# Leucocoria in a boy with Kawasaki disease: a diagnostic challenge

**Che Mahiran C D, Alagaratnam J, Liza-Sharmini A T**


## ABSTRACT

**Retinoblastoma, the most common primary intraocular malignancy of childhood, usually presents in the first three years of life. Atypical presentation of retinoblastoma can masquerade as virtually any ocular or orbital pathology, which may lead to diagnostic dilemmas especially in the presence of other systemic diseases. We report a 20-month-old boy who was diagnosed with coronary aneurysm as a complication of Kawasaki disease, and presented with sudden left eye redness. His mother noticed the presence of white pupillary reflex three months earlier. Atypical acute ocular presentation secondary to Kawasaki disease was initially suspected, but the presence of multiple calcification and mild proptosis on imaging suggested characteristics of advanced retinoblastoma. Histopathological examination of the enucleated eye, which revealed a classical rosette pattern appearance, confirmed the diagnosis. Atypical presentations of retinoblastoma are usually associated with advanced disease. The presence of other systemic conditions further complicates the diagnosis. Early diagnosis is important to reduce the mortality and morbidity.**


**Keywords: enucleation, extraocular extension, leucocoria, Kawasaki disease, retinoblastoma**

*Singapore Med J 2009;50(7):e232-e234*

## INTRODUCTION

Tomisaku Kawasaki, a Japanese paediatrician, first described the Kawasaki disease, also known as mucocutaneous lymph node syndrome in 1967.[1] Kawasaki disease is a triphasic systemic disease without a well-established aetiology. The most consistent early clinical sign is bilateral bulbar conjunctiva hyperaemia without any discharge. Bilateral mild anterior uveitis with the presence of keratic precipitates is not uncommon, and usually resolves without sequelae.[2] Other rare findings include bilateral inner retinal ischaemia, posterior uveitis, vitritis and optic nerve swelling.[3-6] Rarer ocular manifestation may be observed in Kawasaki disease.

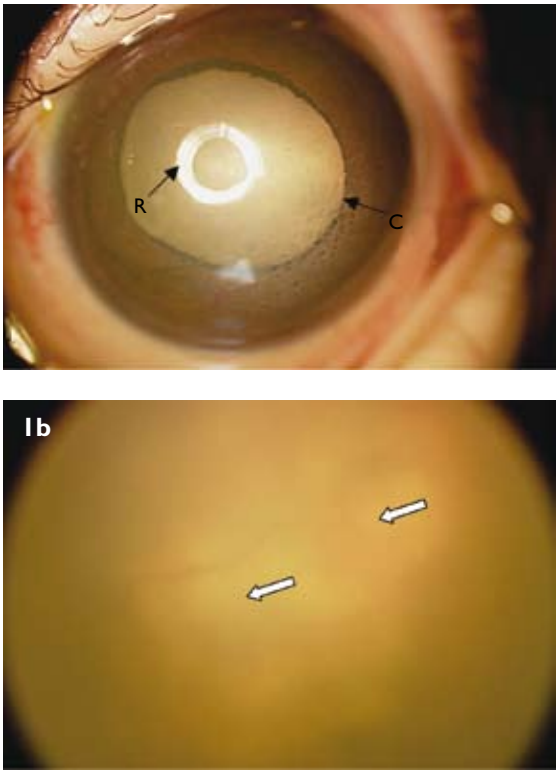


**Fig. 1** (a) Photograph of the left eye shows corneal oedema (C) with epithelial bullae and retrolental mass (R). (b) Fundus photograph of the left eye shows yellowish subretinal mass and multiple subretinal exudations (arrow).

Retinoblastoma is the common cause of leucocoria, which may be associated with mortality, especially if the diagnosis is delayed. However, anterior and posterior uveitis in ocular manifestation of Kawasaki disease may result in secondary cataract, which may also present as leucocoria in young children. Atypical presentations of retinoblastoma in the presence of systemic disease may lead to diagnostic dilemmas.

## CASE REPORT

A 20-month-old boy, who was diagnosed and treated for Kawasaki disease with coronary artery aneurysm, was referred to the ophthalmology clinic for sudden onset of left eye redness associated with swelling of eyelids for three days. His mother claimed she had noticed white pupillary reflex in his left eye three months earlier. There was no history of trauma or fever. She denied any squint or proptosis. He was born at full term without any history

**Department of Ophthalmology, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, Kubang Kerian, Kota Bahru 16150, Malaysia**

Che Mahiran CD, MD
Medical Officer

Liza-Sharmini AT, MBBS, MMed
Senior Lecturer and Consultant

**Department of Ophthalmology, Hospital Kuala Lumpur, Jalan Pahang, Kuala Lumpur 50586, Malaysia**

Alagaratnam J, MBBS, MS
Consultant Paediatric Ophthalmologist

**Correspondence to:**
Dr Liza-Sharmini Ahmad Tajudin
Tel: (60) 9766 4585
Fax: (60) 9765 3370
Email: liza@kb.usm.my

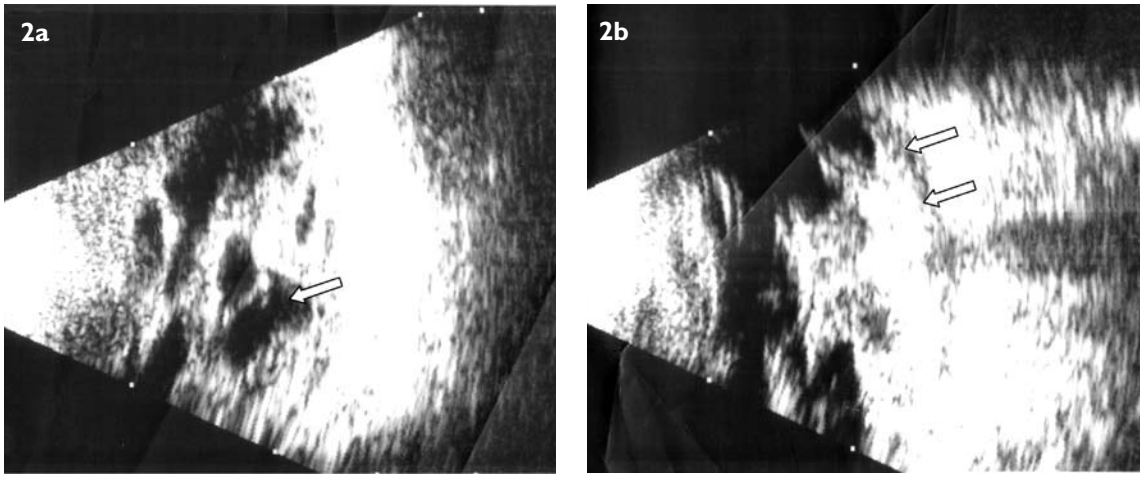


**Fig. 2** (a) B-scan US image of the left eye shows a non-homogenous loculated mass (arrow). (b) B-scan US image shows a subretinal mass with multiple calcifications (arrows).

of neonatal infection. There was also no family history of retinoblastoma or other malignancy. Nine months prior to the presentation, he was diagnosed to have Kawasaki disease complicated with coronary artery aneurysm, which was succesfully treated with intravenous immunoglobulin and methylprednisolone. He was on regular follow-up, and his recent echocardiogram showed good contractility and an ejection fraction of 68%. He was still on oral aspirin and dipyridamole. In spite of his condition, he remained active and was feeding well.

On examination, he demonstrated poor visual fixation in the left eye, but good fixation to light and object in his right eye. The young boy was able to pick up 100 and 1,000 beads with both eyes but he cried upon occlusion of his right eye. There was minimal left eye proptosis, which was hard on digital palpation, with mild swelling of the upper eyelid. He was empirically treated with oral acetazolamide, topical steroid and timolol prior to examination under anaesthesia (EUA). EUA revealed intraocular pressure of 16 mmHg in the left eye and 10 mmHg in the right eye using Perkins tonometer. The left conjunctiva was diffusely injected with oedematous cornea and corneal epithelial bullae formation (Fig. 1a). The left anterior chamber was shallow, with a mid-dilated pupil and extensive presence of posterior synechiae. Several turf of iris new vessel was noted. The lens was also cataractous. Funduscopy through the opaque media revealed multiple subretinal exudations in the left eye (Fig. 1b). The right eye was sound, without any evidence of ocular pathology.

Ultrasonography (US) confirmed a non-homogeneous loculated mass (Fig. 2a) with subretinal opacification and calcification (Fig. 2b). Orbital and brain computed tomography (CT) demonstrated mild proptosis of left eye with a minimal-enhancing retrolental mass, and multiple calcifications were observed within the mass. The optic nerve was not thickened. The extraocular muscles looked normal. Left eye enucleation with primary orbital implant was immediately performed. The histopathological examination revealed classical pathological features of retinoblastoma with the presence of a classical rosette pattern (Fig. 3a). The tumour cells were found to have invaded the Bruch's membrane, choroidal vessels, sclera and extrascleral tissue, and optic nerve head without any extension posterior to lamina cribrosa (Fig. 3b).

The full blood picture, renal and liver function tests, bone marrow aspiration and lumbar puncture were normal. He then underwent a six months' course of adjuvant chemotherapy and 20 cycles of radiotherapy. He is still under our regular follow-up. There was no evidence of retinoblastoma recurrence for the past 36 months.

## DISCUSSION

Leucocoria, or cat's eye appearance, in a young child is an alarming sign. Cataract is usually the common cause followed by retinoblastoma and other diseases such as Coats's disease. In the presence of a confirmed case of Kawasaki disease and its known complications, the ocular manifestation of Kawasaki disease could be one of the most possible differential diagnoses. Anterior segment involvement in Kawasaki disease is well documented, especially mild to moderate bilateral nonexudative conjunctivitis and acute anterior uveitis.[3-6] Posterior involvement in Kawasaki disease is rare. Ohno et al reported bilateral optic disc swelling in two patients and bilateral vitreous opacity in another two patients, in a prospective study involving 18 children with Kawasaki disease.[4] Jacob et al reported another case of optic disc

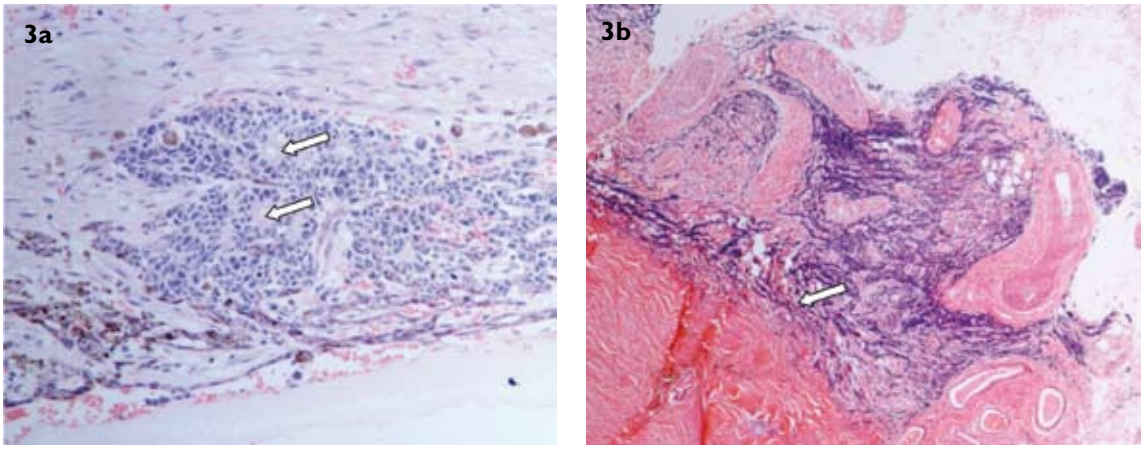


**Fig. 3** Photomicrographs show (a) cells arranged in a classical rosette pattern (arrows) seen in the enucleated eye (Haematoxylin & eosin, × 200) and (b) tumour cells invading into the extrascleral tissue (arrow) (Haematoxylin & eosin, × 50).

swelling and also a case of unilateral retinal exudates, macular and disc oedema, vitritis and preretinal membrane in his prospective study of clinical presentation of Kawasaki disease.[3] In the presence of vitritis, these retinal exudates and membrane may be suspected as a retinal mass. Furthermore, complications of severe posterior uveitis such as retinal detachment or secondary cataract may present as leucocoria. The presence of secondary glaucoma and iris neovascularisation has been reported in advanced cases of retinoblastoma,[7] which may also be the possible complication in cases of retinal ischaemia secondary to systemic vasculitis in Kawasaki disease. These findings may easily mimic retinoblastoma presentation and obscure the actual diagnosis, especially when presented in children younger than four years of age. The fundus findings in this case were inconclusive. A fundus fluorescence angiogram may easily ascertain the diagnosis by providing evidence of bilateral disc swelling and posterior uveitis in Kawasaki disease.[8] However, it is quite impossible to perform this procedure in such a young patient especially with opaque media.

The presence of multiple calcifications on US and CT findings further heightens the suspicion of intraocular mass. Although calcification is commonly associated with retinoblastoma in the presence of leucocoria, calcification following chronic retinal detachment as a complication of severe uveitis is not uncommon. The most confirmative finding was based on histopathological examination post-enucleation. The presence of a classical rosette pattern definitely excludes the possibility of severe posterior uveitis as ocular presentation of Kawasaki disease. To the best of our knowledge, there is no association of Kawasaki disease and retinoblastoma. There is no available report of a higher risk of coronary artery aneurysm in Kawasaki disease with retinoblastoma, or poorer prognosis of retinoblastoma in the presence of Kawasaki disease. Thus, we believed that these two pathologies are independent and manifested coincidentally. This case highlights the diverse spectrum of ocular manifestation of retinoblastoma, which was further complicated by the presence of a systemic disease. Atypical presentations of retinoblastoma are usually associated with advanced disease. An extensive, prompt investigation is essential to provide a conclusive early diagnosis.

## REFERENCES

1. Kawasaki T. [Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children]. Aerugi 1967; 16:178-222. Japanese.
2. Burns JC, Joffe L, Sargent RA, Glode MP. Anterior uveitis associated with Kawasaki syndrome. Pediatr Infect Dis 1985; 4:258-61.
3. Jacob JL, Polomeno RC, Chad Z, Lapointe N. Ocular manifestations of Kawasaki disease (mucocutaneous lymph node syndrome). Can J Ophthalmol 1982; 17:199-202.
4. Ohno S, Miyajima T, Higuchi M, et al. Ocular manifestations of Kawasaki's disease (mucocutaneous lymph node syndrome). Am J Ophthalmol 1982: 93: 713-7.
5. Burke MJ, Rennebohm RM. Eye involvement in Kawasaki disease. J Pediatr Ophthalmol Strabismus 1981; 18:7-11.
6. Font RL, Mehta RS, Streusand SB, O'Boyle TE, Kretzer FL. Bilateral retinal ischemia in Kawasaki disease. Postmortem findings and electron microscopic observations. Ophthalmology 1983; 90:569-77.
7. Balasubramanya R, Pushker N, Bajaj MS, et al. Atypical presentations of retinoblastoma. J Pediatr Ophthalmol Strabismus 2004; 41:18-24.
8. Anand A, Yang YC. Optic disc changes in Kawasaki disease. J Pediatr Ophthalmol Strabismus 2004; 41:177-9.