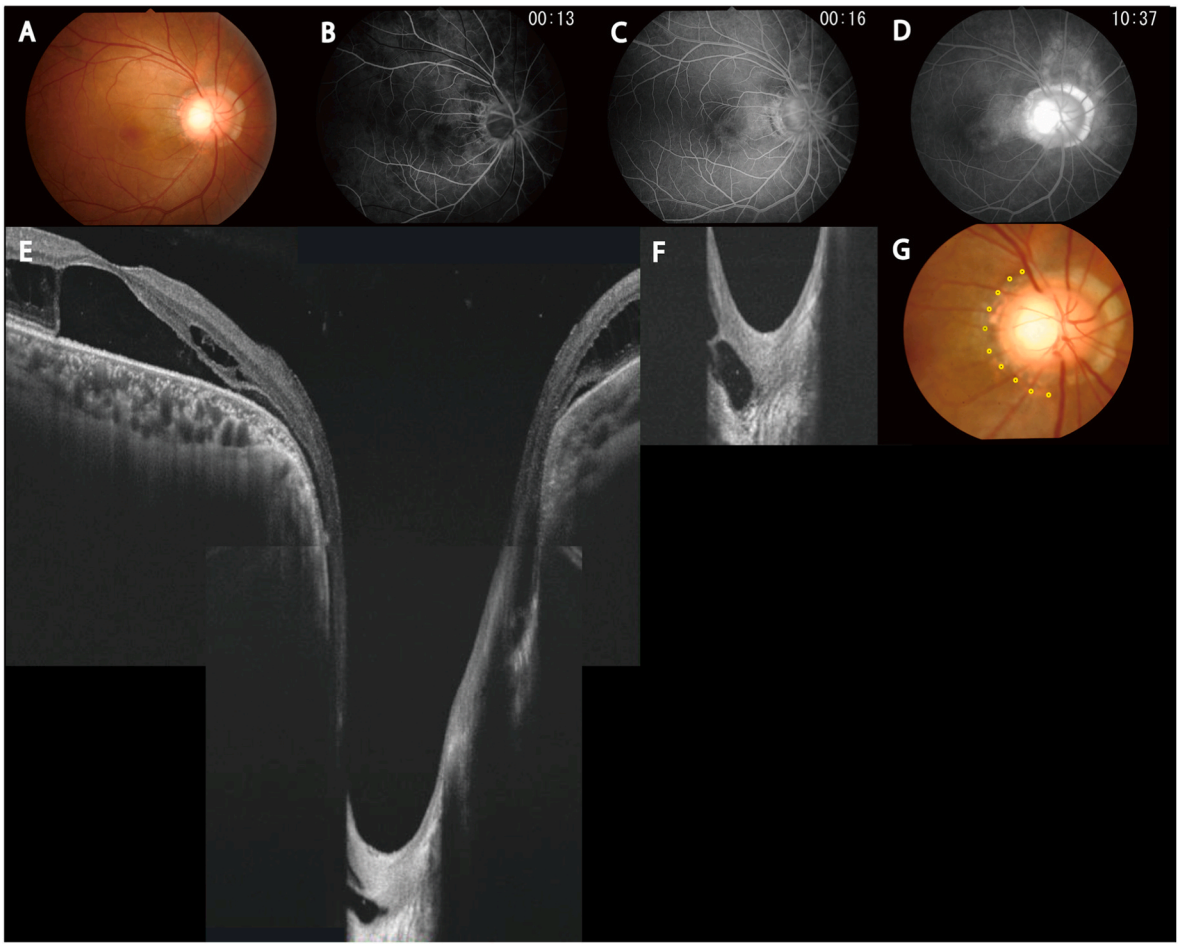

Fig. 1. Fundus photograph of the left eye of a 44-year-old woman with a peripapillary staphyloma and a retinoschisis in her right eye at the initial visit. The sites of a laser photocoagulation can be seen.
A: Fundus photograph showing a peripapillary staphyloma and abnormal appearing optic disc. The disc is large and has an optic disc pit in the temporal area of the optic disc cup. A thin retinal detachment (RD) can be seen extending from the peripapillary staphyloma to the macula area..B ~ D: Fluorescein angiogram of the optic disc pit on the temporal side of the optic disc cup. This area is hypofluorescent in the arterial phase (B) and hyperfluorescent in the venous phase (C). Leakage is observed and area of the RD and it is slightly hyperfluorescent in the late phase (D). E: Optical coherence tomographic (OCT) image shows deep peripapillary staphyloma. The RD can be seen to extend from the middle wall of the staphyloma to the macula area. F: OCT image of the bottom of the optic disc. A white excavated abnormality showing the herniated retina with highly reflective horizontal ridge-like protrusion can be seen. A hypo-reflective space containing hyperreflective granular and dot structures suggestive of the subarachnoid space can be seen just posterior to the bottom of the pit. A hyperreflective horizontal protruded structure is present posterior to the subarachnoid space. The lamina cribrosa is separated from the temporal sclera and it and the retrolaminar optic nerve fiber bundles appear to be shifted away from the optic disc pit. G: Laser photocoagulation was performed around the staphyloma edge in the temporal area.

resembled an optic disc pit in the temporal area of the optic disc of her right eye. A shallow RD was observed extending from the edge of the peripapillary staphyloma to the macular area and to the upper area of the optic disc cup (Fig. 1A). The margin of the optic disc was clearly detected, and a coloboma and morning glory disc anomaly were not present. In addition, there was no prepapillary membrane on the optic disc, and the blood vessels ran normally on the optic disc.

Fluorescein angiography (FA) showed that the optic disc pit-like anomaly was hypofluorescent in the arterial phase (Fig. 1B) and hyperfluorescent in the venous phase (Fig. 1C). There was fluorescein leakage from the abnormality in the late phase, and the area of the RD was slightly hyperfluorescent in the late phase of FA (Fig. 1D). Her left eye had no abnormalities in the FA images.

Swept-source OCT showed a deep and steep peripapillary staphyloma (Fig. 1E). A retinoschisis was observed in the macular area. A RD was detected mainly in the macular area, and it was connected to a thin retina within the staphyloma in the OCT images. A posterior vitreous detachment (PVD) was not present. The OCT images did not show the staphyloma wall from the middle to the deeper area due to a shadow cast by the steep and deep staphyloma. No retinal tear was observed within the staphyloma. The OCT image of the bottom of the optic disc showed the herniated retina as a slightly highly reflective horizontal ridge-like protrusion corresponding to the optic disc pit-like abnormality. A hypo-reflective space contained hyperreflective granules, and dot structures were seen just posterior to it. The findings of this cavity were similar to what Ohno-Matsui et al. described as a subarachnoid space in eyes with an optic disc pit.[6,8] A hyperreflective horizontal protruding structure was present posterior to this subarachnoid space. The lamina cribrosa was separated from the temporal sclera, and it and the retrolaminar optic nerve fiber bundles appeared to have shifted away from the defect in the lamina cribrosa (Fig. 1F).

Based on the findings of the color fundus photographs and FA and OCT images, we identified the whitish round optic disc pit-like abnormality as a large optic disc pit with subarachnoid space that had moved through the lamina cribrosa defect. We then diagnosed the eye with a RD and retinoschisis related to the peripapillary staphyloma with an optic disc pit.

Pars plana vitrectomy with a 27-gauge system was performed to treat the RD in the right eye. A PVD was not detected during vitrectomy, and it was created. The adhesion between the vitreous and retina and optic

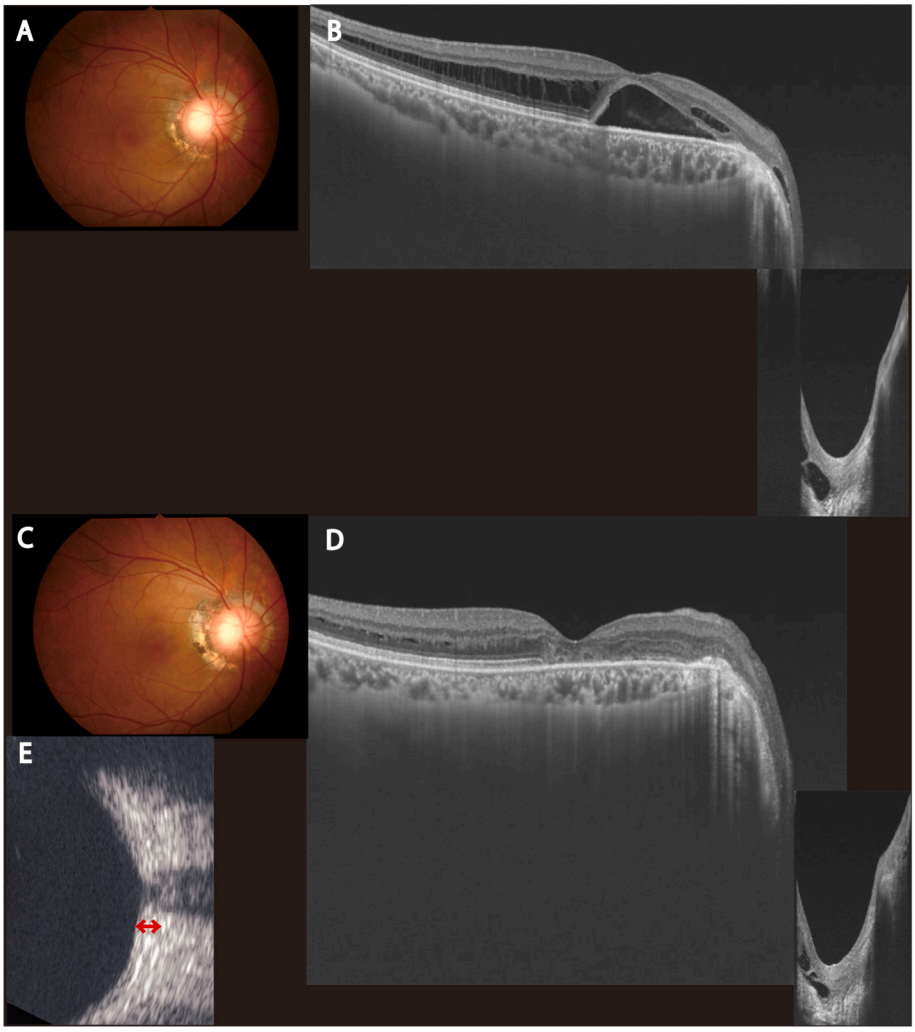

Fig. 2. Postoperative fundus findings in the same patient.
A: Color fundus photograph taken one month after the vitrectomy showing a thin RD which is similar to the preoperative finding. The laser scar can be seen at the temporal edge of the staphyloma. B: OCT image one month after the vitrectomy. The RD and retinoschisis remain in the area of the macula and staphyloma but a small hyperreflective abnormality can be seen as a scar of the laser photocoagulation at the edge of staphyloma. The scar separates the RDs of the macular area and staphyloma area. The hyporeflective subarachnoid space is also present. C: Color fundus photograph 11 months after the vitrectomy. The RD is resolved and laser photocoagulation scar around temporal area of optic disc can be seen as a slightly pigmented area of the retina. D: OCT image 11 months after the vitrectomy. The RD and retinoschisis are not present in the OCT images. A small hyperreflective laser photocoagulation scar is seen at the edge of staphyloma. The subarachnoid space also remains. E. Postoperative ultrasound image 17 months after the vitrectomy. A two-way arrow shows a peripapillary staphyloma. The depth of it is about 19 mm. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

disc was not tight. We searched for retinal tears but none was detected. Laser photocoagulation was performed around the temporal staphyloma edge before fluid air exchange was performed (Fig. 1G). Then, 20% sulfur hexafluoride (SF_6) gas was used to tamponade the retina.

One month after the surgery, the scar from the laser photocoagulation at the edge of staphyloma was observed as a hyperreflective retinal pigmented epithelium proliferation in the OCT images, and it was separated by the subretinal fluid from the macular area and staphyloma area (Fig. 2B). The RD in the peripapillary staphyloma was not present three months after the surgery, and the macular detachment had completely disappeared ten months after the surgery. The decimal BCVA improved to 0.7, and the retinoschisis was almost completely absent 11 months after the surgery (Fig. 2D). The optic disc pit and the subarachnoid space at the bottom of the optic disc were not altered. (Fig. 2B and D). The decimal BCVA at the final examination was 0.7 at 17 months after the surgery. At that time, we performed ultrasonography to measure the depth of staphyloma. It was about 19 mm (Fig. 2E).

3. Discussion

We have presented our findings in a rare case with a RD and retinoschisis associated with a peripapilary staphyloma combined with an optic disc pit. The OCT images showed a subarachnoid space under the optic disc pit. We suggest that the subarachnoid space was created by the movement of tissue through a defect of the lamina cribrosa of the optic disc. Vitrectomy led to a successful reattachment of retina. It is known that relieving the traction of the vitreous over the optic disc pit can contribute to the resolution of an optic disc pit maculopathy in most cases. However, the exact mechanism that caused the optic disc pit maculopathy has not been determined.[3] When we decided to treat this eye with vitrectomy, we were concerned that the retinal reattachment might be more difficult because the subretinal fluid connected to the optic disc is a risk factor for failure of reattachment of macular detachment as we reported.[9] Thus, we could not completely eliminate the possibility of an undetectable retinal tear in the peripapillary staphyloma. So, we not only created a PVD but also added laser photocoagulation around the edges of the staphyloma to create a barrier between the retina in peripapillary staphyloma and the macular area.

The scar of the laser photocoagulation played a major role by acting as a barrier that prevented subretinal fluid from entering from the staphyloma into macular area, but the subretinal fluid in the staphyloma area disappeared faster than that of the macular area. This suggests that creating a PVD would be more effective in resolving this retinal detachment.

This was a rare case but it is only one case. It will be necessary to observe and analyze more cases with a RD from peripapillary staphyloma in detail to clarify their relationships and to determine the best treatment for eyes with similar abnormalities.

Patients consent

Written consent to publish about details of the case and photographs

was obtained from the patient.

Funding

No funding nor grant support.

Authorship

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

Declaration of competing interest

The following authors have no financial disclosures: Kana Okano.

Tomoka Ishida received lecture fee from NIDIK Co., Ltd and Santen Pharmaceutical Co., Ltd.

Makoto Inoue received grants from Alcon lab and lecture fee from Santen Pharmaceutical Co., Ltd, Bayer Yakuhin, Ltd, Novartis Phama K. K., SENJU Pharmaceutical Co., Ltd., Alcon Japan Ltd., Carl Zeiss Meditec.

Akito Hirakata received grants from Santen Pharmaceutical Co.,Ltd, Bayer Yakuhin, Ltd, Novartis Phama K.K., HOYA Corporation, Pfizer Japan Inc., Kowa Pharmaceutical Co., Ltd., SENJU Pharmaceutical Co., Ltd., Alcon Japan Ltd., Otsuka Pharmaceutical Co., Ltd., NIDEK Co., Ltd., Chuo Sangyio Co., RE Medical Inc., IWASAKI OPTICAL. Co., Ltd., AMO Japan K.K., Ogura Inc., TSUMURA&Co., TOMEY CORPORATION, WAKAMOTO Pharmaceutical Co., Ltd., NIHON TENGANYAKU KEN-KYUSYO Co., Ltd., HEIWA IYOUSHOUKAI Co., Ltd., and he received lecture fee from Santen Pharmaceutical Co., Ltd., Bayer Yakuhin, Ltd, Novartis Phama K.K., HOYA Corporation, Pfizer Japan Inc., Kowa Pharmaceutical Co., Ltd., SENJU Pharmaceutical Co., Ltd., Alcon Japan Ltd., Otsuka Pharmaceutical Co., Ltd., NIDEK Co., Ltd., Chuo Sangyio Co.

Acknowledgments

The authors thank Professor Emeritus Duco Hamasaki of the Bascom Palmer Eye Institute of the University of Miami, Florida for his critical discussion and final manuscript revision.

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