Int Ophthalmol (2019) 39:1603–1606
https://doi.org/10.1007/s10792-018-0960-9

CASE REPORT

Ectopic lens material in an otherwise healthy 5-week-old infant

Axelle Rigaudy · Manoj Parulekar · Caspar Gibbon · Anthony Quinn

Received: 2 December 2017 / Accepted: 13 June 2018 / Published online: 21 June 2018

Abstract
Purpose To report the unusual finding of ectopic lens material in an otherwise healthy 5-week-old infant.
Methods Case report and literature review.
Results An asymptomatic 5-week-old female infant was found to have unilateral ectopic lens material in the retrolental space of the left eye associated with a posterior capsular defect.
Conclusion The abnormality is likely embryological in origin, and the established progression for similar conditions means long-term monitoring is required to ensure the best possible visual outcome.

Keywords Posterior capsular defect · Ectopic lens material · Paediatric cataract

A. Rigaudy (✉)
Ophthalmology Department, Musgrove Park Hospital, Taunton, UK
e-mail: axelle.rigaudy@doctors.org.uk

M. Parulekar
Eye Department, Birmingham Children's Hospital, Birmingham, UK

C. Gibbon
Ophthalmology Department, North Devon District Hospital, Barnstaple, UK

A. Quinn
West of England Eye Unit, Royal Devon and Exeter Hospital, Exeter, UK

Introduction

We report the unusual finding of ectopic lens material posterior to the lens capsule in a 5-week-old infant.

This unusual case provides interesting insights into the embryology of the anterior segment [1, 2]. The abnormality described can mimic a rare form of retinoblastoma, making this distinction vital [3]. The lens develops from the surface ectoderm, and developmental lens abnormailities are well recognized [1, 2]. They can be associated with posterior capsular defects (PCDs) as seen with posterior lenticonus, or posterior polar cataracts (5,8). Congenital lens and posterior capsular abnormalities are one of the common cause of preventable childhood blindness and have a diverse aetiology including antenatal infections and genetic disorders but the majority are idiopathic [1, 2, 4].

The presence of lens material outside the confines of the capsule associated with posterior capsular defects without the presence of a lens opacity is a unique case in the literature. We describe distinguishing features of this rare case and explore possible etiologies.

Case report

A 4-week-old female infant was noted to have an abnormal left red reflex in the nasal aspect on neonatal

screening. She was a healthy twin, born at 36 weeks of gestational age by planned Caesarian section. Antenatal history was negative for infection. Neonatal screening for her twin sister was normal including the red reflex but was not formally examined by an ophthalmologist. Prior to presentation, her parents reported no visual concerns. There was no known family history of cataract in early life or any other ocular morbidities.

On assessment, she was alert, growing, and feeding well and otherwise healthy. Awake examination revealed a white, avascular mass, posterior to the nasal periphery of the lens.

An examination under general anaesthesia was arranged. The anterior segments of both eyes were normal. A dilated examination showed two multilobulated discrete and avascular white lesions. These were situated posterior to, but not in direct continuity with the nasal aspect of the lens.

There were two other opacities in the lens: one at the 9 o'clock position in the mid-peripheral lens visible on retroillumination, and the other opacity in the superonasal quadrant in the posterior part of the lens, coexistent with posterior capsular defects (PCD).

The patient was further assessed using RetCam imaging (Fig. 1a–c), ultrasound B scan and ultrasound biomicroscopy (UBM). The ultrasound B scan and UBM could not adequately delineate the lesions. The rest of the eye examination was normal apart from dot peripapillary retinal haemorrhages which were likely birth hemorrhages. The ciliary body appeared normal on the UBM.

This is a highly unusual appearance and will require monitoring. As the lens opacities were not visually significant, surgical intervention was not required. It was, therefore, not possible to confirm our diagnosis by histopathology. Several genetic conditions can be associated with congenital cataracts and PCDs [1, 2, 5]. Although there is no recognizable pattern in this case, further testing with a congenital cataract genetic panel will be arranged. It will also be interesting to observe whether the ectopic lens matter continues to proliferate over time. We plan in 3 months' time to perform an examination under anaesthesia and if stable in appearance, 6-monthly thereafter until we are confident that there is no progression. Additionally, she will be monitored for the development of a cataract. In the event of a cataract, the posterior lens abnormality will need to be taken into consideration and may complicate surgical management [1].

Discussion

We describe an unusual case of a healthy female infant with unilateral ectopic lens material in the retrolental space. In an otherwise healthy child without signs of lens opacification, this is a unique example.

Considering the findings, a possible aetiology to be excluded is diffuse anterior retinoblastoma. This can present in a similar extra-retinal location anterior to the ora serrata. However, its appearance is distinctive, often described as white and fluffy aggregates [1] thus lacking the delineated crystalline appearance present in this case. A further malignant differential of medulloepithelioma was unlikely given the multiloculated white lesions being separate from the normal ciliary body or iris [2, 6].

The white lesions are most likely lenticular in origin which is supported by the finding of a localized abnormality on the posterior lens surface visible on high magnification. The location of the lesions, their appearance, as well as the overlying abnormality in the posterior lens capsule would suggest that this is a developmental anomaly, with sequestered lens material in the retrolental space. The retrolental lesions and the PCDs are therefore likely to be related.

PCDs associated with unilateral cataract have been more frequently reported, especially in males, which differentiates this case in the literature [1, 2, 6]. PCDs have been described in cases of traumatic cataracts, intraocular tumours and posterior polar cataracts [7]. The aetiology of preexisting PCD such as in this patient is unknown; but intrauterine abnormalities of the embryonic lens may have a role in its pathogenesis [3].

A mechanism responsible for the appearances in this case may be similar to that seen in the commonly described Mittendorf's dot. This is a small axial or nasally paraxial grey-white dot opacity at the posterior apex of the lens and is often associated with PCDs [8]. The PCD is thought to arise through the hyaloid artery's contact with the posterior lens capsule, which itself then degenerates and forms Mittendorf's dot [9]. Though no remnant of the hyaloid vasculature was present on Ultrasound B examination in this case, a

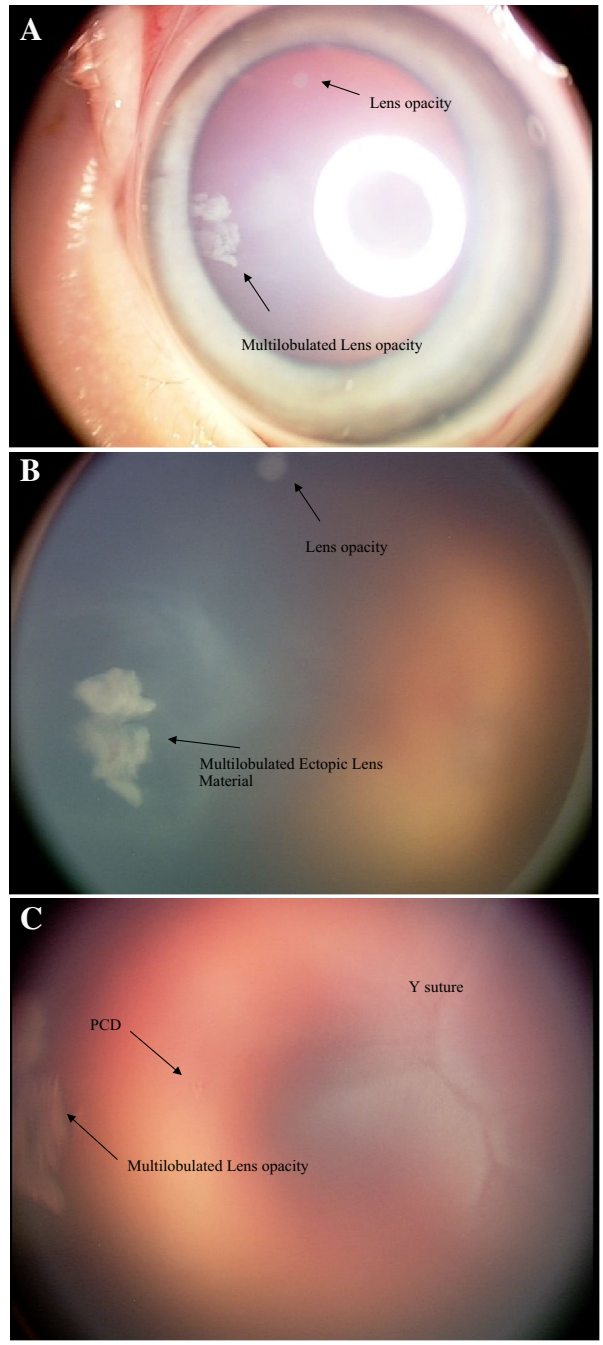

Fig. 1 RetCam images of ectopic lens material. **a** Intraoperative RetCam photograph of the left eye highlighting the multiple opacities posterior to the lens. **b** Intraoperative RetCam photograph of the left eye highlighting the multiple opacities posterior to the lens. **c** Intraoperative RetCam photograph of the left eye highlighting the multiple opacities posterior to the lens, the posterior capsular opacification

comparable mechanism may be responsible for the presence of the PCD and the ectopic lens material.

Though this patient is free of lens opacification and visual disturbance, these may develop in the future and will need ongoing monitoring. If cataracts do develop surgical management will need to be approached carefully to avoid enlargement of the PCD and ensuing complications [10].

Limitations in this case were the absence of a biopsy and histological assessment. As the patient remains symptom free, current intervention is not required. The case may also have benefitted from

further genetic testing as many genetic conditions are associated with congenital cataracts and PCDs [7].

Conclusion

This is a rare presentation of ectopic lens material associated with a PCD. Though no visual symptoms have been reported so far, given the likely embryological aetiology and the established progression for similar conditions, long-term monitoring is required to ensure the best possible visual outcome.

Compliance with ethical standards

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

Informed consent Informed consent from legal guardians obtained.

References

1. Graw J (2003) The genetic and molecular basis of congenital eye defects. Nat Rev Genet 4(11):876–888. https://doi.org/10.1038/nrg1202
2. Amaya L, Taylor D, Russell-Eggitt I, Nischal KK, Lengyel D (2003) The morphology and natural history of childhood cataracts. Surv Ophthalmol 48(2):125–144. https://doi.org/10.1016/S0039-6257(02)00462-9
3. Jijelava KP, Grossniklaus HE (2013) Diffuse anterior retinoblastoma: a review. Saudi J Ophthalmol 27(3):135–139. https://doi.org/10.1016/j.sjopt.2013.06.006
4. Yeung HH (2014) No red reflexes and retrolental opacities in the newborn nursery. J Pediatr Ophthalmol Strabismus 51(2):73–74. https://doi.org/10.3928/01913913-20140220-01
5. Tandon AK, Oltra EZ, Velez FG (2015) Bilateral congenital posterior capsular defects and ectopic cataracts. J Pediatr Ophthalmol Strabismus 52 Online:e48–e51. https://doi.org/10.3928/01913913-20150915-02
6. Shields CL, Schoenberg E, Kocher K, Shukla SY, Kaliki S, Shields JA (2013) Lesions simulating retinoblastoma (pseudoretinoblastoma) in 604 cases: results based on age at presentation. Ophthalmology 120(2):311–316. https://doi.org/10.1016/j.ophtha.2012.07.067
7. Bozkurt E (2011) Bilateral preexisting congenital posterior capsular defects with accompanying membranes. J Clin Exp Ophthalmol. https://doi.org/10.4172/2155-9570.1000148
8. Peshtani A, Kaliki S, Eagle RC, Shields CL (2014) Medulloepithelioma: a triad of clinical features. Oman J Ophthalmol 7(2):93–95. https://doi.org/10.4103/0974-620X.137171
9. Vajpayee RB, Angra SK, Honavar SG et al (1994) Preexisting posterior capsule breaks from perforating ocular injuries. J Cataract Refract Surg 20:291–294
10. Zetterstrom C, Lundvall A, Kugelberg M (2005) Cataracts in children. J Cataract Refract Surg 31(4):824–840. https://doi.org/10.1016/j.jcrs.2005.01.012