



# Bleb-like posterior combined retinal detachment in severe retinopathy of prematurity: clinical characteristics, management challenges and outcome

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## To the Editor:

Retinal detachments associated with retinopathy of prematurity (ROP) are predominantly tractional [1], occasionally exudative [2], and rarely rhegmatogenous. The configuration of the tractional retinal detachment in ROP is guided by the fibrous tissue growing over the shunt at the junction of the vascular- avascular retina [3] and the hyaloid stalk. Various tractional forces in the vitreous and previous treatment, if any, modulate these changes. The tractional forces can include ridge to ridge (trans-ridge), ridge to the retina, lens, hyaloid or ciliary body, etc. ROP detachment assumes a tent-shaped configuration when the tractional vectors from the contracting fibrovascular tissue over the ridge and around the optic disc predominate [1]. Isolated exudative detachment in ROP is often peripheral and can extend posteriorly but is relatively uncommon and can, at times, follow laser treatment. The detachment, occasionally,

can have a bullous configuration when it is secondary to exudation. Rhegmatogenous retinal detachment, uncommon in active ROP, can happen in subjects with regressed cicatricial ROP at a later stage secondary to firmly attached contracting posterior hyaloid or fibrous tissue. In this article, we report a series of 5 babies (10 eyes) with atypically configured bleb- shaped posterior combined tractional and exudative retinal detachment in severe ROP, not yet reported in the literature (Medline search).

The demographic details, mode, sequence, and timing of various interventions are summarized in Table 1. All babies had an eventful neonatal course and were treated at the NICUs with 100% oxygen supplementation using a hood or nasal prongs. The mean gestational age (GA), postmenstrual age (PMA), and birth weight (BW) were 29.8 (range 28–31) weeks, 36.3 (35–39) weeks, and 1252 (range 1060–1600) grams respectively. All babies had a delayed presentation, from 5 to 8 weeks after birth (Average:6.6 weeks).The detachments were-located posteriorly around the optic disc with attached but avascular periphery (Supplementary Table 1, Fig. 1, and Supplementary Figs. 1–4).The retinal vessels were scanty, disorganized, indistinct with a very posterior location in and around the bleb. Each eye received a combination of intravitreal bevacizumab half adult dose, visible area laser, and vitrectomy (lens sparing in 7 of 10 eyes) within a span of 1–3 weeks after presentation. Three eyes had good anatomical outcome, in 4 eyes; the disease was stabilized at cicatricial stage 4B, and it progressed to stage 5 ROP in 3 eyes

The atypically configured bleb-shaped predominantly exudative and combined with focal tractional posterior only detachments in babies with ROP described here are not reported before (Medline search with keywords, ‘bleb or blister-like detachment’, ‘ROP’, ‘atypical detachment’, ‘configuration of Stage 4B ROP’, ‘combined ROP detachment’). These are different from classical tractional or

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Case1 was presented as a challenging case at the 3rd annual Indian Retinopathy of Prematurity (iROP) Society meet, July 21–22, 2018

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**Table 1** Profile and outcome of babies treated for posterior bleb shaped combined exudative and tractional ROP detachment.

Case No.	GA	BW	Neonatal care		PMA at Presentation (wks)	Eye	Intervention timing (PMA in wks)			Follow up Duration (months)	Outcome (Till last follow up)
			NICU stay (days)	O <sub>2</sub> suppl Days Mode			Anti VEGF	Laser	Surgery(s)		
1	28	1145	12	08 100% hood	36	OD	36–37	36–37	LSVS (38) Vit lavage (42) Lens +Revit. (60)	24	TRD (posterior pole) Peripheral retina attached
						OS			LSVS (38)		Very good Attached retina, Undistorted macula
2	31	1305	28	03 100% hood	38–39	OD	38–39	39–40	OU Lens +Vit. (40) OU Revit. (59)	30	Very poor Progressed to Stage 5 ROP
						OS					Stage 4B Cicatricial ROP
3	30	1150	30	25 100% hood	35	OD	35–36	35–36	Lens+Vit. (38)	04	Progressed to Stage 4B/5 ROP
						OS			LSVS (38)		
4	30	1060	30	21 100% hood	35	OU	35–36	36–37	LSVS (37)	04	Progressed to stage 4B cicatricial ROP
5	30	1600	12	08 100% Nasal prongs	37	OU	37–38	37–38	LSVS (37–38)	20	Very good Undistorted macula, attached retina

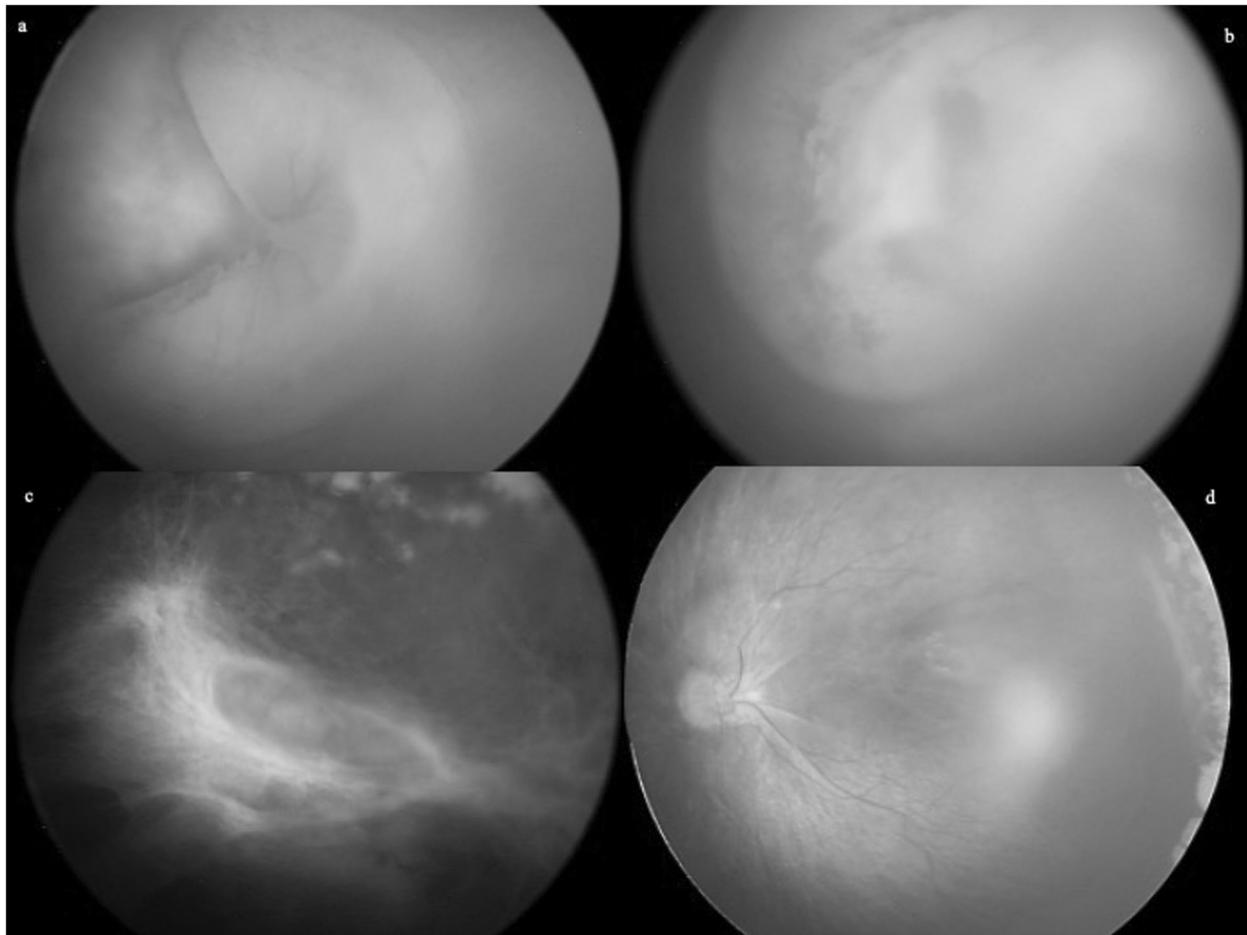
GA Gestational Age, BW Birth weight, NICU Neonatal intensive care unit, O<sub>2</sub> suppl oxygen supplementation, PMA post menstrual age, wks weeks, anti VEGF anti vascular endothelial growth factors, LSVS Lens sparing vitreous surgery, Vit lavage vitreous lavage, Lens+Revit lensectomy+ revitrectomy, TRD Tractional retinal detachment, Revit revitrectomy, Lens+Vit lensectomy+vitrectomy.

exudative retinal detachments in ROP by their physical configuration (Fig. 1 and Supplementary Figs. 1–4) and response to treatment. (Online supplement Table 1). Because all affected babies had a stormy post-natal course and received 100% unblended oxygen, we suspect oxygen-induced retinopathy could be the predominant pathogenesis. However, the extensive posterior exudation along the developing arcades has an unknown mechanism and needs further studies. The disorganized retinal vasculature and relatively posterior location than expected for the age (GA 28–30 weeks; PMA 36–39 weeks) or birth weight seen in these babies strike a resemblance to oxygen-induced retinopathy in the experimental animal [4]. We suspect the extensive avascular periphery with high vascular endothelial growth factor (VEGF) drive could have accounted for severe fibrovascular proliferation over the posterior pole, as reported by Yokoi et al. [3].

The exact reason for the predominant posterior pole location and presence of large cystic exudation with sanguineous fluid is unclear. We hypothesize exudation from these immature blood vessels, aggravated further by the

shearing force from the contracting fibrovascular tissue limited to the posterior disc and immaturely developed macular tissue, could have contributed to the bleb-like configuration to these detachments. These posterior detachments had several management challenges due to the atypical configuration of the detachment, disorganized retinal vasculature located within the posterior zone I, inadequate laser in the first sitting because of poor systemic health, severe-plus component, intra, and postoperative hemorrhages, and anti-VEGF crunch effect. The response to intravitreal bevacizumab in these babies was peculiar- the fibrovascular proliferation became more organized, white with increased traction, similar to the ‘crunch’ effect described by Yonekawa et al. [5]. We also encountered extreme difficulty in dissecting the traction tissues safely overlying the optic disc. The outcome was good, where intravitreal bevacizumab, laser, and surgery were performed in quick succession, all within a week (as in the left eye of baby #1 and both eyes of baby #5).

In conclusion, bleb like posterior combined exudative and tractional retinal detachment in severely sick babies



**Fig. 1 Fundus images of both eyes of case #1 at presentation and 5 months post surgery.** **a** Color fundus photograph of right eye showing a bleb like localized retinal detachment with overlying fibrovascular tissue obscuring the disc details. Retinal vascular architecture is absent except for few loops with tortuosity superotemporally and straightened retinal vessels inferotemporally within posterior zone I. The detachment is bordered by a zone of sub retinal/ sub-retinal pigment epithelium (sub-RPE) hemorrhages superiorly and nasally. **b** Color fundus photograph of left eye shows a localized detachment with overlying central fibrovascular tissue obscuring the disc details.

Supero-nasal part of detachment is dome shaped with sub-retinal and sub-RPE hemorrhage nasally and inferiorly similar to the right eye. Superotemporal dilated and tortuous vessels are restricted to zone I with pre-retinal hemorrhages. **c** Fundus picture of the right eye 5 months post-surgery shows a plaque of fibrous tissue overlying the posterior pole with lasered and attached peripheral retina. **d** Fundus picture of the left eye shows a stable and lasered peripheral retina with a glial tissue bridging between two retinal folds along supero- and inferotemporal arcades (The greyish reflex at the mid periphery is an artifact).

with ROP who receive 100% unblended oxygen is a possibility. This probably represents a form of oxygen-induced retinopathy. Quick and rapid combination therapy using the laser, anti-VEGF injection, and surgery could salvage some eyes. However, closely monitored oxygen therapy could possibly prevent such events that do not have a good prognosis.

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## Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

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