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Bilateral Coats Disease in a 2-Year-Old Boy (Case Report)

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Abstract **Case Report**

***Introduction*:** Coats disease is a rare idiopathic retinal vascular disorder characterized by telangiectasia, retinal and sub retinal exudates, potentially leading to retinal detachment, neovascular glaucoma, and vision loss. It typically presents in male children with a mean age of onset of 5 years and is usually unilateral. This report describes a case of bilateral Coats disease in a 2-year-old boy. The objective is to outline the clinical manifestations and management of this disease in the context of its rare presentation in both eyes and at a young age. ***Results*:** A 2-year-old boy presented with bilateral leukocoria and inward deviation of his left eye. Anterior segment examination revealed ectropion uveae with anterior polar cataract in the right eye and ectropion uveae with a subluxated lens in the left eye. Fundus examination showed hard exudates and fixed yellow retinal detachment in both eyes. B-scan excluded calcifications. Ocular magnetic resonance (MRI) imaging indicated bilateral exudative retinitis. The patient undergone pars plana vitrectomy in his left eye and is programmed for pars plana vitrectomy of the fellow eye. ***Conclusion*:** This case underscores the importance of considering Coats disease as a differential diagnosis in cases of bilateral leukocoria in infants, following the exclusion of retinoblastoma.

Keywords: Retinal Disease, Telangiectasis, Leukocoria, Exudates.

INTRODUCTION

Coats disease, a clinical entity known from early 1900s, remains a diagnostic challenge due to its varied presentation and resemblance to other vascular and exudative retinopathies. Coats disease was described by George Coats in 1908 as unilateral retinal vascular abnormality with exudation mainly in young males [1]. A similar condition with multiple aneurysm-like degeneration of the retina was discussed further by Leber [2]. In 1955, Reese recognized the similarities between both findings and summarized both entities as part of a spectrum in retinal telangiectasia, which results redundantly in progressive exudation and finally in retinal detachment [3].

This sporadic, non- hereditary condition is predominantly unilateral (95-100%) and more common in males, with no clear explanation for theses patterns [4]. In younger patients, the disease tends to present more severely and often leads to poorer visual outcomes [5]. Management strategies vary depending on the stage of the disease, and typically involves interventions such as laser photocoagulation or cryotherapy, aiming to prevent further exudation and to preserve vision.

Where the initial descriptions by Coats, as well as subsequent statements by Leber and Reese, provided the fundamental knowledge, modern clinical experience would suggest that such variable presentations still have the potential to pose a diagnostic challenge in rare instances of both bilateral involvement and early onset.

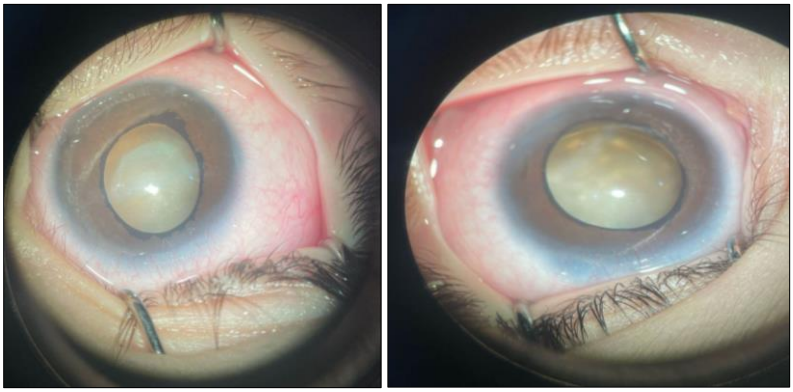
This case report will try to contribute to the literature by discussing the clinical challenges and the management approach toward such atypical presentations, aiming improvement of diagnostic accuracy and optimization of the outcomes of similar cases.

CASE REPORT

A 2-year-old boy was presented with bilateral leukokoria and inward deviation of his left eye, first noted by his parents at 6 months of age (**Figure 1)**. The child's visual acuity could not be documented as the child was uncooperative during the initial examination. Under anesthesia, anterior segment examination revealed an ectropion uveae with anterior polar cataract in the both eyes **(Figure 2)**. Fundus examination revealed yellowish hard exudates, retinal telangiectasia,

and total retinal detachment in both eyes. The general physical developmental milestones of the patient were unremarkable.

Figure 1: Bilateral leukocoria and inward deviation of the left eye



2A **2B**

Figures 2:

- **A**: *image of anterior segment of the right eye showing ectropion uvea associated with cataract*
- ***B****: Image of the left eye showing ectropion uvea as well, with noticeable hard yellowish exudates posterior to the crystalline lens.*

Imaging studies confirmed no calcifications with bilateral retinal detachment **(Figure 3)**. Due to the child's uncooperative behavior, fundus fluorescein angiography and OCT could not be performed. The ocular MRI revealed hyperintense bilateral intraocular lesions (**(Figure 4)** with no retrobulbar extention, thereby confirming stage 3B bilateral Coats disease according to the classification system by Shields and colleagues **(Figure 5)** [6].

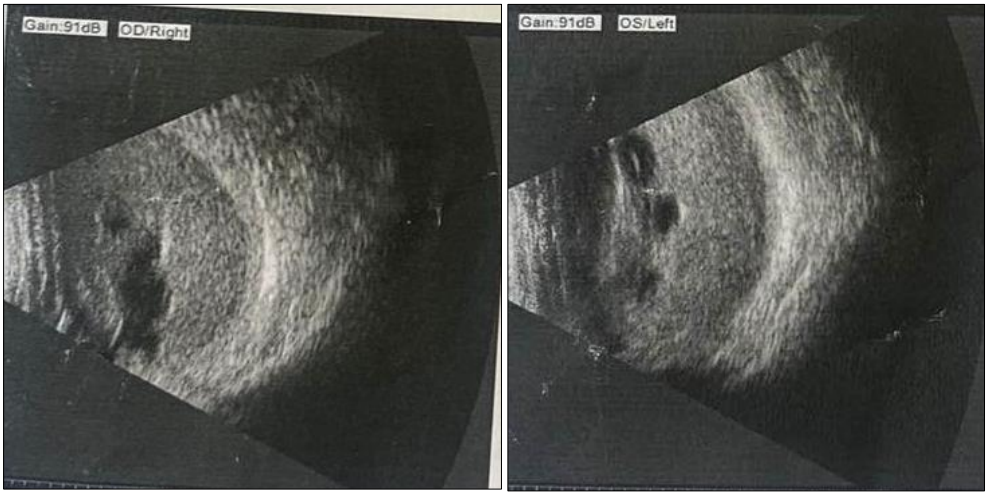
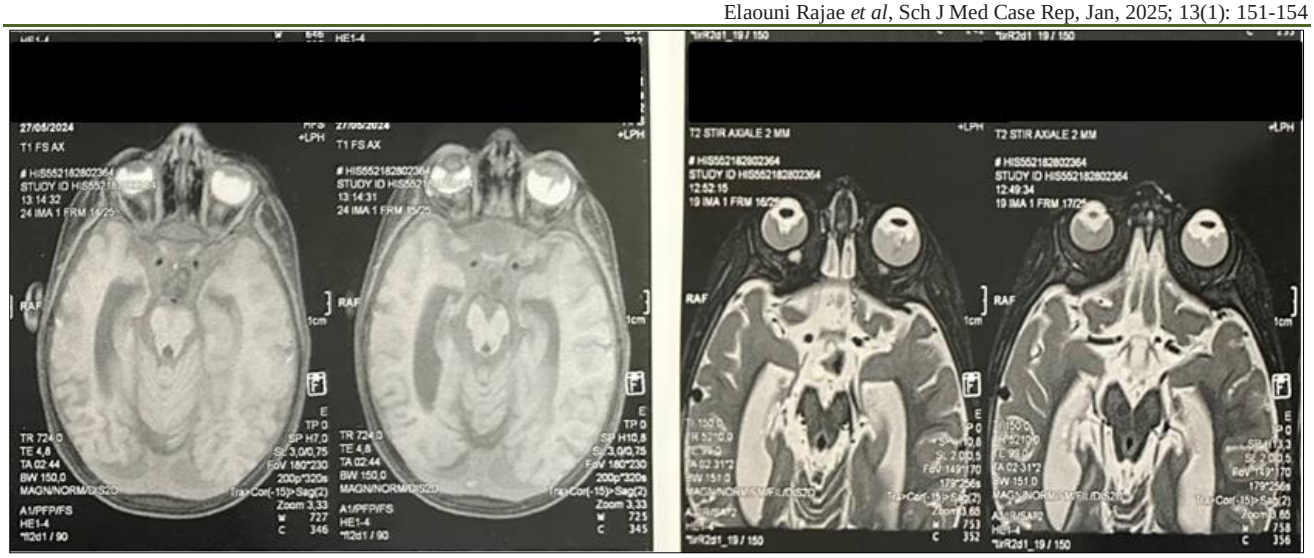


Figure 3: B-scan of both eyes ruled out any calcifications, providing further diagnostic clarity



Figures 4: The T1-weighted and T2-weighted MRI images reveal hyperintense lesions in both eyes

Stage	Findings
1	Retinal telangiectasia only
2	Telangiectasia and exudation
3a	Exudative subtotal retinal detachment
3b	Exudative total retinal detachment
4	Total retinal detachment and glaucoma
5	Advanced end-stage disease

Shields, JA et al. Classification and Management of Coats Disease: the 2000 Proctor Lecture. Am J Ophthalmology. 2001;31(5):572-583.

Figure 5: Shied's classification of Coats disease

Left eye pars plana vitrectomy was done, and surgical intervention for the right eye is scheduled. On follow-up, successful retinal reattachment could be confirmed by B-scan (**Figure 6)**, although the long-term visual prognosis remains guarded.

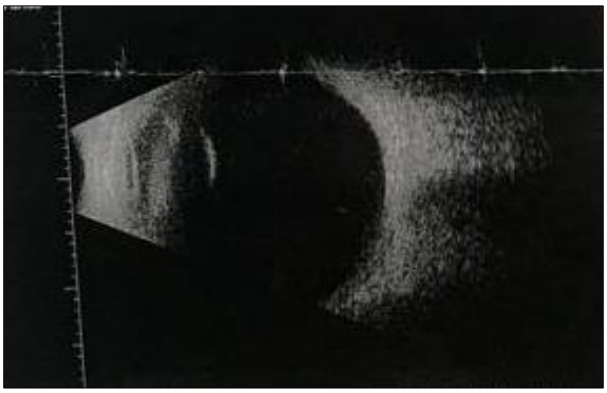


Figure 6: B-scan of the left eye showing a complete reattached retina

DISCUSSION

Coats disease, first reported by George Coats in 1908, for all practical purposes is a unilateral disease of young males [7], with a predilection for declaration at about 10 years. Bilateral involvement, as in our patient, is rare, occurring in less than 5% of cases [8]. If not treated, the disease will frequently progress to significant visual loss, and in far-advanced cases, enucleation [9].

Coats disease can present a significant challenge, especially in differentiating it from retinoblastoma. In the case of our patient, the early age at onset and bilateral involvement were both unusual for Coats disease. Imaging was key to making a definitive

diagnosis. B-scan and MRI both helped eliminate the possibility of calcification or solid masses which are often present in retinoblastoma tumors.

Management for our patient included pars plana vitrectomy with successful reattachment of the retina. The visual prognosis remains guarded given the extent of the disease; However, surgical intervention was needed to prevent the formation of neovascular glaucoma, a common complication which is painful. Silodor *et al*., [10], demonstrated that early intervention with drainage of subretinal fluid and supplemental cryotherapy or photocoagulation can reduce the risk of neovascular glaucoma.

Poor visual outcomes are common since subretinal fluid and subfoveal fibrosis often persist after Coats disease has been treated. Even with successful reattachment, the long-term visual prognosis remains poor due to chronic exudation and fibrosis causing irreversible damage.

CONCLUSION

Coats disease is a challenge for diagnosis because it seems to mimic other grave ophthalmologic diseases in children, particularly retinoblastoma. Misdiagnosis can lead to wrongful enucleation, as exudates in Coats disease may resemble calcifications seen in retinoblastoma, particularly in bilateral presentations. This case highlights the importance of thorough clinical examination and advanced imaging, particularly ocular MRI, in distinguishing Coats disease from other conditions, thus avoiding unnecessary surgical interventions. Early and precise diagnosis is crucial for appropriate management and better visual outcomes in affected children.

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