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Coats' disease in adulthood with preserved vision after intravitreal aflibercept injection combined with laser photocoagulation : a case report

Safaa Abatli[1*†] and Sameeha Ziad Shweiki[2*†]

Abstract

Background This case report describes a rare case of Coats disease in adult female patient with preserved vision after intravitreal Aflibercept injection and laser photocoagulation.

Case presentation A female patient of Asian Palestinian descent, aged 20, exhibited a progressive and painless deterioration in the vision of her left eye over a period of two weeks. She exhibited no additional ocular symptoms. Prior to her presentation, she had no notable medical history and her vision was normal in both eyes. Inferotemporal telangiectasia, sausage-like blood vessels with perivascular sheathing in the peripheral fundus, extensive exudate involving the macula, severe macular edema, and localized inferotemporal exudative retinal detachment were observed upon examination of the posterior segment of her left eye. Following this, optical coherence tomography (OCT) identified subretinal exudate, intraretinal and subretinal fluid. After establishing the diagnosis of stage 3 Coats' disease, the patient was treated with intravitreal Aflibercept (Eylea) injections and sectoral laser photocagulation. The third injection resulted in the absence of intraretinal and subretinal fluid by OCT, but the subretinal exudate remained unresolved. One month subsequent to the previous injection, FFA guided sectoral laser photoagulation was applied to the inferiotemporal ischemic area. The patient was subsequently monitored monthly, and her vision improved. Five months after treatment, her vision has improved to 0.7 (6/8.7) and she has remained stable ever since. At present, the patient is undergoing routine outpatient follow-up.

Conclusion Coats disease is an idiopathic, progressive disease that mostly affects male infants, yet adult cases have been documented. Our case and the existing body of literature indicate that adult individuals have a favorable visual prognosis in the small proportion of cases where this occurs. It appeared that the implementation of intravitreal

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therapies and increased use of lasers led to enhanced visual outcomes. It is recommended to perform lifelong follow-up to monitor for recurrences and complications.

Keywords Adult onset Coats' disease, Retinal telangectasia, Exudative maculopathy, Aflibercept injection

Introduction

Coats' disease is defined as a non-hereditary idiopathic peripheral retinal telangiectasia (IPT) characterized by an abnormal development of telangiectatic and aneurysmal retinal vessels with progressive deposition of intraretinal and/or subretinal exudate, leading to exudative retinal detachment without appreciable retinal or vitreal traction. The most frequent complaint is decreased vision, but it can also unexpectedly manifest as discomfort, vitreous hemorrhage, or abnormalities of the anterior segment [1–3]. Although it usually affects children, Coats' disease can also strike adults. The presenting complaints and prognosis vary when Coats' disease manifests in adulthood [2, 4]. The gold standard for diagnosing Coats' disease is an indirect ophthalmoscopy fundus examination. Fundus Fluorescein Angiography (FFA) and Optical Coherence Tomography (OCT) can help confirm the diagnosis and rule out other diagnoses [5]. The course of treatment for Coats' disease is primarily determined by how severe the condition is, to always preserve the anatomy of the eyes and vision [1, 6]. Herein, we present a case of a 20-year-old Asian Palestinian lady who presented to our clinic with a painless decrease in her vision. The diagnosis of Coats' disease was then established, and the patient received the needed treatment for her case.

Case presentation

A 20-year-old Asian Palestinian female patient presented with a gradual painless diminution of vision in her left eye for 2 weeks. She had no strabismus, xanthocoria, or other ocular symptoms. She has no significant medical history, and her vision was normal in both eyes before her presentation, i.e., 1.0 (6/6). Ocular examination on presentation revealed a vision of 0.05 (6/120) in her left eye with no improvement. The vision in her right eye was normal, i.e., 1.0 (6/6). The intraocular pressure (IOP) of the left and right eye, measured using Goldmann applanation tonometry (GAT), was 16 mm of Hg in the right and left eyes. Slit lamp examination revealed no signs of inflammation or cataract in the anterior segment and no rubeosis. Posterior segment examination of her left eye revealed inferiotemporal telangiectasia with large aneurysms and sausage-like blood vessels with perivascular sheathing in the peripheral fundus, extensive exudate involving 12 clock hours and macula, severe macular edema, and localized inferotemporal exudative retinal detachment that extends to less than 4 clock hours with no vitreous hemorrhage. Fundus Fluorescein Angiography (FFA) revealed an area of hyperfluorescence corresponding to telangiectatic vessels, with peripheral hypofluoresence indicating capillary dropout and non-perfusion in that area and perivascular dye leakage in the arteriovenous phase. Optical Coherence Tomography (OCT) done subsequently revealed subretinal exudate with intraretinal and subretinal fluid and a central subfield thickness (CST) of 620 µm. The right eye examination, including fundoscopy, FFA, and OCT, was all within normal limits (WNL).

The diagnosis of stage 3 Coats' disease was established, and the patient was managed with intravitreal Aflibercept (Eylea) injections followed by sectoral laser photocoagulation. The patient received Aflibercept 2 mg/0.05 ml injections on day one, at 4 weeks, and at 8 weeks post-presentation. She was followed up every 2 weeks after each intravitreal injection, and improvement was assessed based on visual acuity and OCT findings. Two weeks after the third injection, her visual acuity improved from 0.05 (6/120) to 0.4 (6/15). OCT showed resolution of intraretinal and subretinal fluid, with the presence of subretinal exudate. The central subfield thickness (CST) improved from 620 µm to 178 µm after the third injection.

One month after the third injection, FFA-guided sectoral laser photocoagulation was performed on the inferotemporal ischemic area. The patient was then followed up monthly. Her vision improved to 0.4 (6/15) three months after the treatment. She has been stable since then, with her vision improving further to 0.7 (6/8.7) at 5 months post-treatment. Currently, the patient is being regularly followed up on an outpatient basis.

Discussion

Coats' disease is an idiopathic retinal vasculopathy that is non-hereditary in its nature. It is characterized by telangiectasia and aneurysm of retinal vessels, intraretinal and subretinal exudation, and exudative retinal detachment [1–3]. Coats disease is more common in Caucasian [7]. However, there are few studies of Coats' disease based mainly in Asian populations. One of those studies was done on Korean patients, 45% of those having childhood-onset disease, mean age at presentation was 8.6 years (range 0.2–17.8) and 55% of those having adult-onset disease, mean age at presentation was 39.8 years (range 18.3–69.0) [8].

Coats' disease commonly presents unilaterally with strong male predominance [5]. Herein, we present a case of a 20-year-old female, which is unusual because male predominance is a regular feature of the disease in the

literature [5, 9–11]. However, a recent extensive study of 351 eyes of 351 instances of Coats' disease from a single hospital revealed that 56 cases (16%) were female [7]. Therefore, despite the fact that it is uncommon to identify coats' disease in females, it still can be detected.

Our patient presented with a gradual, painless decrease in her left-eye vision. The IOP was normal. The slit lamp examination was also normal, with no evidence of any rubeosis. In the literature, decreased vision is the most common complaint [12, 13]. Unusual symptoms such as discomfort, vitreous hemorrhage, anterior segment abnormalities, and elevated IOP can also present [13]. However, in adults-onset coats' disease, a subset of patients remain asymptomatic for an extended period of time and are diagnosed on routine fundus examinations [2, 4].

Indirect ophthalmoscopy fundus examination is the gold standard for diagnosing Coats' disease [5]. Every case reveals retinal telangiectasia, which typically has a fusiform shape. The most frequently affected quadrants are the inferior and temporal ones [5, 14]. Nearly all cases have intraretinal exudation, which can be widespread and extend far from the telangiectasia [5]. Additionally, the density of macular exudate serves as a significant indicator of visual prognosis besides poor vision at presentation, even with appropriate treatment [1, 7]. In our case, the patient had inferotemporal telangiectasia that is sausage-like in shape with large aneurysms, extensive exudate in all 12 clock hours involving macula, extensive macular edema, and exudation, which align with the typical presentation with poor prognostic factors. However, the patient's results after the treatment were good, and the patient's vision improved.

Shields classified Coats disease into five stages: stage 1, which only shows retinal telangiectasia; stage 2, which shows both telangiectasia and exudates; stage 3, which shows the development of exudative retinal detachment; stage 4, which shows total retinal detachment and glaucoma; and stage 5, where the disease has progressed to the end stage and is occasionally accompanied by phthisis bulbi [1, 7]. We classified our patient as stage 3 due to the observation of retinal detachment. However, this is not concordant with the previous studies that report that nonjuvenile cases of Coats' disease are usually diagnosed at stage 2 [7].

The treatment modality for coats' disease depends mainly on the severity of the disease, and it always aims to preserve vision and ocular anatomy [1, 6]. Cryotherapy, laser photocoagulation, scleral buckling, external subretinal fluid drainage, and pars plana vitrectomy (PPV) are among the treatment options. We have also utilized adjunctive intravitreal injections of corticosteroids and anti-VEGF in patients with subretinal exudates and macular edema. Ocular VEGF levels have been shown to be elevated in patients with Coats' disease. This finding has prompted the use of anti-VEGF therapy as both primary and adjuvant treatment [7, 15]. Laser photocoagulation and cryotherapy are the first-line treatments for Coats' disease, particularly for stage 1. However, stages 2 to 3 A do not respond well to these treatments when used independently because retinal exudative changes and serous retinal detachment obstruct the effectiveness of laser therapy. To drain subretinal fluid before photocoagulation, surgical intervention might be necessary, or alternatively, anti-VEGF injections as adjuvant therapy might prove effective [16].

Our patient had extensive retinal exudation with massive subfoveal fluid and exudate, so the decision was made to initiate treatment with intravitreal Aflibercept injections. A reduction in subretinal fluid, a decrease in the size of telangiectatic vessels, improved retinal detachment, and improved visual acuity were observed. Kang et al. [8] reported that the use of more frequent anti-VEGF injections during the initial treatment period was associated with an improvement in final visual acuity by approximately 9.4 letters.

While multiple recent reports have demonstrated resolution of severe retinal detachments following anti-VEGF therapy [17], a study by Ramasubramanian et al. [18] cautioned against the use of anti-VEGF agents due to the development of vitreoretinal fibrosis and tractional detachments in eight pediatric patients. Another study by Daruich et al. [19] showed that 40.6% of 69 patients exhibited findings associated with extramacular fibrosis, which were not linked to cryotherapy, laser therapy, or anti-VEGF treatment.

Recurrences can also occur several years later, characterized by the return of exudates after the disease appears to have successfully remitted. A side consequence of Coats disease could be cataract development [20]. Therefore, even once the disease has stabilized, we advise close patient monitoring due to potential consequences and recurrences. Patients typically schedule lifetime follow-up appointments, ideally once every six months [21].

Conclusion

Coats disease is a progressive, idiopathic disease that affects male infants predominantly, although it can also manifest in adults and adolescents. Our case demonstrates that the visual prognosis is good in the few instances in which adult people are affected. The introduction of intravitreal therapies and more frequent use of lasers seemed to improve visual outcomes. We recommend conducting lifelong follow-up to monitor for complications and recurrences, longer follow will be reported in future cases.

Appendix

See Figs. 1, 2, 3, 4, 5, and 6.

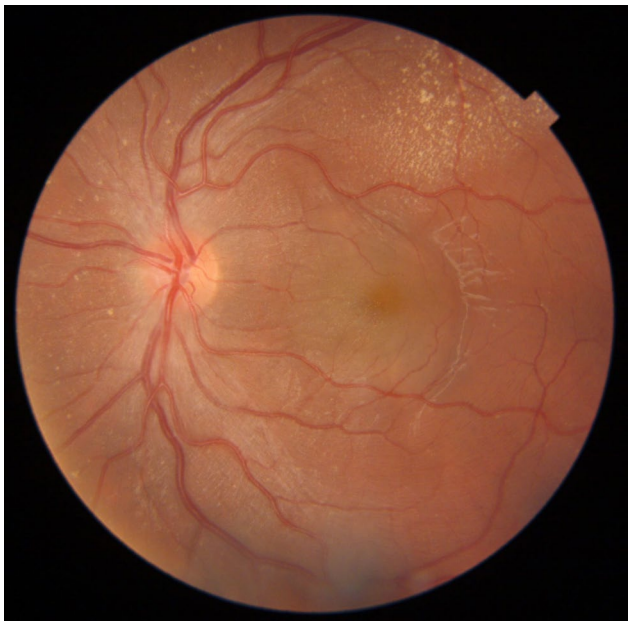


Fig. 1 Colored fundus photo for left eye before treatment, exudate involving all quadrant with macular edema

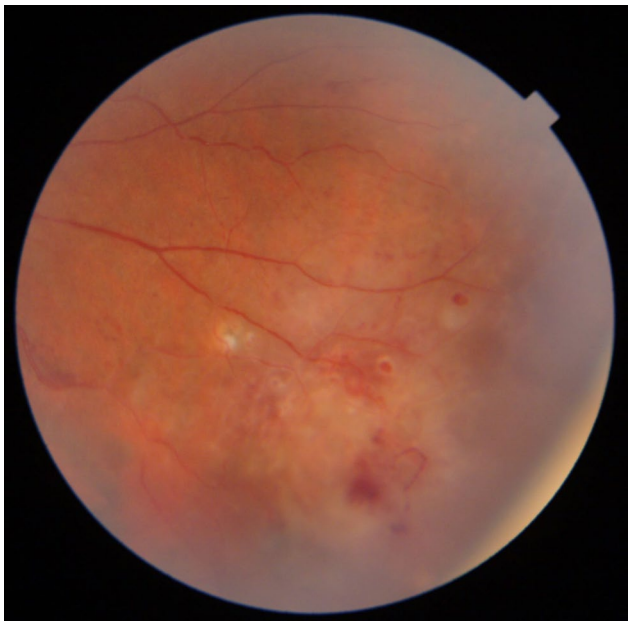
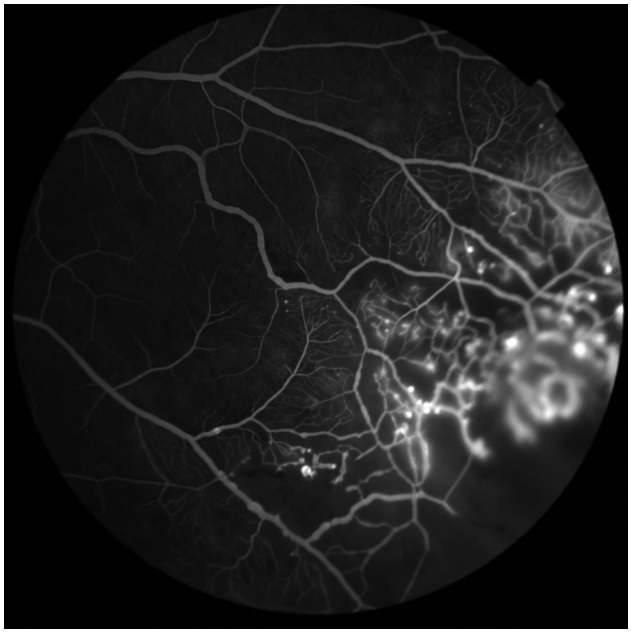


Fig. 2 Colored fundus photo for left eye before treatment, Inferiotemporal telangiectasia with aneurysmal dilated blood vessels in peripheral fundus, surrounded by localized exudative retinal detachment

Fig. 3 Fluorescein angiography (FFA) arteriovenous phase before treatment: capillary drop out and non-perfusion, perivascular dye leakage, and arterial beading



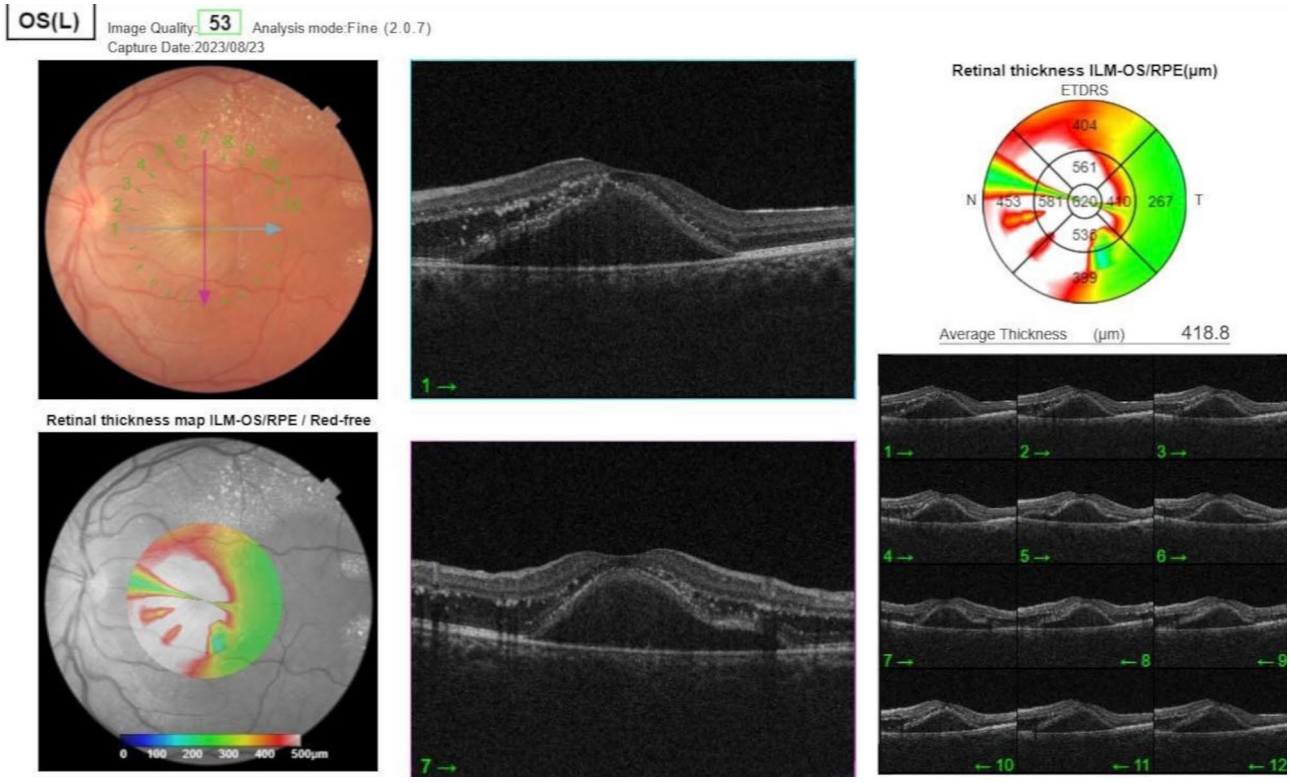


Fig. 4 SD OCT of left eye before treatment: intraretinal fluid and exudate, and sub-retinal fluid, CST:620

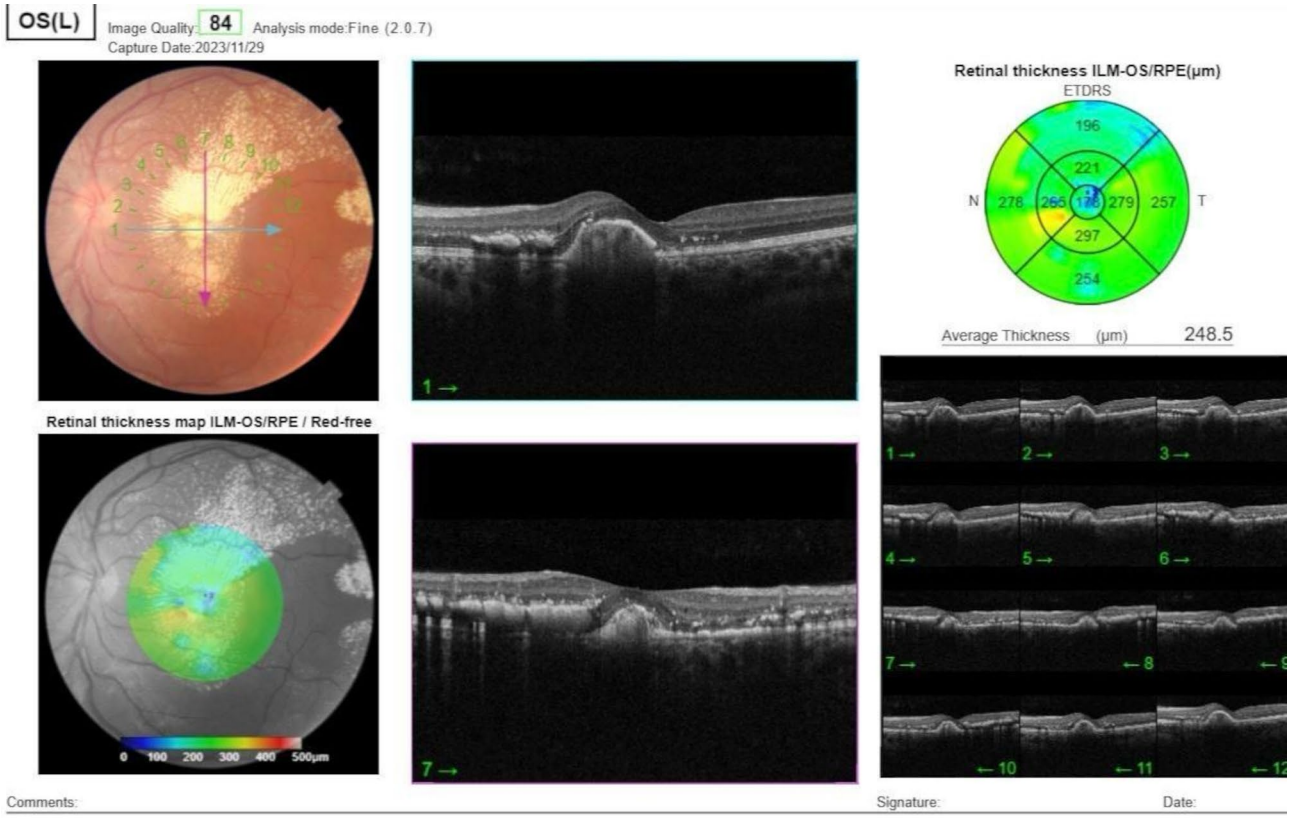


Fig. 5 SD OCT of left eye 2 weeks after the 3rd intravitreal aflibercept injection, Resolved intraretinal (IRF) and subretinal (SRF) fluid and persistent of subfoveal exudate, CST:178

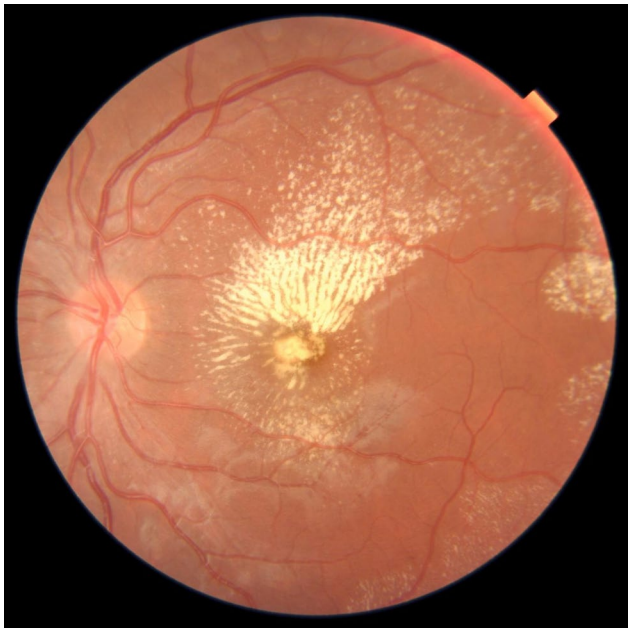


Fig. 6 Colored fundus photo after treatment: Extensive exudate involving macula and resolved macular edema

Abbreviations

OCT	Optical Coherence Tomography
IPT	Idiopathic Peripheral Telangiectasia
FFA	Fundus Fluorescein Angiography
IOP	Intraocular Pressure
GAT	Goldmann Applanation Tonometry
CST	Central Subfield Thickness
WNL	Within Normal Limits
IRF	Intraretinal Fluid
SRF	Subretinal Fluid
PPV	Pars Plana Vitrectomy
VEGF	Vascular Endothelial Growth Factor

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
Ethical approval was not sought for the present study because it is not reporting research findings.

Consent for publication
Written informed consents for publication were obtained from the patient.

Competing interests
The authors declare no competing interests.

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