

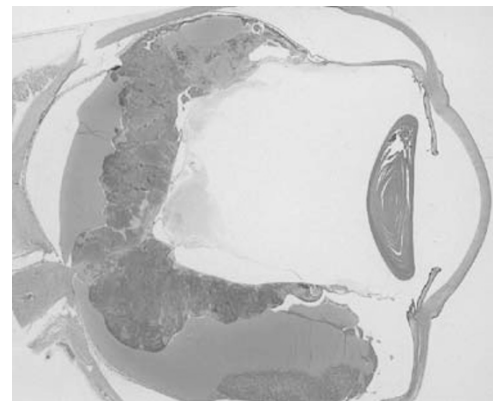
Sir,  
**Retinoblastoma presenting as Coats' disease**  
We present a case of atypical presentation of retinoblastoma masquerading as Coats' disease.

**Case report**

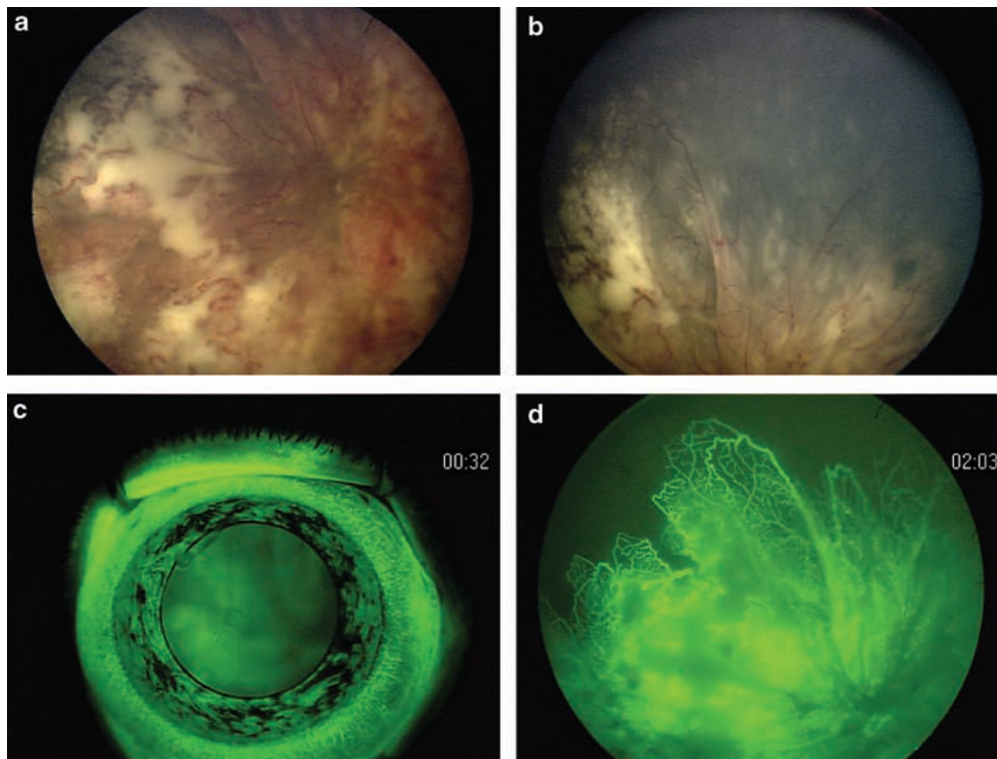
A healthy 4-year-old boy presented with leukocoria of the right eye. On examination, the patient demonstrated poor visual fixation in the right eye and steady fixation in his left eye. During examination under anaesthesia, intraocular pressures were within normal ranges. Ultrasonography confirmed a detached and diffusely thickened retina with no intraretinal calcification. Indirect ophthalmoscopy revealed an unusual exudative retinal detachment. The retina was diffusely thickened and associated with intraretinal and preretinal haemorrhages, a white flocculent material in between areas of retinal folds, and an anomalous retinal vasculature pattern throughout (Figure 1a). Intraoperative fluorescein angiography confirmed this unusual vascular pattern with arterio-venous communication, prominent telangiectatic vessels, leakage, and 360° of peripheral capillary non-perfusion (Figure 1b and d). Anterior segment evaluation showed diffuse iris hyperfluorescence consistent with iris neovascularization (Figure 1c).

The differential diagnosis included Coats' disease, persistent hyperplastic primary vitreous, retinal dysplasia, and an atypical presentation of retinoblastoma. Given the uncertainty over the clinical

diagnosis and the poor visual prognosis, enucleation of the eye was performed. Pathologic study revealed an endophytic mass arising from the retina extending from ora to ora, with vitreous seeding and extensive exudate in the subretinal space. There were large basophilic nuclei with mitotic figures (Figure 2). The diagnosis of retinoblastoma with Coat-like response was confirmed.



**Figure 2** Macrophotograph of haematoxylin and eosin-stained section of the enucleated eye. The retina is replaced by an intraretinal mass extending from ora to ora, with extensive subretinal exudate.



**Figure 1** Indirect funduscopy photographs show (a) exudative retinal detachment with white flocculent material, retinal folds, anomalous vasculature, and haemorrhage. (b) Peripheral retinal vasculature with arterio-venous communication. (c) Fluorescein angiogram of the anterior segment shows prominent iris neovascularization and (d) FA of the posterior segment shows telangiectasia, leakage, and peripheral capillary non-perfusion.

### Comment

Atypical manifestations of retinoblastoma (presenting at older age, with unusual features such as pseudohypopyon, hyphaema, or vitreous haemorrhage) can pose diagnosis challenges. Retinoblastoma masquerading as Coats' disease has been described previously.<sup>1</sup> This case is unique because ultrasonography and fluorescein angiography strongly mimicked typical findings in Coats' disease, including lack of calcification, dilated telangiectatic vessels with leakage, and peripheral non-perfusion. Shields and Shields<sup>2</sup> have suggested that the calibre and distribution of retinal blood vessels can help distinguish Coats' from retinoblastoma. In this case, fluorescein angiography was misleading. It is important for ophthalmologists to maintain a high index of suspicion for retinoblastoma in all children with unusual ocular diseases in which the diagnosis is not clear.

### Acknowledgements

This study was conducted without any outside commercial interests and has not been previously published.

### References

- 1 McCaffery S, Wieland MR, O'Brien JM, Cooper KL, Wieland MR, Wendel RT. Atypical retinoblastoma presentations: a challenge for the treating ophthalmologist. *Arch Ophthalmol* 2002; **120**(9): 1222–1225.
- 2 Shields JA, Shields CL. Differentiation of Coats' disease and retinoblastoma. *J Pediatr Ophthalmol Strabismus* 2001; **38**(5): 262–266.

HD Lam<sup>1,2</sup>, MA Samuel<sup>1</sup>, NA Rao<sup>1</sup> and AL Murphree<sup>1</sup>

<sup>1</sup>Children's Hospital Los Angeles, Doheny Retina Institute, Keck School of Medicine, University of Southern California, Los Angeles, CA, USA

<sup>2</sup>Department of Ophthalmology, UCSF medical Center, San Francisco, CA, USA  
E-mail: hhdawnlam@yahoo.com

The authors have no financial interest.

*Eye* (2008) **22**, 1196–1197; doi:10.1038/sj.eye.6703046; published online 30 November 2007

Sir,  
**Reply to Wong *et al***

Wong *et al*<sup>1</sup> claim to have reported the first case of non-tuberculous *Mycobacterium* (NTM) infectious crystalline keratopathy in a 'non-traumatized' eye. However, their patient had suffered a dendritic ulcer 6 weeks before and had received topical steroids, two significant risk factors for NTM keratitis. Four cases of corneal ulcer in which NTM were isolated with no predisposing risk factors have been described previously.<sup>2</sup>

Wong *et al*<sup>1</sup> recommend the use of fourth generation fluoroquinolones as a first-line treatment for this

condition, although no *in vitro* sensitivity testing was demonstrated for the 0.3% gatifloxacin they used. *Mycobacterium abscessus* is the most resistant NTM, typically showing sensitivity only to clarithromycin, azithromycin, and amikacin, and cannot be distinguished from the closely related but less resistant *M. chelonae* in routine microbiological cultures.<sup>3</sup> DNA sequencing of the suspected pathogen was not carried out in this case, and it is not clear whether repeat microbiological sampling was done when the keratopathy recurred. The true identity of the causative organism is therefore in doubt.

NTM are ubiquitous organisms whose pathogenesis is far from completely understood; a genetic variation in individual susceptibility to infections with these atypical mycobacteria has been described.<sup>4</sup>

### References

- 1 Wong R, Luo YH, Al-Zahawi MF, Poole TRG. Infective crystalline keratopathy secondary to non-tuberculous mycobacterium (*Mycobacterium abscessus*) in a non-traumatized eye. *Eye* 2007; **21**: 1123–1124.
- 2 Lalitha P, Rathinam SR, Srinivasan M. Ocular infections due to non-tuberculous mycobacteria. *Indian J Med Microbiol* 2004; **22**: 231–237.
- 3 Petrini B. *Mycobacterium abscessus*: an emerging rapid-growing potential pathogen. *APMIS* 2006; **114**: 319–328.
- 4 Koh WJ, Kwon OJ, Kim EJ, Lee KS, Ki CS, Kim JW. NRAMP1 gene polymorphism and susceptibility to nontuberculous mycobacterial lung diseases. *Chest* 2005; **128**: 94–101.

G Clare<sup>1</sup> and S Mitchell<sup>2</sup>

<sup>1</sup>Moorfields Eye Hospital, London, UK

<sup>2</sup>Chelsea and Westminster NHS Trust, London, UK  
E-mail: gaclare@doctors.org.uk

*Eye* (2008) **22**, 1197; doi:10.1038/sj.eye.6703056; published online 29 February 2008

Sir,  
**Reply to D Shome *et al***

We read with great interest the article by D Shome *et al*, 'Ptosis caused by orbicularis myokymia and treated with botulinum toxin'.

We wish to raise the following points:

The patient did not have a ptosis but blepharospasm, resulting in a decrease in palpebral aperture size in the left eye compared to the right.

The fact that the left upper lid was relatively lower than the right would presumably be a function of increased left orbicularis oculi tone relative to the right.

Furthermore, comparing the two photographs Figure 1a and Figure 2, it would appear that the patient had been squinting, that is, the palpebral apertures on the right eye were also greater in Figure 2 compared to Figure 1a. The authors have not made any attempt to explain this. Was there in fact an element of right