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CASE REPORT

Atypical Presentation of Coats Disease in a Teenager: A Case Report

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Abstract

Coats disease is an idiopathic retinal telangiectasia with retinal exudation. It has a bimodal distribution – in children, with extensive disease and poor prognosis, and in adults, after the age of 35 years, with focal involvement and favorable outcomes. We report the case of a 15-year-old boy, whose routine fundus examination incidentally revealed a localized lipid exudation with telangiectatic vessels, without any retinal detachment or macular involvement. Fluorescein angiography showed classic "light bulb" appearance of the vessels. A diagnosis of Coats disease was made and the lesion was treated with laser photocoagulation and intravitreal ranibizumab. This case is unusual as despite occurring in a child, the features are similar to the rarer adult onset variant, with a better prognosis.

Introduction

Coats disease is an idiopathic retinal telangiectasia with subretinal or intraretinal exudation, first described in 1908.[1] It has two variants – in children, within the first two decades of life, with extensive severe disease and poor prognosis; and in adults, after the age of 35 years, with focal involvement and favorable outcomes. The presenting features in young patients are usually severe and may end in a painful blind eye. We report an unusual case of Coats' disease that presented in a 15-year-old boy but had limited disease involvement without any visual deterioration.

Case Description

A 15-year-old male patient was referred to our tertiary care center with a suspected mass lesion detected incidentally on dilated fundus examination of his left eye. He had no visual complaints and no significant family history. On examination, he had a best-corrected visual acuity of OD 20/20 and OS 20/20 and a normal anterior segment, with briskly reacting pupils and a normal intraocular pressure of 14 and 16 mmHg on applanation tonometry. Dilated fundus examination was normal in the right eye but revealed a well-defined subretinal exudation in the superonasal quadrant in the left eye with a normal posterior pole [Figure 1a and b].

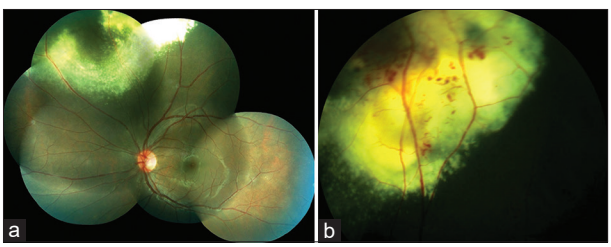

Figure 1: (a) A well-defined subretinal exudation, between the equator and the ora, in the superonasal quadrant in the left eye with a normal posterior pole. (b) The lesion had telangiectatic vessels with multiple small pre-retinal hemorrhages and some surrounding sub- and intraretinal exudation

There was no macular involvement, retinal detachment or traction bands, and no other lesion elsewhere. Our differentials included Coats' disease, granuloma secondary to sarcoidosis or tuberculosis, capillary hemangioma associated with Von Hippel-Lindau syndrome, late-onset retinoblastoma, retinopathy of

prematurity, and familial exudative retinopathy. Ultrasound B-scan of the same eye is shown in Figure 2.

Optical coherence tomography of the macula was normal with preserved architecture and central macular thickness of 263 microns. Autofluorescence and fluorescein angiography of the involved eye are shown in Figure 3. The latter revealed the classical "light bulb" appearance of the telangiectatic vessels and hypofluorescence in the areas of exudation in the early venous phase and leakage and staining in the area of exudation in the late phases.

Extensive blood work was normal [Table 1]. Other tests such as Mantoux, contrast-enhanced computed tomography chest, and magnetic resonance imaging brain with orbit revealed no significant abnormality. A diagnosis of atypical Coat's disease was made and management was conservative. Sectoral laser photocoagulation with a prophylactic laser barrage was done to ablate the abnormal vessels and limit the exudation from gravitating to the posterior pole, respectively. Intravitreal ranibizumab was given to regress the vasculature. The lesion responded to treatment, with decrease in telangiectasia and bleeds, and loss of volume of the exudation [Figure 4] but did not regress fully. The patient has remained on regular follow-up and has remained stable at 6 months.

Discussion

Coats disease is idiopathic retinal telangiectasia with exudation, which can have variable severity and presenting features.[1] It is bimodal in distribution; the commoner classical type seen in young males with unilateral, progressive, and visual threatening eye disease[1] and the less common adult-onset disease, which has a localized disease and milder course.[2]

The presenting features in classical Coats disease are usually severe, ranging from xanthocoria, strabismus, and pain to severe vision loss. Typical signs include irregular "light bulb"-shaped telangiectatic vessels, subtotal or total exudative retinal detachment, and peripheral capillary non-perfusion areas[3] and have a stage-wise staging system proposed by Shields *et al.*[4] This disease is classically seen in the first decade of life, peaking between

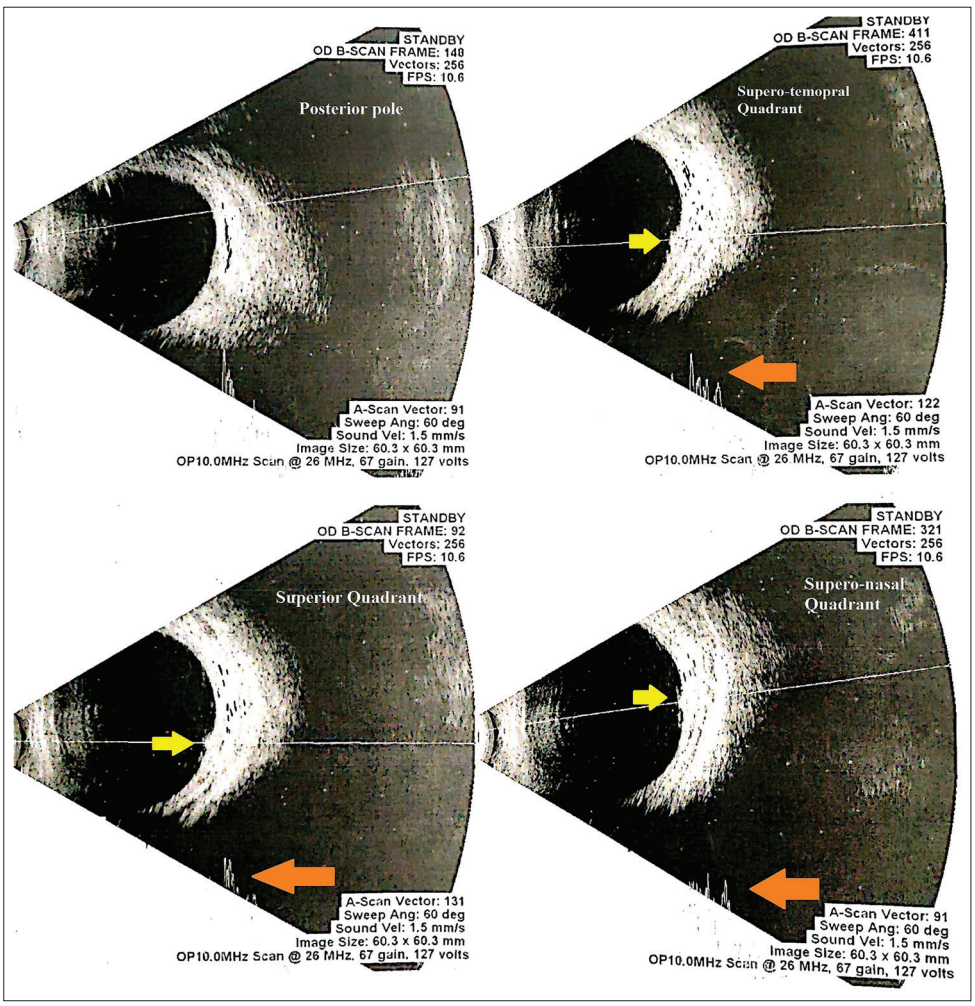

Figure 2: Ultrasound B-scan of the left eye showed a hyperechoic lesion (yellow arrows) with high spikes (orange arrows) in the superior, superonasal, and superotemporal quadrant without any overlying detachment

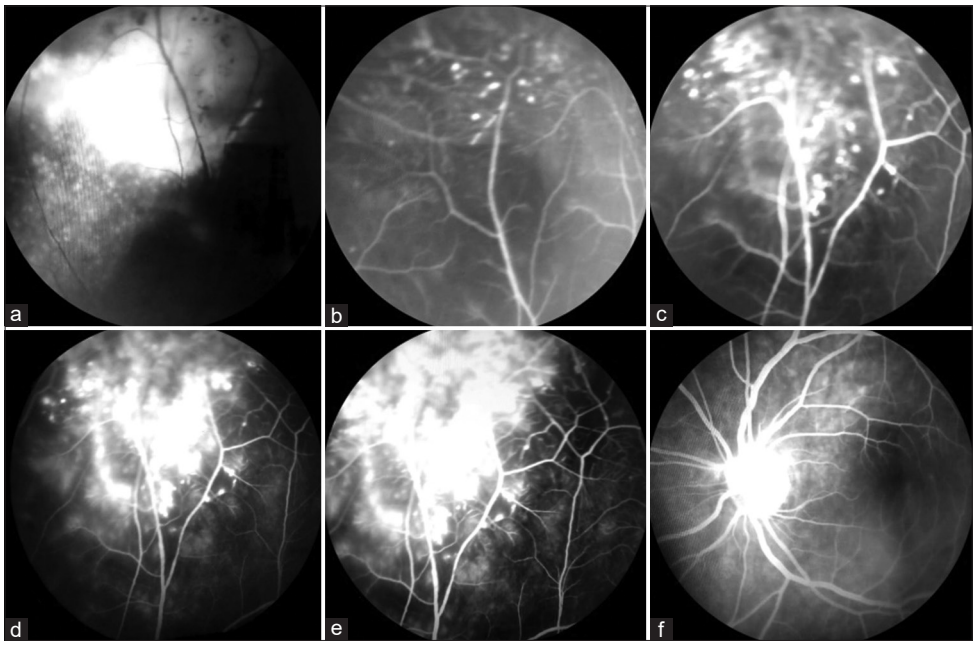

Figure 3: (a) Enhancement of the lesion on autofluorescence. Fundus fluorescein angiography was revealed hyperfluorescence of the telangiectatic vessels with bulbous terminals, similar to a "light bulb" and hypofluorescence in the areas of exudation in the early venous phase (b and c), and leakage from these vessels and staining in the area of exudation in the late phases (d and e). The disk and macula were normal (f)

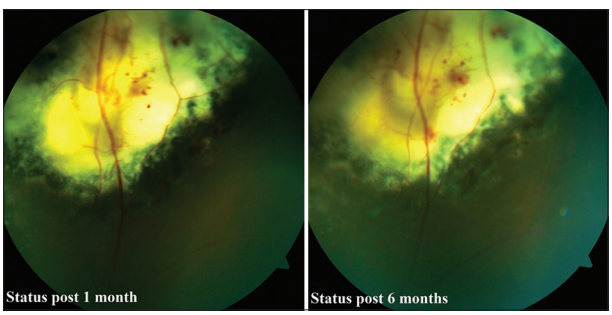

Figure 4: Post-treatment photos at 1 month and 6 months. There were decrease in telangiectasia and pre-retinal bleeds, and loss of volume of the exudation. While the lesion did not regress fully, it has remained stable on follow-up

6 and 8 years of age;[5] features previously described in the second decade are comparable to the childhood type, with telangiectasia and exudative detachment involving the fovea,[6] subfoveal exudation,[7] and macular edema and exudates.[8] Complications from long-standing disease include vitreous hemorrhage, retinal macrocysts, and neovascular glaucoma.[1]

Coats disease of adult onset is usually diagnosed above 35 years of age.[2] These patients may a have limited and peripheral area of retinal telangiectasia and exudation, slower disease progression, and resultant good visual acuity. This variant has been linked to systemic diseases such as diabetes, hypercholesterolemia, and hypertension.[5]

Our case was unusual because our patient was 15 years old but had presenting features similar to the adult variety, leading to a diagnostic dilemma. With extrafoveal exudates and telangiectasia, he had Stage 2A disease as per the Shield's classification. There were no retinal detachment and foveal involvement, and no systemic affliction. Our management involved was conservative with long-term follow-up.

Management in Coats disease is stage wise.[1,9] Observation can be done for patients with good vision and telangiectasia with little to no exudation. Laser photocoagulation may be done for Stages 1–3a with no or shallow retinal detachment. Cryotherapy is recommended for peripheral lesions and retinal detachment up to Stage 3B. Vitreoretinal surgery may be done under guarded prognosis for advanced diseases with extensive retinal detachment. Ultimately, enucleation may be the only option for neovascular glaucomas and painful blind eyes. Anti-VEGFs are under research for their ability to regress abnormal vessels and decrease exudation and macular edema.[6-8]

The peripheral lesion in our young patient responded well to sectoral laser photocoagulation and intravitreal ranibizumab, with decrease in telangiectasia and loss of volume of the exudation. Since these exudations have a tendency to gravitate toward the posterior pole, a prophylactic laser barrage was also done, and the patient has remained on regular follow-up, with no change at 6 months.

Statement of Equal Authors' Contribution

All the authors meet the requirements for authorship. The manuscript has been read by all the authors and each author believes that the manuscript represents honest work. There has been no prior publication of this work and none of the authors

Table 1: Hemodynamic parameters

Test	Value	Reference
Hemoglobin	15.3 g/dl	13–17
Total leukocyte count	6×10^3/µl	4–10
Hematocrit	44.9%	40–50%
RBC	5.10 million/mm^3	4.50–5.50
MCV	88.7FL	83–101
MCH	30.3pg	27–33
Platelet count	130×10^3/µl	150–410
ESR	02 mm/1^{st}h	0–10
Neutrophils	46.1%	40–75
Lymphocytes	41.9%	20–45
Monocytes	8.1%	01–10
Eosinophils	3.7%	01–06
Basophils	0.2%	00–02
Creatinine	0.79 mg/dl	0.9–1.3
Uric acid	5.2 mg/dl	3.5–7.2
Calcium	9.9 mg/dl	8.8–10.6
Sodium	143 mmol/L	136–146
Chloride	106 mmol/L	101–109
Blood urea	16 mg/dl	17–43
Serum bilirubin (total)	0.77 mg/dl	0.3–1.2
AST	27 U/L	3–50
ALT	40.30 U/L	3–50
Serum albumin	4.14 g/dl	3.5–5.2
Serum globulin	2.56 g/dl	3–4.2
Total cholesterol	146 mg/dl	<200
Serum triglycerides	87 mg/dl	<150
HDL	43.5 mg/dl	40–60
LDL	93 mg/dl	<100
VLDL	17.4 mg/dl	06–30
Serum potassium	5.5 mmol/L	3.5–5.5

RBC: Red blood cell count, MCV: Mean corpuscular volume, MCH: Mean corpuscular hemoglobin, ESR: Erythrocyte sedimentation rate, AST: Aspartate transaminase, ALT: Alanine transaminase, HDL: High-density lipoprotein, VLDL: Very low-density lipoprotein, LDL: Low-density lipoprotein

have any financial interest in any of the materials and methods used in this study. I would like to mention that the manuscript, to the best of the author's knowledge, does not infringe upon any copyright or property right of any third party.

Conclusion

Coats disease is classically described in the first decade of life, but it should be still kept in mind as a differential in older children with an atypical presentation. Multimodal imaging is helpful in its diagnosis and a wide range of therapeutic options with long-term follow-up may preserve vision in such cases.

Declaration of Consent

Written consent to publish was obtained from the patient.

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Conflicts of Interest

None declared.

References

1. Sen M, Shields CL, Honavar SG, Shields JA. Coats disease: An overview of classification, management and outcomes. Indian J Ophthalmol 2019;67:763-71.
2. Smithen LM, Brown GC, Brucker AJ, Yannuzzi LA, Klais CM, Spaide RF. Coats' disease diagnosed in adulthood. Ophthalmol 2005;112:1072-8.
3. Shields CL, Schoenberg E, Kocher K, Shukla SY, Kaliki S, Shields JA. Lesions simulating retinoblastoma (pseudoretinoblastoma) in 604 cases: Results based on age at presentation. Ophthalmology 2013;120:311-6.
4. Shields JA, Shields CL, Honavar SG, Demirci H, Cater J. Classification and management of Coats disease: The 2000 proctor lecture. Am J Ophthalmol 2001;131:572-83.
5. Mandura RA, Alqahtani AS. Coats' disease diagnosed during adulthood. Cureus 2021;13:e16303.
6. Kodama A, Sugioka K, Kusaka S, Matsumoto C, Shimomura Y. Combined treatment for coats' disease: Retinal laser photocoagulation combined with intravitreal bevacizumab injection was effective in two cases. BMC Ophthalmol 2014;14:36.
7. Lin CJ, Hwang JF, Chen YT, Chen SN. The effect of intravitreal bevacizumab in the treatment of Coats disease in children. Retina 2010;30:617-22.
8. Venkatesh P, Mandal S, Garg, S. Management of coats disease with bevacizumab in 2 patients. Can J Ophthalmol 2008;43:245-6.
9. Adeniran JF, Duff SM, Mimouni M, Lambert N, Ramasubramanian A. Treatment of Coats' disease: An analysis of pooled results. Int J Ophthalmol 2019;12:668-74.

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