TENT-SHAPED RETINAL DETACHMENTS IN RETINOPATHY OF PREMATURITY

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Background: Most retinal detachments associated with retinopathy of prematurity (ROP) are radial, segmental, or circumferential with cicatricial extraretinal fibrovascular proliferation (EFP) at the apex of the detachment. This report describes peculiar tent-shaped retinal detachments that developed among eyes with ROP.

Methods: An observational case series consisting of 9 patients and 13 eyes with tent-shaped retinal detachments. Their morphology, clinical baseline, surgical course, and final retinal status were extracted from medical records.

Results: Eight had simple tent-shaped retinal detachments, three had a double tent-shaped retinal detachment, one had a chevron-based retinal detachment, and one had a star-shaped retinal detachment. Each case had a disk based stalk that extended to the apex of the traction retinal detachment and continued anteriorly; 12 stalks inserted in the retrolental space and 1 terminated in the mid vitreous. Six eyes were stage 4A, two eyes were stage 4B, and five eyes were stage 5. Vitrectomy surgery was performed on 11 eyes. Surgery resulted in retinal attachment in nine eyes, and two retinas remained detached.

Conclusion: Tent-shaped retinal detachments are seen in patients with ROP. A stalk should be sought in evaluation of these eyes. Vitreous surgery focused on relieving this traction is often successful.

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In his description of the pathogenesis of late stages of retinopathy of prematurity (ROP), Machemer presented observations about the development of retinal detachments in ROP.1 He proposed a mechanical schema that showed that contraction of stage 3 ROP could account for the progression to segmental, circumferential, and radial retinal detachments. He concluded with a general statement that “All retinal changes seen in ROP can be explained by proliferation of tissue originating in the shunt area.” Extending these observations, the international classification of ROP further described the retinal detachment configurations.2

The presence of a stalk originating on the optic nerve in retinal detachments associated with ROP has been frequently noted.3–6 The dissection of the stalk was a well-documented step in membranectomy for ROP. Eller et al. studied the association of the persistent hyaloid artery with ROP and suggested that it was a likely part of the disturbance of vasculogenesis and angiogenesis that describe the advance stages of ROP.7 They also speculated that the hyaloid system could provide a scaffold for fibrovascular proliferation. The stalk also had practical implications since when using the open sky technique this stalk may increase the intraoperative hemorrhage,4 and using closed vitrectomy, aggressive surgery on the stalk may lead to posterior retinal breaks.6 Recently, the disk stalk was identified as one of the many vectors of traction involved in development of retinal detachment in ROP.8 Although frequently observed, the identification of a disk stalk as a sufficient cause of retinal detachment separate from the extraretinal fibrovascular proliferation (EFP) was often impossible.
because the vitreoretinal relationships were dominated by the EFP.

In this article, we describe infants with ROP that presented with tent-shaped retinal detachments rather than radial, segmental, or circumferential folds. This tent-shaped morphology is associated with persistent fetal vasculature (PFV), and very different from the retinal detachments generally seen in ROP. Careful inspection revealed that these retinal detachments were not a result of the contraction of the extraretinal fibrovascular proliferation, but rather the contraction of a hyperplastic disk stalk probably related to the hyaloid artery. Identification of these unexpected retinal detachment configurations is helpful for diagnosis by ophthalmologists who examine or treat ROP.

**Materials and Methods**

Tent-shaped detachments were defined as a traction retinal detachment with five criteria: one, a well-defined small apex and relatively broad base; two, retinal detachment apex well within the vascularized retina; three, tubular fibrous stalk connected to the retinal apex; four, stalk emanating from the optic nerve disk; and five, traction from the stalk directed into central vitreous. Records of patients with traction retinal detachments from ROP seen in a pediatric retina practice were reviewed. Cases with a documented tent-shaped retinal detachment were selected. Eyes with a radial or circumferential fold or detachment as the predominant morphology were excluded. All eyes that had an optic nerve disk stalk discovered during surgical dissection were also excluded.

The baseline characteristics were recorded, including the estimated gestational age and birthweight. The history was reviewed using the postmenstrual age as the standard. Preoperative evaluation focused special attention on the retinal examination. The form of the retinal detachment was studied. The EFP was considered for extent, location, vascularity, and its position relative to both the retina and the lens. Postoperative follow-up ranged from 6 months to 6 years and included patient history, examination with indirect ophthalmoscope, and slit lamp examination (when age appropriate).

**Results**

Tent-shaped retinal detachments were found in 13 eyes of 9 patients with ROP (Table 1). All patients were born prematurely and also had severe ROP in the contralateral eye. One was first examined as an adult; of the remainder, five infants were seen during the active phase and three infants were referred during the cicatricial phase of ROP. The birthweights ranged from 550 g to 1800 g. Among the pediatric cases (excluding Patient 9) the average birthweight was 611 g. The infants were born at estimated gestational ages that ranged from 23 to 26 weeks with a mean of 24.5 weeks. Three of the infants were inborn. Five were referred for treatment. The one adult, ex-preterm neonate, was referred for long-term observation. The retinal detachments evolved under supervision among the inborn patients. All but two eyes among the infants reached threshold before development of retinal detachment. Eight were in zone 1 and four were in zone 2. All threshold eyes were treated with laser photocoagulation.

There were four configurations of retinal detachments. Eight eyes showed simple tent-shaped retinal detachment (Figure 1). In these eight eyes, fibrous elements originated on the optic disk and connected...
along the retina surface to the apex of the retinal detachment within the vascularized retina, and inserted anteriorly in the central vitreous, anterior vitreous, or posterior lens (Figure 2). In eight eyes there were accompanying peripheral segmental folds due to contraction of persistent stage 3 ROP. In Cases 1, 7, 8, and 9 the EFP involuted and exerted no traction on the retina. Since the media were clear, ultrasounds were only occasionally performed. In one eye the ultrasound showed the stalk extending from adjacent to the nerve to the retrolental space and in the second eye it extended from the disk to the central vitreous (Figure 3). Connection to the condensed anterior vitreous was noted in five eyes, and in one a connection to vitreous that bridged across a circumferential fold was recorded. Among all the eyes with tent-shaped retinal detachments, the area of retinal detachment was limited by the disk and the laser induced chorioretinal atrophy. Some vascular and retinal dragging was always present among cases with tent-shaped retinal detachment.

In five eyes more complicated retinal detachment configurations developed. One eye presented with a tent-shaped detachment with a chevron-shaped base (Figures 4 and 5). This shape reflected the straightening of the arcade blood vessels proximal and distal to the apex forming a chevron as a result of the anterior traction at the apex. Three eyes presented with a double tent with the stalk connected to two retinal detachment apices, one nasal and one temporal. A fifth eye showed a thick stalk with tight connection 360 degrees and presented with a star shape (Figure 6).

Peripapillary fibrosis was noted in two eyes (Figure 7). Vitreous hemorrhage was seen in two eyes. Using the international classification of ROP six eyes reached stage 4A, two eyes reached stage 4B, and five eyes reached stage 5. Initial scleral buckle was performed on three eyes. In two cases the buckle reduced
the peripheral component of the traction retinal detachment; however the central traction retinal detachment progressed in all three cases. Vitrectomy with membranectomy was performed on 11 eyes. Seven eyes underwent lens sparing vitrectomies; repeat lens sparing vitrectomies were performed in two eyes, and four eyes had combined lensectomy and vitrectomy.

Fig. 5. A tent-shaped retinal detachment with a chevron base. The origin is seen at the disk and the stalk is connected to the vessels near the disk. The apex is seen along the inferior arcade blood vessels labeled as “on end view of anterior extent of stalk.” The proximal and distal portions of the vessels are elevated, straightened, and dragged so that the detachment base forms a chevron. The blood vessel distal to apex forms the crest of the detachment more peripherally.

Fig. 6. A star-shaped retinal detachment with 360 degree connections of the retina elevated and drawn towards the stalk with draping of the retina.

Fig. 7. Fibrosis of the optic nerve head with infiltration of vitreoretinal interface. Striae as seen inferiorly are a common manifestation and may indicate the alterations that precede the connection of the stalk to the retina. Inferior nasal there was a disk stalk.
procedures. Two eyes failed treatment and nine reattached. The two cases with surgical failure were preoperatively classified as stage 5 ROP.

Discussion

Machemer identified the key factors in the development of retinal detachments from ROP as the contraction of the circumferential shunt with stretching and elevation of the retina, the traction from intravitreal proliferation, and the position of the shunt along the anterior posterior axis of the eye. The resultant vitreoretinal configurations in eyes with a radially symmetric distribution of disease centered on the disk are circumferential folds and funnel-shaped detachments. In eyes with radially asymmetric distributions of fibrovascular shunt tissue the retina developed radial folds, segmental folds, incomplete funnels, eccentrically displaced funnels, and spiral funnels. Recognizing the additional pathologic forces responsible for retinal detachment and understanding the vector combinations that lead to the different patterns of detachment is especially important in determining the effective surgical approach since the membranes often obscure much of the retina. In this report, we described eyes with ROP that developed tent-shaped retinal detachments. Recognition of this uncommon pattern of retinal detachment may help during monitoring of the ROP patient and surgical management.

The tent-shaped retinal detachments reported were most similar to the typical radial folds described in ROP. However, they presented distinct features. First, in each case the tractional element was a stalk emanating from the disk rather than a flat, pleated, or bunched sheet that originated from the EFP. Second, the apex of the tent-shaped retina detachment was well within the vascularized retina rather than at the junction of the vascular and avascular retina. Third, the stalk extended from the apex centrally into the midvitreous, anterior vitreous, and posterior lens rather than the peripheral vitreous, ciliary body, and peripheral lens. Some of the cases revealed retinal traction from both the disk stalk and extraretinal fibrovascular proliferation. However, we excluded the cases in which major EFP was associated with extensive opaque vitreous proliferation to emphasize the stalk component as a sufficient cause of retinal detachment.

Sporadic persistent fetal vasculature syndrome is quite rare, but findings of PFV are common among premature neonates. In normal eyes the primary vitreous vasculogenesis stops growing at 10 weeks, and the branch vessels slowly atrophy. The posterior branches of the hyaloid trunk, vasa hyaloidea propria, regress first and are completely atrophic by 8.5 months gestational age. The hyaloid artery itself closes at 8 months and regresses soon afterwards. The neonatal cases in this report evolved in the setting of extreme prematurity, generally followed complete laser treatment, and were either inborn or early referrals. In our cases both retinal vasculogenesis and hyaloidal artery involution were disturbed.

The development of the normal eye in humans and most mammals includes a chronological overlap and proportional relationship between retinal vascularization and vascular involution of the primary vitreous. In many animal models of ROP the hyaloidal system persists and in some the hyaloidal blood vessels proliferate. Bischoff et al displayed persistent hyaloidal vessels in the mouse models of ROP using scanning electron microscopy. They described the hyaloid vasculature as a high resistance passive system and the retinal vasculature as a low resistance active system. They suggested that oxygen exposure constricts the retinal vasculature and raises the hydrostatic pressure in the retinal vessels, driving the blood into the functionally closed hyaloidal vasculature. This diversion of blood causes a persistence and distention of the tunica vasculosum lentis and vasa hyaloidea propria. Larrazabal and Penn showed the arborizing hyaloid system with fluorescein angiography of newborn rats recovering in room air after 14 days of 80% oxygen exposure. They demonstrated an extensive and tortuous hyaloid vascular system as well as tortuosity of the major retinal vessels. This model may be the most similar to the neonatal human condition. Berkowitz et al used a noninvasive method, carbogen enhanced MRI, to study the hyaloidopathy in newborn rats. They found that the functional volume of hyaloidal circulation occupies a larger volume of the vitreous in ROP rats compared with controls. These findings confirm and refine the essential finding of Patz and although not in complete agreement, build on the work of Ashton. Bischoff also suggested a role for the persistent vasculature in some cases of retinal detachment. Indeed, the cicatrization of the hyaloid vascular system was a major contributor in the beagle model of ROP retinal detachment.

Finally, we reported special cases in which the relationship between the stalk and the tent-shaped retinal detachments were isolated and easy to appreciate. More frequently, a complex mix of forces must be analyzed and treated. We believe that early referrals for failure of laser treatment may reduce the complexity of the retinal detachments. If this is the case, observation of the hyperplastic disk stalk as a vector contributing to retinal detachment may achieve greater importance for diagnosis and treatment of late stages of ROP.
Key words: hyaloid artery, persistent fetal vasculature (PFV), persistent and hyperplastic primary vitreous (PHPV), retinal detachment, ROP, pediatric retinal detachment, tent-shaped retinal detachment.

References