

Spontaneous Globe Rupture in Infant with Retinoblastoma

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Abstract

This article describes spontaneous globe rupture as a rare but serious complication of retinoblastoma. This warrants acknowledgement and discussion, given the implications for metastatic spread should it not be managed appropriately.

Spontaneous Globe Rupture in Infant with Retinoblastoma

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Abbreviations

MRI	Magnetic resonance imaging
RB-1 Gene	Retinoblastoma 1 gene
mmHg	Millimetre of mercury

Spontaneous globe rupture in infant with retinoblastoma

Abstract: This article describes spontaneous globe rupture as a rare but serious complication of retinoblastoma. This warrants acknowledgement and discussion, given the implications for metastatic spread should it not be managed appropriately.

Article:

We report the rare complication of spontaneous globe rupture secondary to unilateral retinoblastoma in an infant with Group E retinoblastoma. A 5-month-old previously healthy infant presented with a 1 month history of right leukocoria. On examination under general anaesthesia, the right intraocular pressure was 50mmHg, left 26mmHg. The right eye had an uninflamed conjunctiva, clear cornea and normal diameter. The anterior chamber was shallow with iris that bowed anteriorly. There was no rubeosis or anterior chamber cells. A white tumour with overlying retinal detachment filled the entire vitreous cavity and posterior chamber, moulding to the posterior curvature of the lens. B-scan ultrasonography showed a solid, homogenous lesion with hyperechoic foci. The left eye was normal.

MRI scan of her brain confirmed the tumour, but showed no scleral invasion or extraocular tumour extension. Enhancement of the right central retinal vessel was noted. No contralateral or intracranial tumour was identified. Lumbar puncture showed no evidence of tumour cells within the cerebrospinal fluid. Following a multidisciplinary team discussion and counselling of parents, right enucleation was scheduled for the following week.

Six days following the initial assessment, the patient attended the emergency department with an injected right eye with creamy discharge. It was noted that her crystalline lens had expelled. No trauma was reported prior. The patient was diagnosed with spontaneous right globe rupture and proceeded to undergo an urgent enucleation that same evening (Figure 1A). Peritomy exposed thin sclera at the limbus with a rupture at 9 o'clock. Prior to enucleation, the site of rupture was carefully exposed without opening any tissue planes and submerged in distilled water for 3 minutes. The limbal defect was closed with 6-0 vicryl suture (Figure 1B). The ruptured area was then resubmerged in distilled water to mitigate further spillover during the enucleation process. No implant was inserted due to concern this may hinder the detection of orbital recurrence. The patient was commenced on emergent chemotherapy postoperatively as per ARET0332 using vincristine, etoposide and carboplatin for 6 cycles.

Histology showed a moderately differentiated retinoblastoma with haemorrhage, mixed inflammation, necrosis and dystrophic calcification within the lesion. The site of the rupture at the limbus showed interstitial keratitis and scleritis but there was no tumour invasion of the sclera, iris or choroid. No infectious organisms were identified. Tumour genetics found non-heritable homozygous RB1 whole gene deletion. 3-monthly MRI scans, the latest at 16 months post-diagnosis, have shown no recurrence.

Spontaneous globe rupture secondary to retinoblastoma is rare. The only reports found in the literature are dated in the 1900s as a complication of end-stage disease, whereby perforation would occur at the limbus, often preceded by buphthalmos and rupture of Descemet's membrane or sclera (1). This case is noteworthy in that globe rupture occurred without warning or clear cause. Whilst the intraocular pressure was elevated, this in itself is not unusual for many cases of group E disease (2, 3). Furthermore, static and dynamic studies of post-mortem eyes have shown that the human eye has the capacity to maintain globe integrity up to 900mmHg before globe rupture may occur (4). The presence of kerato-scleritis on histology may have contributed to scleral weakness.

Globe rupture carries a high risk of tumour contamination and metastatic spread. To mitigate this risk, surgery was expedited, the surgical site was irrigated with distilled water prior to enucleation, and patient commenced on emergent chemotherapy. Submersion of retinoblastoma cells in distilled water leads to cell lysis and may be useful to limit surface tumour spread (5).

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