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Case Report

An Unusual Presentation of Coats Disease in a 1 Year Old Girl

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Abstract

We report a 1 year old female who unusually presented with retinal hemorrhages, neovascularization, subtotal serous retinal detachment and no exudation in one eye at her first visit, and found to have a classic "Coats disease".

A 1 year old female presented with a history of exodeviation of one eye, fundoscopic findings showed retinal hemorrhages, disc and retinal neovascularization with sub total serous retinal detachment and no exudation. Few weeks later during fundus examination under general anesthesia found to have a typical "Coats disease", showing at that moment massive exudation.

Laser photocoagulation alone to vascular lesions was effective enough to reduce the exudative changes and sub retinal fluid suppressing the worsening of the disease and recurrence of exudation in the first 5 months.

Typically, vascular telangiectasias and exudation are the hallmark of "Coats disease", but our patient's first presentation was atypical showing retinal hemorrhages, neovascularization and serous retinal detachment with no exudation. After laser photocoagulation therapy alone showed great improvement with no recurrence of the disease despite the advanced stage.

Keywords: *Coats' Disease; Treatment; Follow-Up; Atypical Presentation; Laser Photocoagulation; Retinal Hemorrhages*

Introduction

Coats disease its and exudative retinopathy characterized by idiopathic retinal telangiectasia, retinal exudation and retinal detachment and was first described by George Coats, in 1908 [1]. It may be present as earlier as the first month of life and up to the eight decade, but typically appears in childhood (6 - 16 years of age) and the most common signs are leukocoria, strabismus and/ or visual impairment. Males are more affected (70 - 90%) and no racial or ethnic predilection has been shown. It is sporadic and nonhereditary [2].

Clinical findings are characterized by changes of the retinal vessels that shows an irregular caliber, focal telangiectasias, aneurysmal dilatations ("light bulbs") and sheathing by yellow cholesterol deposits associated with large amounts of yellowish subretinal and intraretinal exudates [3].

Ablation of abnormal retinal vessels, either by laser photocoagulation or by cryotherapy in earlier stages and surgery for advanced stages of the disease are the standard treatment. In the recent years vascular endothelial growth factor (VEGF) showed to play an important role on the disease [4-6]. If left untreated the disease may progress to retinal detachment, glaucoma, and painful eye and may even require enucleation.

The hallmark finding in the disease are the vascular telangiectasias and massive exudation that actually do not typically bleed. In this article we report a 1 year old female who unusually presented with retinal hemorrhages, neovascularization and subtotal serous retinal detachment with no exudation in one eye, and found to have a classic "Coats disease".

Case Report

Clinical History

A 1 -year-old girl was referred to our clinic with a 3 weeks history of exodeviation and apparent visual impairment of her left eye noted by the mother. The girl is the first child in the family. She was born at term (40 weeks) by vaginal delivery, and her birth weight was 3790 g. with no history of supplemental oxygen therapy. Past medical and family histories were unremarkable and there was no history of previous ocular disease or treatment.

Visual acuity was not measured because of young age but was not able to fix or follow objects. Divergent strabismus of 5 - 10 degrees was documented and slit lamp examination of anterior segment was normal. IOP was 15 mm HG using a rebound tonometer (ICARE Finland Oy, Vantaa, Finland).

Fundus biomicroscopy revealed retinal and disc neovascularization, marked venous tortuosity with an irregular "sausage like" dilatations of arterioles, multiples intra and pre retinal hemorrhages in all quadrants and a subtotal flat serous retinal detachment, the vitreous cavity was clear (Figures 1 and 2). There were no apparent signs of trauma. Right eye was completely normal. Laboratory testing, including blood chemistry, white cell counts, glucose/protein monitoring, and coagulation panel were within expected normal ranges. IgG and IgM antibodies were not detected for Toxoplasma Gondii and HSV I/II. Ultrasound examination demonstrated an inferior retinal detachment partially involving the macula, no other lesions were found (Figure 3).

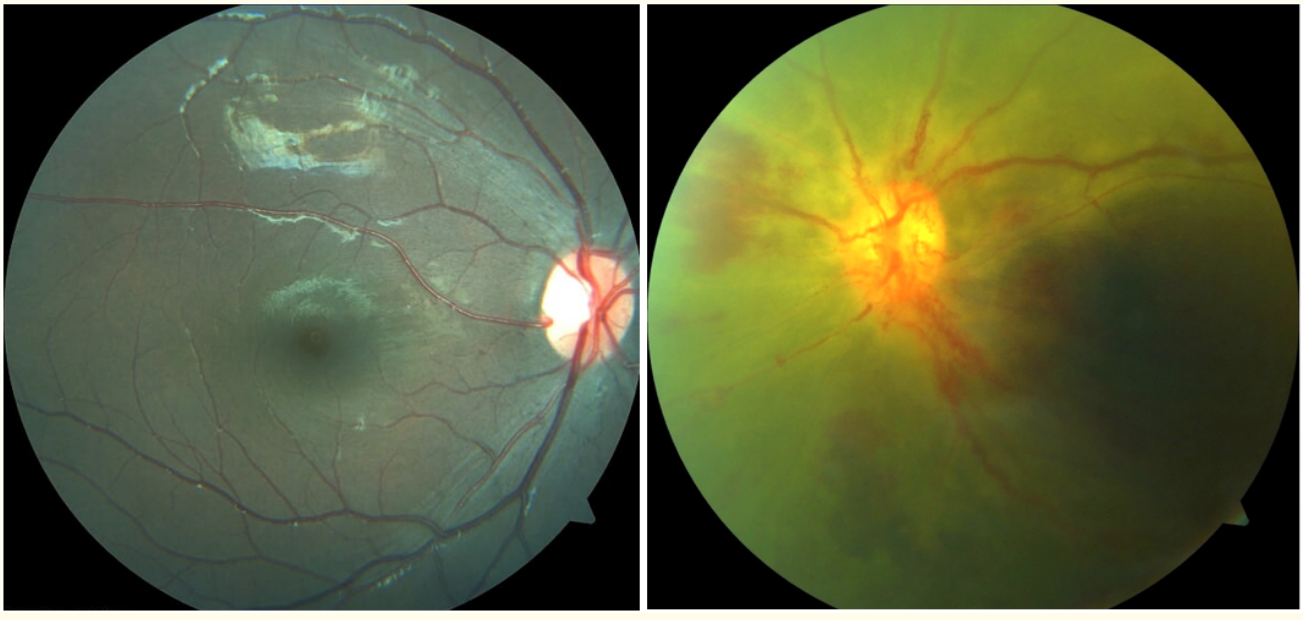

Figure 1: *Fundus biomicroscopy showing normal right eye and left eye showing retinal and disc neovascularization, marked venous tortuosity, multiples intra and pre retinal hemorrhages in all quadrants and an serous inferior retinal detachment, vitreous was clear.*

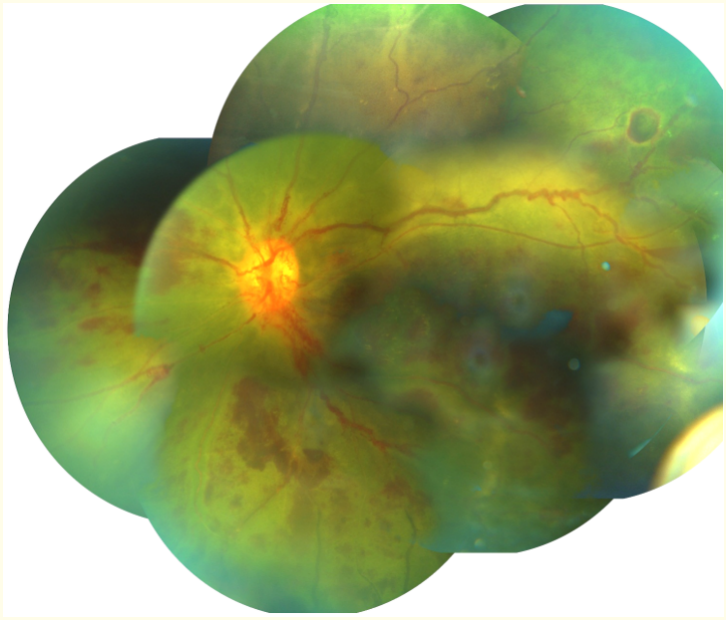

Figure 2: *Left eye fundus evaluation showing retinal and disc neovascularization, marked venous tortuosity with an irregular "sausage like" dilatations of arterioles, multiples intra and pre retinal hemorrhages in all quadrants and a sub total exudative retinal detachment, vitreous was clear.*

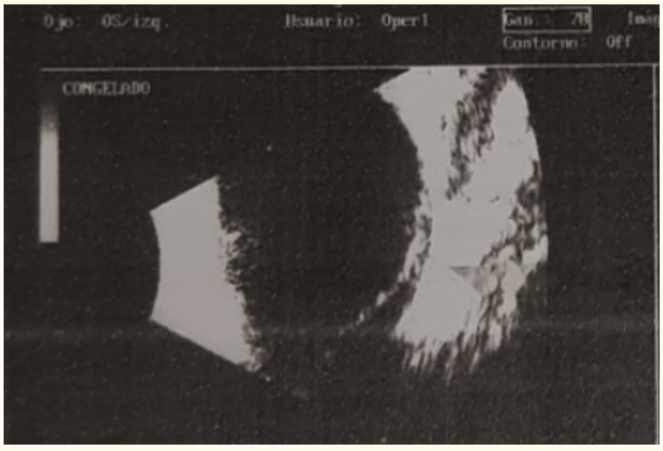

Figure 3: *Ultrasound examination of the left eye demonstrated an inferior retinal detachment partially involving the macula, no other lesions were found.*

3 weeks later

The child was evaluated under general anesthesia and the indirect ophthalmoscopic examination, revealed severe lipid exudation around the disc and temporal arcades and multiple telangiectatic vessels on the inferior and temporal retina with exudative retinal detachment (Figure 4). A clinical diagnosis of stage 3 A 2 "Coats" disease was made and laser photocoagulation was applied to all telangiectatic vessels using a 532 nm diode-laser (OcuLight Tx; Iridex Corp, Mountain View, CA, USA).

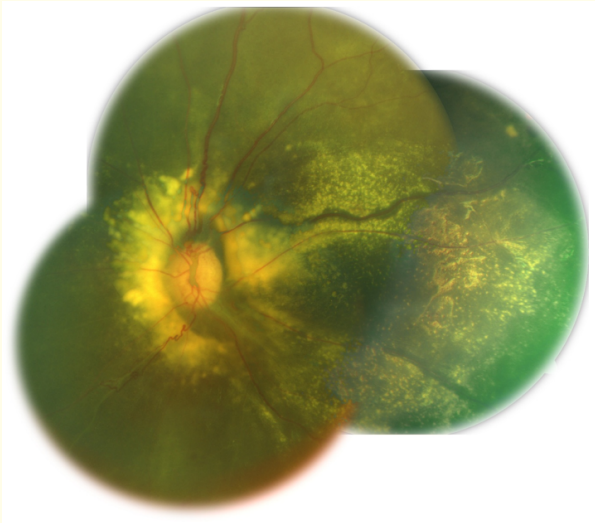

Figure 4: *Fundus examination of the left eye, 3 weeks later, showing extensive retinal exudation and inferior exudative retinal detachment.*

Five months after presentation, the exudates had decreased and there was resolution of the angiomatous changes, exudation and sub retinal fluid (Figure 5 and 6). There was no recurrence of disease after 5 months of follow-up.

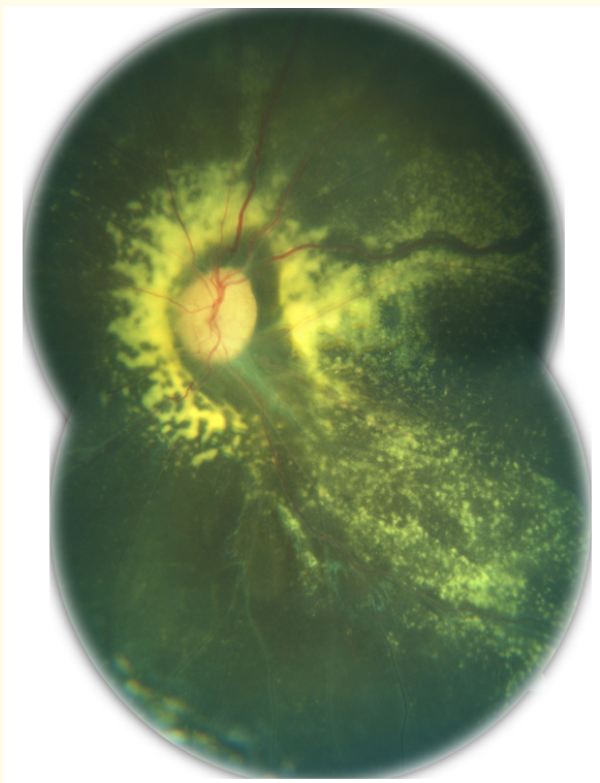

Figure 5: *Fundus examination of the left eye, 1 month after laser therapy, showing improvement of exudation and sub retinal fluid.*

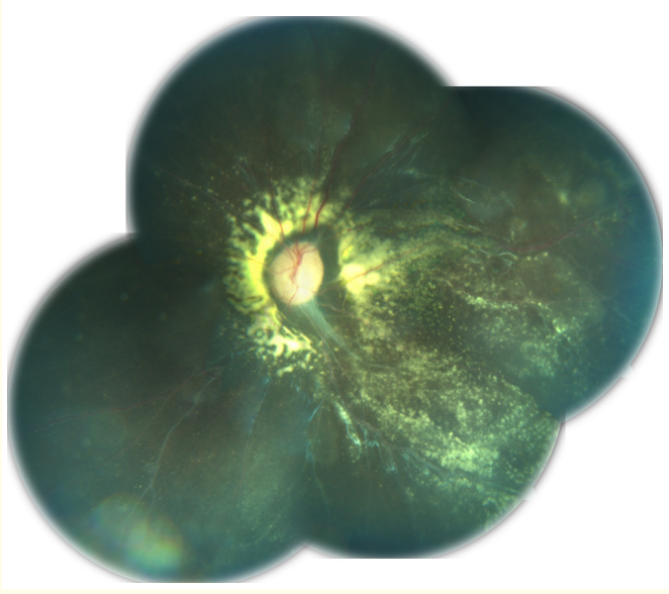

Figure 6: *Fundus examination of the left eye, 5 months after laser therapy, showing improvement of exudation and sub retinal fluid, associated with sclerotic vessels and glial tissue inferior to the disc.*

Discussion

This case presented an initial unusual presentation of the disease, since hemorrhages and retinal detachment without exudation are atypical for childhood "Coats" and when are present, are accompanied with rubeosis iridis and neovascular glaucoma which are also rare.

The hallmark finding of the disease are the vascular telangiectasias and massive exudation that actually do not typically bleed [3,7]. In this article we report a 1 year old female who unusually presented with retinal hemorrhages, neovascularization, subtotal flat serous retinal detachment and no exudation in one eye and found to have a classic "Coats disease". The sex and age of our patient are not typical for the disease since "Coats" typically occurs in male patients and the usual age of presentation it's from 6 to 15 years of age [2].

The standard treatment of "Coats" disease includes laser or cryoretinopexy to the telangiectatic vessels [8] and anti-VEGF injections seems to be an adjunctive treatment for these methods, according to some reports [4-6,9]. Surgery are suitable for more advanced cases. Laser photocoagulation in the early stages, either alone, or in combination with cryotherapy, has proven to be effective especially in young cases [8].

In our case, laser photocoagulation alone to vascular lesions was effective enough to reduce the exudative changes and sub retinal fluid and more importantly, have suppressed the worsening or recurrence of exudation for at least 5 months. The visual function seemed not improved but the treatment showed a dramatic resolution of exudation and the sub retinal fluid during the follow up with only laser photocoagulation.

Even though many specialist are more aggressive for the treatment of these patients at this stage, using anti VEGF therapy with laser and/or surgery [5,6,9]; our patient showed a dramatic response with laser therapy alone, with clinical improvement of the disease and no early recurrence. Five months of follow up it's a short time to establish the effectiveness of our treatment despite the advanced stage of the disease and no practicable conclusion could be drawn from our report because of his own nature.

We would like to highlight the atypical initial presentation of our case, that weeks later showed a classic presentation and how rare it's to find it at this stage.

Conclusion

In conclusion, for the best of our knowledge, this is an unusual report of an initial atypical presentation of "Coats disease" with retinal hemorrhages, neovascularization and subtotal serous retinal detachment without exudation, that was successfully treated with laser photocoagulation alone. Conventional laser photocoagulation therapy still seems to be effective for more advanced disease like this.

Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient parents has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patient parents understand that their names and initials will not be published and due efforts will be made to conceal their identity but anonymity cannot be guaranteed.

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Conflicts of Interest

The authors have no financial disclosures nor conflict of interest.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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