Clinical Management of Coats Disease: A Case Study

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ABSTRACT

Coats disease, is a rare unilateral retinal vascular disease of unknown aetiology though there may be a genetic predisposition to the disorder. If left untreated, severe and permanent vision loss can occur due to total exudative retinal detachment. Early intervention and close monitoring remains the most effective way to prevent potential vision loss and the progression to a blind and painful eye. This report describes the case of a 15 year-old healthy male who presented with a one-month history of unilateral blurred central vision. Fundus examination revealed a peripheral retinal vascular lesion which resulted in lipid deposits in the macular region. The patient was treated with argon laser panretinal photocoagulation and monitored over an 18-month period. He demonstrated a slow though significant resolution of the maculopathy which correlated with an improvement in visual acuity. This case highlights that early presentation followed with an appropriate management regime can result in a successful visual outcome.

Keywords: Coats disease, retinal telangiectasia, retinal detachment

INTRODUCTION

Coats disease, also known as retinal telangiectasia, is a rare unilateral retinal vascular disease[1] first identified by George Coats in 1908.[2] Though generally of unknown aetiology, some evidence has suggested a mutation of the Norrie disease gene could be a possible cause of Coats disease.[3] The exact incidence and prevalence of Coats disease is unknown, however a prospective population-based study in the United Kingdom has estimated an incidence of 0.09 per 100,000 of the population.[4] Those typically affected are males under the age of 20 years,[5] however the peak incidence of the disease occurs between six and eight years of age.[6] Coats disease is considered to be a severe form of retinal telangiectasia, a congenital retinal vascular anomaly.[6] Other types of retinal telangiectasia include idiopathic juxtafoveal telangiectasia, parafoveal telangiectasia and Leber's miliary aneurysms.[7]

The presenting signs of Coats disease, the stages and age of presentation contribute to its diagnosis as well as considerations for differential diagnosis. Children, who often present in the advanced stages of Coats disease can present with leukocoria, strabismus and intraocular mass lesions.[2] Differential diagnosis includes retinoblastoma, retinopathy of prematurity, ocular toxocariasis, choroidal hemangioma, and familial exudative vitreoretinopathy.[3] In older children and adults Coats disease must be differentiated from diabetic retinopathy, hypertensive retinopathy, retinal vein occlusions, vasculitis, melanoma, choroidal hemangiomas and juxtafoveal telangiectasia.[6] In each of these eye diseases patients present with retinal vascular anomalies with associated visual loss or disturbances.

The first sign of Coats disease is retinal telangiectasia.[8] When quiet, the telangiectasia does not cause any reduction in visual acuity and thus the patient remains asymptomatic. However, symptoms such as reduction in visual acuity occur when the retinal appearance alters and the vessels become dilated and tortuous. Aneurysms may form, giving rise to subsequent haemorrhages and exudates. Initially, the patient is usually unaware of any visual disturbance due to the often peripheral retinal location of the telangiectasia.

Recommendations regarding the management of Coats disease are well described in the literature. The primary goal of treatment is to remove areas of active retinal telangiectasia and allow resolution of lipid deposits by arresting leakage of exudates from retinal vessels.[9] This can be achieved through argon laser pan retinal photocoagulation (PRP) or cryotherapy.[6] In the presence of exudation with no retinal detachment, argon laser PRP is appropriate.[9] In those severe cases which present with a shallow detachment,

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cryotherapy is more effective in sealing retinal vessels and repairing detachments.[6] In the presence of extensive retinal detachment, vitreo-retinal surgical techniques are utilised to reattach the retina.[9] At end-stage, enucleation is required for a sore, blind and painful eye. This might be indicated if the patient presents late in the progression of the disease.[6]

Visual outcomes vary considerably between patients with Coats disease and depend largely on the nature or the stage of the retinal change and thus the time at which the patient is seen. Table 1 summarises the various stages of Coats disease and management options at each of these stages.

Table 1. Stages and classifications of Coats disease (adapted from Shields et al[9])

Stage		Criteria	Management Options
Stage 1		- retinal telangiectasia	- observation
Stage 2	2A	- retinal telangiectasia - extrafoveal exudation	- observation (2A) - photocoagulation - cryotherapy
	2B	- retinal telangiectasia - foveal exudation	
Stage 3	3Ai	- extrafoveal execdative retinal detachment	- photocoagulation - cryotherapy - retinal detachment repair - enucleation (to relieve ocular pain associated with glaucoma)
	3Aii	- exudative foveal retinal detachment	
	3B	- total exudative retinal detachment	
Stage 4		- total retinal detachment - glaucoma	
Stage 5		- end stage	

CASE REPORT

A 15 year-old male presented complaining of blurred central vision in the right eye for one month. His general health was good and no past ocular history was reported. No family history of eye disease was reported. Snellen visual acuity was R 3/36 (no improvement with pinhole) and L 6/4. Intraocular pressures were on the higher side of normal, measuring R 21 mmHg and L 19 mmHg. A right relative afferent pupil defect was noted and fundus examination revealed a macular star exudate at the posterior pole of the right eye, with slight lipid deposition in the retinal periphery (Figure 1A). A retinal vascular anomaly was also found in the upper temporal quadrant of the right eye fundus which was identified with fundus flourescein angiography (Figure 1C). No retinal abnormality was detected in the left eye (Figure 1B).

At this visit, the patient was treated with argon laser PRP over the area of retinal telangiectasia. Over an 18-month period the patient was continually reviewed and required a total of four PRP treatments to his right eye. A slow and steady improvement in his visual acuity throughout this time was evident as the macular exudate resolved (Figure 2).

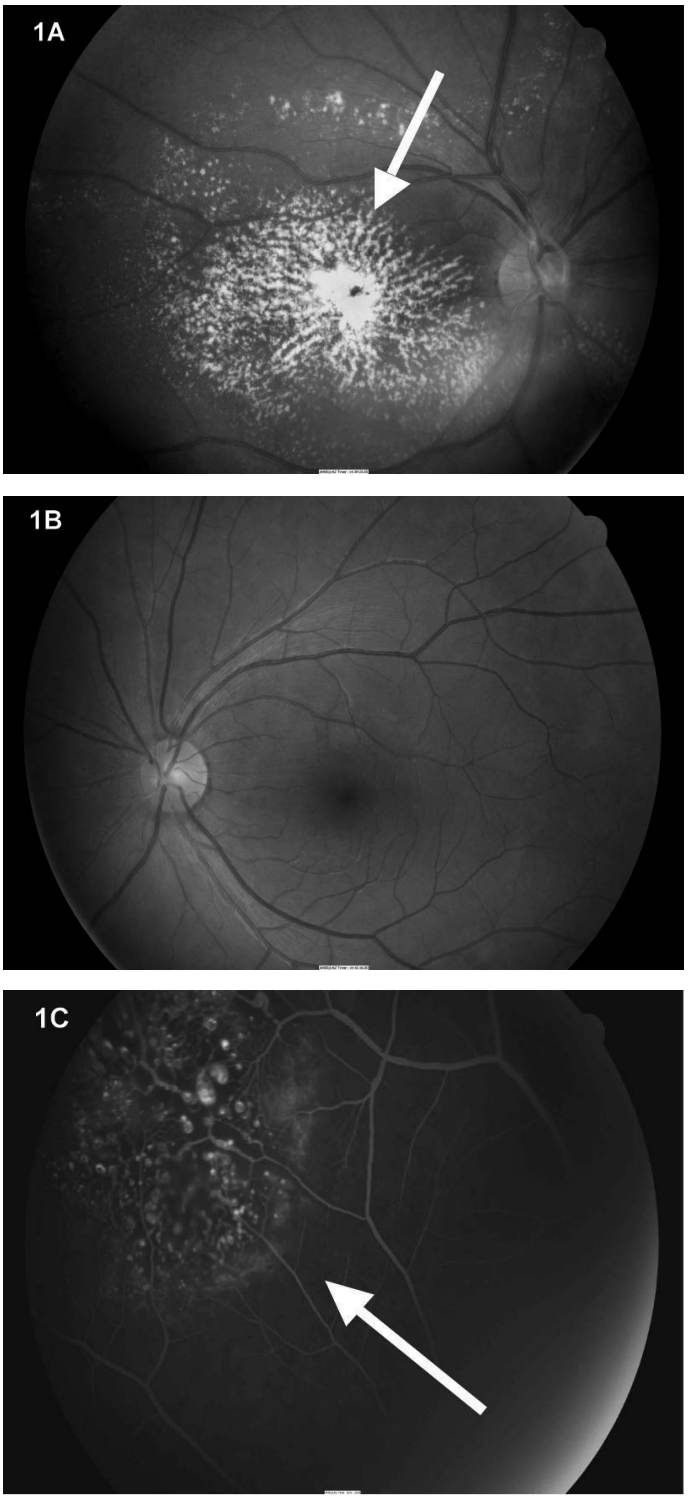

Figure 1. Fundus photographs show (A) a right eye macular star exudate; and (B) no abnormality in the left eye. Flourescein angiography (C) clearly shows an area of retinal vascular anomaly in the upper temporal quadrant of right eye.

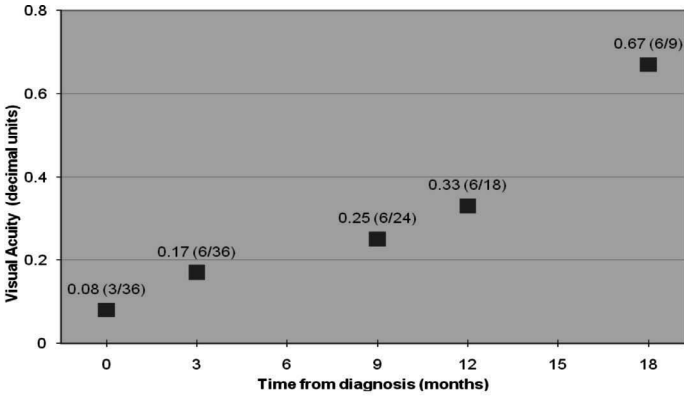

Figure 2. Improvement in visual acuity over time expressed in decimal units (Snellen equivalents shown).

At his last visit, visual acuity in the right eye had improved to 6/9 which was commensurate with an improvement in retinal appearance as most of the exudate at the posterior pole had resolved (Figure 3).

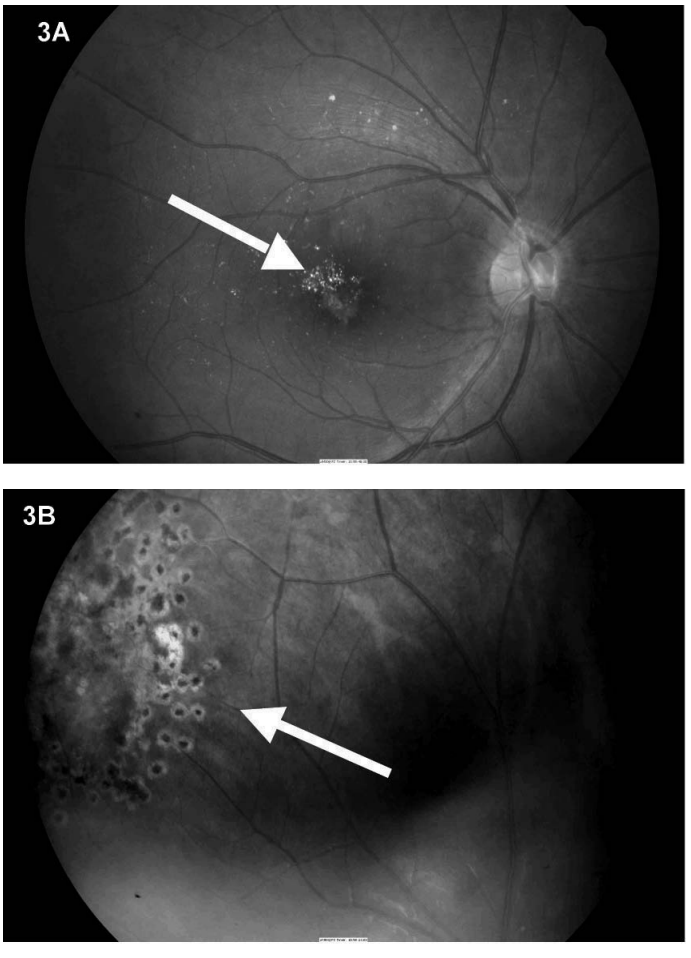

Figure 3. 18 months following initial presentation, (A) shows the remaining exudate at the posterior pole and (B) shows the area of retinal telangiectasia following multiple laser treatments.

DISCUSSION

This male patient presented before the age of 20 years with a complaint of unilateral vision loss. There was no systemic disease to indicate a cause for his visual disturbance, and fundus examination findings were typical of progressive Coats disease.

The vision loss described by the patient was due to the large area of exudate deposited at the posterior pole. The exudate had migrated from an area of retinal telangiectasia that had formed aneurysms with subsequent haemorrhaging and lipid leakage. It is most likely that the area of congenital retinal telangiectasia had remained quiet throughout early childhood, giving rise to a later presentation than the reported peak incidence of 6 to 8 years.[6]

The leakage of lipid in the posterior pole most often causes initial visual disturbances in Coats disease, particularly when the fovea is affected, as was noted in this case. The arrangement of nerve fibres in the fovea, which run parallel to the retinal surface, allow exudates to migrate there and remain in the outer plexiform layer.[2] This parallel radial distribution of exudate can be likened to a star – hence the term macular star, which can also be seen in other retinal disorders such as hypertensive and diabetic retinopathies.[2] This can occur when there is chronic retinal oedema and deposition of hard exudates around the fovea.[2] The macular region is slow to absorb the leaking exudates and the build-up of this lipid exudate can lead to an exudative retinal detachment causing severe and permanent vision loss.[5]

The patient in this case study presented at stage 2B (see Table 1). His peripheral retina showed signs of retinal telangiectasia and marked exudation at the fovea. This early presentation allowed treatment to commence immediately and arrest leakage from the area of retinal telangectasia via argon laser PRP. Table 1 shows how those presenting with stages 1 to 3 have the best visual prognosis due to viable treatments of the disease.[7] If left longer, the progression of the disease leads to a total exudative detachment with permanent vision loss and further complications of potential glaucoma.[10]

CONCLUSION

This case highlights an appropriate management regime applicable to an early-presenting case of Coats disease. In such instances, a successful visual outcome can result. This case serves as an important reminder that a dilated fundus examination in all patients, particularly children, is essential in order to adequately detect retinal abnormalities. This is of particular importance in patients with retinal telangiectasia, although initially asymptomatic, if left untreated can lead to severe visual disturbances.

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