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CASE REPORT

# Intravitreal dexamethasone implant with retinal photocoagulation for adult-onset Coats' disease

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**Abstract**
*Purpose* To report a case of adult-onset Coats' disease with secondary retinal vasoproliferative tumor managed with dexamethasone intravitreal implant and retinal photocoagulation.
*Methods* Case study.
*Results* A 41-year-old female with counting finger vision was diagnosed with Coats' disease with secondary retinal vasoproliferative tumor in right eye. Fundus examination revealed exudative retinopathy involving posterior pole and a retinal tumor located in the inferotemporal quadrant. Optical coherence tomography scan confirmed massive exudative neurosensory detachment and fundus fluorescein angiography showed areas of telangiectatic vessels with capillary non-perfusion. Intravitreal injection of dexamethasone implant was done initially followed by laser photocoagulation when the detachment resolved. There was significant improvement in patient's visual acuity with no further recurrence of exudation.
*Conclusion* Intravitreal dexamethasone implant Ozurdex(®) (Allergan, Inc., Irvine, Calif., USA) may be an effective initial therapeutic approach for Coats' disease with massive exudation.

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**Keywords** Exudative vitreo-retinopathy · Idiopathic retinal telangiectasia · Intravitreal steroid implant · Retinal vasoproliferative tumor

## Introduction

Coats' disease is a unilateral (95%), idiopathic retinal vascular abnormality characterized by telangiectatic retinal vessels in association with lipid exudation [1]. Although most cases are diagnosed before the age of 20 years (75%), Coats' disease may manifest in adult patients [2]. Treatment of Coats's disease depends on the stage of the disease. The disease is classified as per the staging proposed by Shields et al.: Stage 1, telangiectasia only; Stage 2, telangiectasia and exudation (2A, extrafoveal exudation; 2B, foveal exudation); Stage 3, exudative retinal detachment (3A, subtotal; 3B, total); Stage 4, total retinal detachment and secondary glaucoma; Stage 5, advanced end-stage disease [3].

Conventional therapeutic strategies involve observation, photocoagulation, or cryotherapy for retinal telangiectasias and surgical intervention for retinal detachment. Adjunctive treatment modalities such as intravitreal steroid triamcinolone and/or anti-VEGF agents may be employed to improve anatomic and visual outcome in cases with a significant amount of subretinal fluid but with suboptimal outcomes [4–7]. Efficacy of dexamethasone intravitreal steroid implant

in the initial management of Coats' disease has been reported recently [8, 9].

Here, we report a case of adult-onset Coats' disease with retinal vasoproliferative tumor (RVT) managed with adjunctive dexamethasone intravitreal implant Ozurdex(®) (Allergan, Inc., Irvine, Calif., USA).

## Case report

A 41-year-old female was referred to our department with a 6-month history of progressive vision loss in her left eye. Best-corrected visual acuity (BCVA) was 20/20 in RE and Counting Finger 1Meter in LE. Anterior segment examination was unremarkable.

Whereas the RE fundus did not show any significant findings, in the LE massive exudation within the inferior temporal quadrant of the retina was evident, with posterior pole involvement and intraretinal lipid accumulation. In addition, a yellowish mass with overlying hemorrhages and neo-vessels was observed in temporal peripheral retina (Fig. 1). SD-OCT confirmed the presence of exudative retinal detachment with macular involvement with numerous hyperreflective intraretinal lesions corresponding to lipids (Fig. 2). B-scan ultrasonography (USG B-scan) showed an intraretinal mass located in the temporal periphery, associated with the exudative retinal detachment. FFA (Fig. 3) confirmed the presence of telangiectatic vessels with peripheral capillary nonperfusion areas; the peripheral temporal mass was identified as a retinal vasoproliferative tumor (RVT). The patient was classified as Stage 3A Coats' disease with RVT. Patient was treated with an intravitreal injection of the sustained-release dexamethasone implant Ozurdex® (Allergan, Inc., Irvine, Calif., USA). One month later, BCVA improved to 20/120 and SD-OCT revealed complete resolution of the exudative retinal detachment with persistent macular edema (central foveal thickness—CFT 513 microns). Intraocular pressure (IOP) was 16 mm Hg. At this point, retinal photocoagualtion was done using laser machine (VISULAS 532 nm, double-frequency Nd-YAG retinal laser, Carl Zeiss Meditec, Germany). Laser was done in one session with an average power of 200 mW, 200 ms duration, and spot size of 200 microns. The entire surface of telangiectatic vessels and/or area of exudative detachment was treated. The clinical endpoint was the complete whitening of the telangiectatic vessels. Treatment was also applied to the avascular retinal areas peripheral to telangiectatic vessels. At 3 months, macular edema had resolved (CFT—253 microns) with BCVA 20/60 and IOP of 14 mm Hg. The volume of the RVT decreased progressively. Through the follow-up of 4 months, no further recurrences of the exudation were noted, and the extensive lipid accumulation progressively regressed with stable vision (Fig. 4). No adverse effect of steroid on IOP or on lens was observed during the course of treatment.

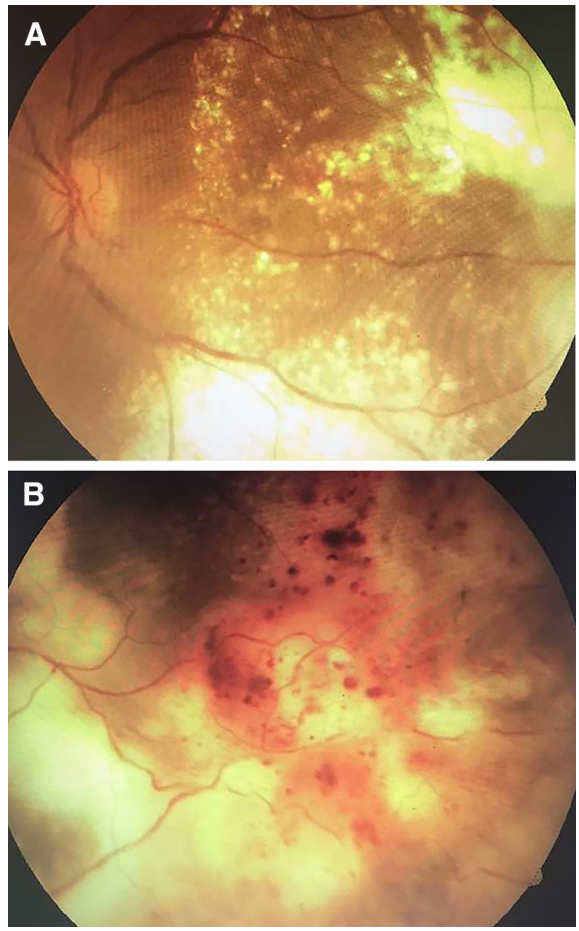


**Fig. 1** Fundus photographs of left eye. **a** Massive intraretinal lipid accumulation in retina. **b** Temporal periphery showing a mass lesion with overlying new vessels and hemorrhages suggestive of secondary retinal vasoproliferative tumor (RVT)

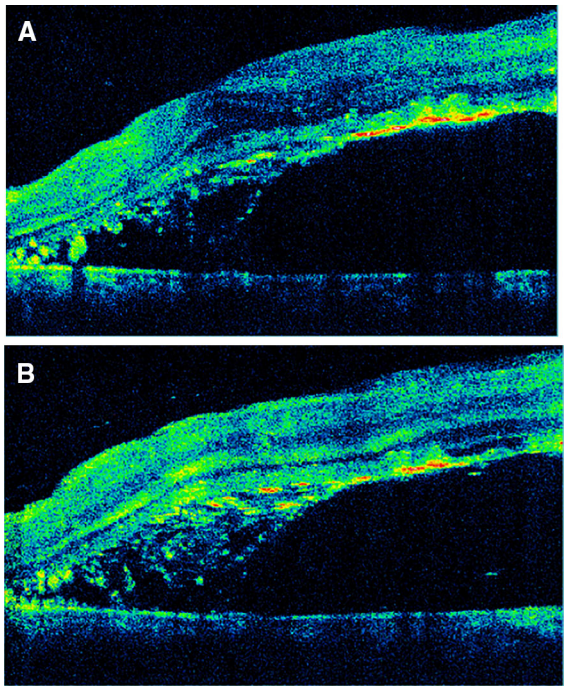


**Fig. 2** SD-OCT image of left eye (**a**, **b**). At presentation showing massive neurosensory detachment, retinal edema, and intraretinal and subretinal lipid exudation

## Discussion

Coats' disease affects men three times as often as women, in the first or second decade of life, with no racial predilection and is usually unilateral (bilateral in 5% of cases) [10]. Retinal vasoproliferative tumors can occur secondarily to preexisting conditions (26%) such as inflammatory, vascular, traumatic, or degenerative retinal diseases as evidenced in our patient who had associated Coats' disease as the primary pathology [11]. This unique presentation in a female patient in fourth decade of life makes an interesting case study.

The pathogenesis of Coats' disease involves increased permeability of the telangiectatic blood vessels, leading to the leakage of lipoproteins into the retina and causing retinal edema. This further leads to exudative retinal detachment. The consequence of this entire process is retinal hypoxia and neovascularization [12]. The treatment goal of Coats' disease is to obliterate the abnormal vasculature and hyperpermeable aneurysmal dilatations. The treatment choice depends on the stage of the disease. Shields and

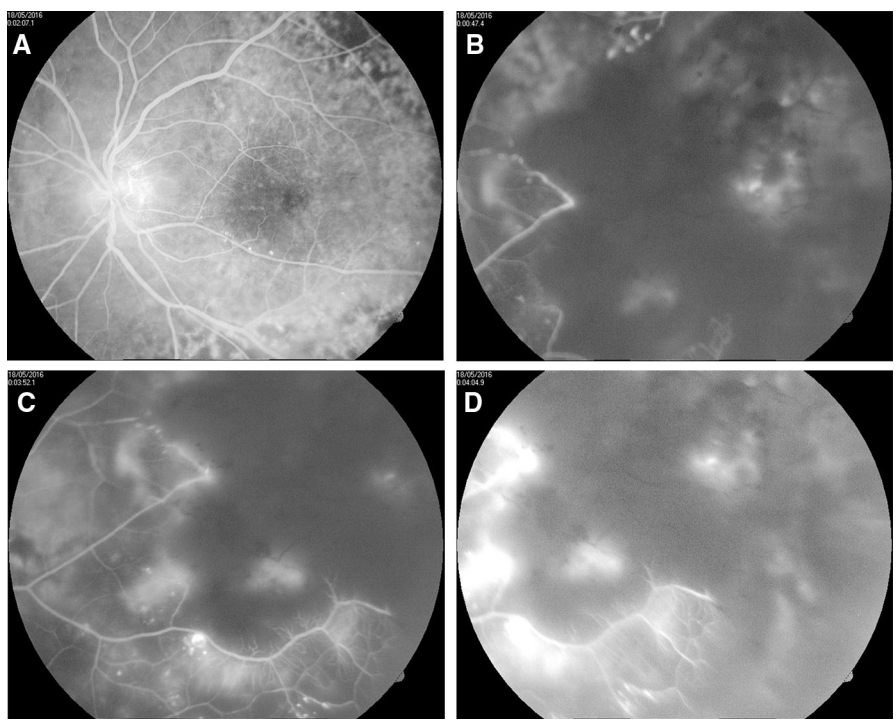


**Fig. 3** Funds fluorescein angiography images of left eye at presentation. **a** Posterior pole showing pinpoint leakages. **b**, **c**, **d** Early and late films showing capillary non-perfusion areas with leakages from telangiectatic vessels and telangiectatic bulbs

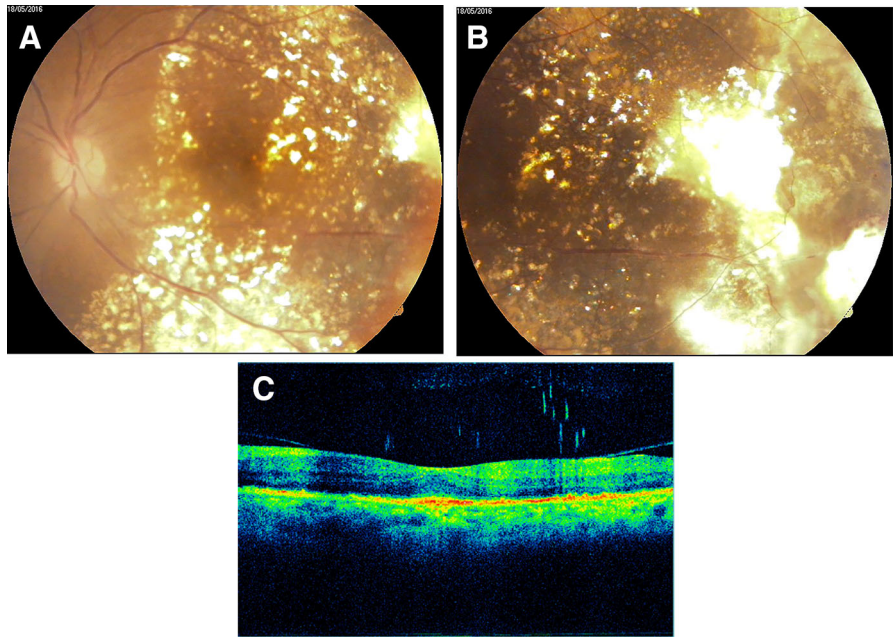


**Fig. 4** Fundus photographs of left eye 3 months post-intravitreal ozurdex injection. **a** Resolution of retinal exudation with clearing of intraretinal lipids. **b** Fibrosed retinal vasoproliferative tumor with regression. **c** SD-OCT 3 months post-injection, showing completely flat macula with resolved macular edema

Shields [10] have recently proposed a staging classification, which might help in selecting treatment and predicting the ocular outcomes of the disease (Table 1).

It is evident that in advanced stages of Coat's disease (Stage 3A and 3B) when there is massive retinal exudation or serous retinal detachment, laser treatment or cryotherapy is often ineffective and therefore with limited treatment options before. More recently, there has been development in the treatment of Coats' disease with the use of anti-VEGF (vascular endothelial growth factor) agents. Several reports on anti-VEGF (bevacizumab and ranibizumab) agents in stage 3 and 4 Coats' disease have shown promising efficacy in decreasing the amount of SRF, macular edema and exudates [6, 13, 14]. However, the patient

**Table 1** Coats' disease: classification and management of Shields and Shields [10]

| | | |
|---|---|---|
| Stage 1 | Retinal telangiectasia only | Periodic observation or laser photocoagulation<br>The visual prognosis is usually favorable |
| Stage 2 | Telangiectasia and exudation | Laser photocoagulation or cryotherapy, depending on the extent of disease and the preference of the ophthalmologist |
| | A: extrafoveal exudation<br>B: foveal exudation | The prognosis is secondary to the extension of exudation (favorable if limited to one quadrant or located nasally)<br>In stage 2A the visual prognosis is generally good, because the fovea is not involved in exudation<br>In stage 2B the visual prognosis is relatively good if the foveal exudation is not advanced |
| Stage 3 | Exudative retinal detachment | 3A: photocoagulation or cryotherapy; even if the retinal detachment involves the fovea, it will resolve when the telangiectasia is eradicated |
| | A: subtotal detachment<br>B: total retinal detachment | 3B: cryotherapy if the retinal detachment is shallow, but may require an attempt at surgical reattachment if the detachment is advanced and immediately posterior to the lens |
| Stage 4 | Total retinal detachment and glaucoma | Often enucleation for the severe ocular pain |
| Stage 5 | Advanced end-stage disease | Patients generally have a blind but comfortable eye and require no aggressive treatment |

may have to undergo multiple injections to achieve the desired effect to suppress the disease progression. Studies have shown aqueous and vitreous levels of VEGF that are approximately 1000 pg/ml in patients with Coats' disease; in contrast, patients with choroidal neovascular membranes secondary to age-related degeneration have aqueous levels of VEGF that are approximately 67 pg/ml [15, 16] and patients with branch vein occlusions have aqueous levels of VEGF that are less than 500 pg/ml [17]. Hence, there is a need for repeated treatment to control the exudation in Coats' disease. Another drawback with use of anti-VEGF agents is the occurrence of vitreo-retinal fibrosis and tractional retinal detachment as found in some studies [14, 18].

Latest in the management of Coats' disease is the use of intravitreal dexamethasone implant. Coats' disease may have a possible underlying inflammatory component, though uncommon [19]. Intravitreal corticosteroid treatment is known to attenuate leukostasis and vascular leakage along with suppression of inflammation [20], therefore dexamethasone implant can play a role in this complex disorder. Intravitreal triamcinolone acetonide injection is another alternative but with a poor safety profile compared to dexamethasone implant. Ray et al. [21] compared their results with bevacizumab injections to those achieved with triamcinolone acetonide injections as an additional treatment modality to ablative therapy in eyes with Coats' disease. They argued that rapid resorption of subretinal fluid was seen after treatment with intravitreal triamcinolone, but this was not the case after bevacizumab injection. Castillo et al. [8] demonstrated complete resolution of exudation with dexamethasone implant and closure of telangiectasias with photocoagulation done subsequently. Similar results were seen in case study of Saatci et al. [9] but in pediatric patients. This case report also illustrates the beneficial effect of steroid implant in Stage 3A Coats' disease, where there was complete resolution of exudative detachment following injection which in turn rendered the retina amenable to laser photocoagulation.

We suggest the use of dexamethasone steroid implant as the primary treatment for cases with massive exudation; when the SRF is absorbed, laser photocoagulation must be added to obliterate the abnormal vasculature and hyperpermeable aneurysmal dilatations. Combined therapy of intravitreal steroid and laser ablation can have sustained and long-term effect in keeping the disease under control.

## Conclusion

Coats' disease can be a rare presentation in an adult female patient. Secondary retinal vasoproliferative tumors are frequent associated finding in such cases. Intravitreal dexamethasone implant (Ozurdex) adds to the armamentarium of Coats' disease management. Studies with a larger sample size and longer follow-up duration are needed to establish the efficacy of dexamethasone steroid implant in advanced stages of Coats' disease.

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