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Lens-Sparing Vitrectomy Effective for Reattachment of Newly Developed Falciform Retinal Detachment in a Patient With Norrie Disease

Norrie disease (ND) is an inherited eye disease caused by mutations in the Norrie disease protein gene,1 and it is characterized by congenital blindness due to malformations of the retina.2 Patients are rarely seen before they develop a retinal detachment (RD), and surgical intervention usually fails because of the longstanding RD. We describe a case of ND in which the clinical course and angiographic findings were examined before the development of an RD. A falciform RD developed and lens-sparing vitrectomy reattached the retina.

Report of a Case. A 4-month-old boy was referred because of leukokoria in his right eye. Informed consent was obtained from his parents to perform genetic analyses, and a single base-pair substitution (c.53 T to A) was detected.3 Slitlamp examination showed a shallow anterior chamber, a clear lens, and a retrolenticular mass in the right eye. Fundus examination showed preretinal and vitreous hemorrhages located between the normal-appearing and dark retina. B-scan ultrasonography showed a pseudoglioma in the right eye.

Fluorescein angiography with a wide-field digital fundus camera (RetCam; Massie Research Laboratories, Inc, Dublin, California) showed that the peripheral dark area was avascular with leakage from the new vessels. The macula was estimated to be at the border of the vascular and avascular retina; however, this region was obscured by the lack of an avascular zone and macular pigment. Confluent photocoagulation with a 532-nm laser was applied to the peripheral avascular retina.

Five months later, a falciform RD, ie, vascularized retinal fold that extends from the temporal margin of the disc and passes across and obscures the macular region with contraction of fibrovascular proliferations (FVPs), had developed (Figure, A). Fluorescein angiography showed fluorescein leakage from the FVPs and nonablated avascular retina located at the nasal peripheral area that had not been observed at the first examination owing to preretinal and vitreous hemorrhages (Figure, B).

Because the retinal fold could lead to a total RD and phthisis bulbi, lens-sparing vitrectomy,5 ie, vitrectomy without removal of the crystalline lens, was performed. After conjunctival peritomy, a 3-port vitrectomy commonly used for retinopathy of prematurity in our insti-
tute\(^6\) was performed. Three sclerotomy sites were made 1 mm from the limbus with a 23-gauge microvitreoretinal blade, and a 23-gauge vitreous cutter, a light pipe (Alcon Laboratories, Inc, Fort Worth, Texas), and a self-retaining 23-gauge infusion tube (Dutch Ophthalmic Research Center International, Zuidland, the Netherlands) with the inner port length of 3 mm were used. The vitreoretinal traction between the FVPs and the peripheral retina and ciliary body was removed. Because the temporal peripheral avascular retina had laser burns, it could be distinguished from the overlying vitreous or FVPs. The posterior hyaloid was then separated from the posterior retina up to the border of the vascular and avascular retina. The dissection of the FVPs that had dragged the retina was performed mainly in a radial direction with 23-gauge scissors (Dutch Ophthalmic Research Center International) to release the circumferential traction. Extensive membrane dissection and delamination were not performed to avoid creating iatrogenic retinal tears. Finally, the avascular retina was ablated with a 532-nm laser.

One month after the surgery, the posterior retina was completely reattached despite residual tractional tension. The status of the retina and the clarity of the lens remained unchanged at the 11-month postoperative examination (Figure, C). The uncorrected visual acuity measured by the Teller Acuity Card was 0.1 OS.

Comment. Laser photocoagulation was performed as the initial treatment, but the disease continued to progress. This may be owing to an incomplete ablation to the avascular retina because of preretinal and vitreous hemorrhages. During the follow-up period, a falciform RD developed. In general, a long-standing falciform RD is difficult to unfold by surgery because FVPs are thick and hard and are firmly attached to the detached retina. However, the adherence between the FVPs and the surrounding tissues was weak in our case, and surgery was effective in unfolding and reattaching the retina.

In conclusion, we have followed up a patient with ND whose left eye developed a falciform RD even after photocoagulation. The falciform RD was managed successfully by lens-sparing vitrectomy. We recommend a careful follow-up of patients with ND especially at younger ages because early surgical intervention can lead to a better visual outcome.

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