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Adult-onset Coats' disease. Case series and literature review



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HIGHLIGHTS

Early diagnosis and initial visual acuity are key predictors of the treatment outcome in adult-onset Coats' disease; combined therapy effectively stabilizes the condition.

ABSTRACT

Adult-onset Coats' disease is a rare idiopathic retinal vascular disorder. We present three cases of unilateral retinal telangiectasia and exudation diagnosed through clinical examination and fluorescein angiography. All patients were treated with laser photocoagulation and/or: two also underwent cryotherapy, one received transpupillary thermotherapy, and two received adjunctive anti-VEGF therapy. A literature review highlights male predominance, variable age of onset, and frequent macular involvement. Initial vision acuity appears to be the primary predictor of post-treatment outcomes. Early diagnosis and combined therapy can stabilise the disease and preserve vision.

Key words: Coats' disease, adult-onset, retinal telangiectasia, retinal vasculopathy, multimodal treatment

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INTRODUCTION

Coats' disease was first described in 1908 by the Scottish ophthalmologist George Coats. This idiopathic disorder is characterised by retinal telangiectasia and focal aneurysms accompanied by intraretinal and subretinal exudates as well as exudative retinal detachment [1, 2]. The aetiology of the disease remains elusive, with suspected microvascular anomalies leading to the breakdown of the blood–retinal barrier and subsequent plasma leakage, resulting in exudate formation [3].

The disease typically occurs in young boys; however, adult-onset cases have also been reported, primarily in men (75%) over the age of 35 [4, 5]. The condition is unilateral, with the most common presenting symptom being visual disturbance (83%), followed by strabismus (8%), floaters (6,2%) and scotoma (2%) [5]. Coats' disease is often associated with systemic conditions such as hypertension, diabetes mellitus and hypercholesterolemia, which are considered risk factors [2, 5].

The adult-onset form of the disease generally presents in a milder form compared to its paediatric counterpart, with fever, exudative retinal detachments and a longer asymptomatic period. This variant of the disease is also named as Leber miliary aneurysm [2, 6]. Both forms are collectively termed 'idiopathic retinal telangiectasia'. Based on disease severity, Coats' disease can be classified into 4 categories, with the most severe form characterised by peripheral exudation, hard macular exudates, and macular edema [6]. Treatment depends on the severity of the disease and includes laser photocoagulation, anti-vascular endothelial growth factor (VEGF) injections, intravitreal triamcinolone injections, cryotherapy, vitrectomy or enucleation [2, 3].

In this short case series, we describe three patients with adult-onset Coats' disease. Two of them were successfully treated with satisfactory visual outcome with the cryo and laser photocoagulation, in one of them vision didn't improve due to subfoveal scarring.

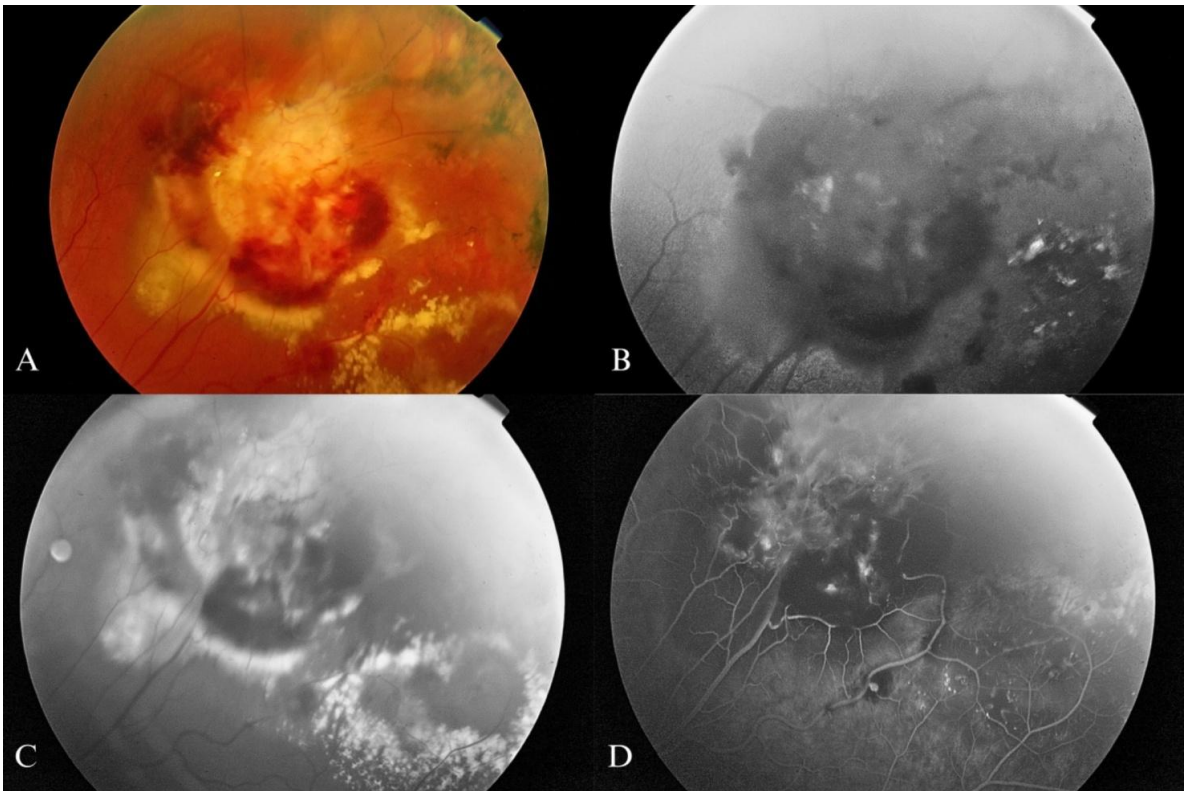
CASE DESCRIPTIONS

Case 1

A 49-year-old patient was referred to our clinic in 2017 due to dark spots affecting his visual field. The patient had no chronic diseases, significant personal or family medical history. His visual acuity was 0.0 logMAR in both eyes, intraocular pressure was within the normal range. Fundoscopic examination revealed hemangiomas, aneurysms with scar tissue and exudates in the posterior pole of the left eye (fig. 1).

FIGURE 1

Patient 1: colour (A) and autofluorescence (B) fundus photography at presentation (May 2017). Early (C) and late (D) arteriovenous phase of fluorescein angiography after the first laser treatment.



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Based on the clinical manifestation and examination, adult-onset Coats' disease was diagnosed. A series of 6 laser photocoagulation sessions and cryotherapy was initiated, after which the patient reported resolution of his visual disturbances.

At the 3-month follow-up visit, exudation around the lesion was noted, and bevacizumab treatment was proposed. The patient underwent 3 intravitreal injections. Following treatment, scar tissue formed over the lesion. Due to macular exudates, adjacent anti-VEGF therapy was initiated. By October 2018, the lesion had stabilised, and the best corrected visual acuity (BCVA) remained at 0.0 logMAR.

In May 2021, the patient was diagnosed with colorectal cancer and underwent chemotherapy, followed by colectomy with ileostomy. No ocular symptoms recurred during this period. However, in June 2024, he reported metamorphopsias. New exudation was observed in the upper temporal arcade requiring another session of argon laser photocoagulation (fig. 2).

Due to the metastatic lesion in the lungs, the patient was started on the FOLFIRI chemotherapy scheme with panitumumab together with radiotherapy. On last follow-up visit, in April 2025, the vision was 0.0 logMAR in both eyes and adjacent laser treatment was applied.

Case 2

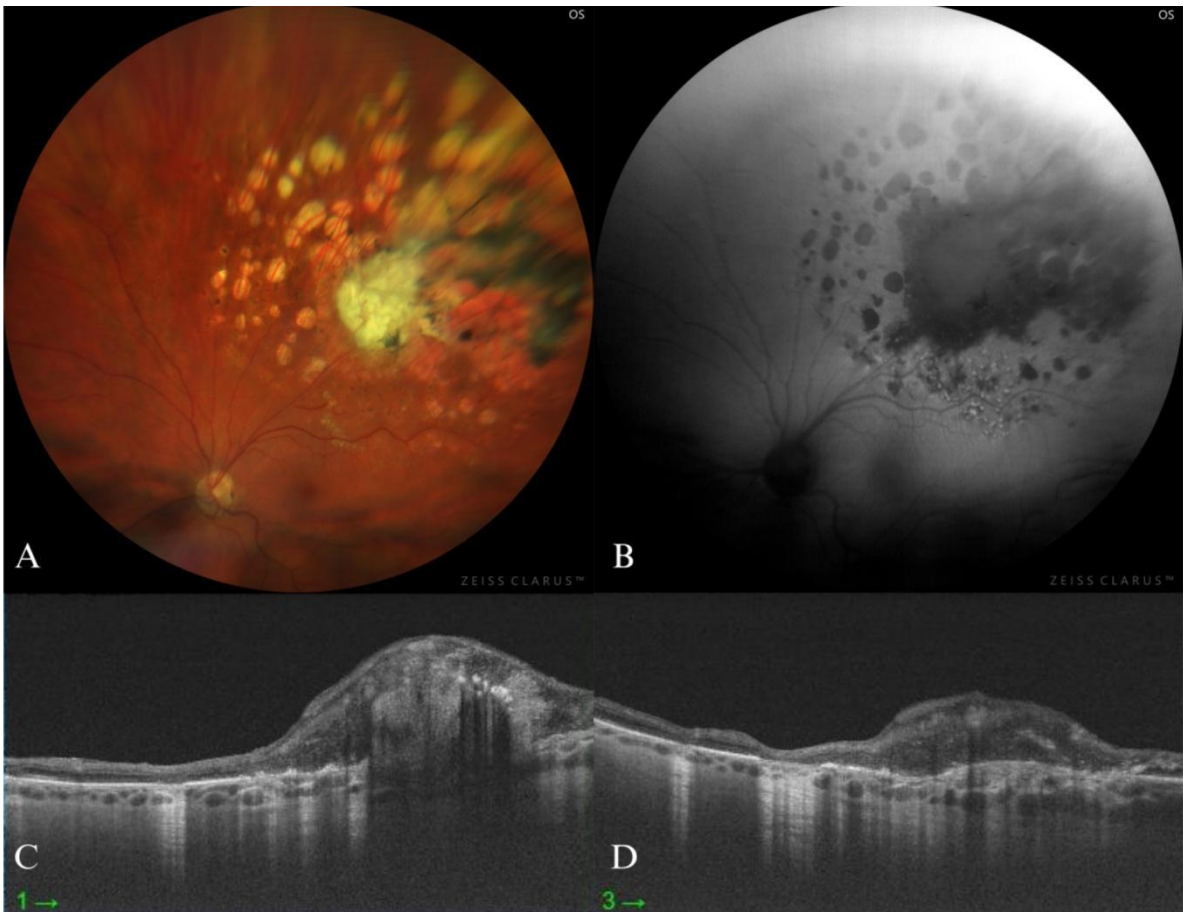
A 31-year-old man was referred to our clinic in October 2024 with a suspicion of a retinal vasoproliferative tumour. He was asymptomatic, and the lesion was detected during a routine ophthalmic examination. His medical history was unremarkable for prior ocular procedures or ongoing ophthalmic treatment. The patient had a history of hypothyroidism.

On the initial examination, his visual acuity was 0.0 logMAR in both eyes, and intraocular pressure was within normal limits. Anterior segment examination was unremarkable. Fundoscopic evaluation revealed a large area of exudation, with aneurysms and retinal pigment epithelium proliferation over the upper vascular arcade of the left eye (fig. 3).

Based on the clinical features and fluorescein angiography (fig. 4 A, B) the diagnosis of the adult-onset Coats' disease was established. The patient was qualified for the

FIGURE 2

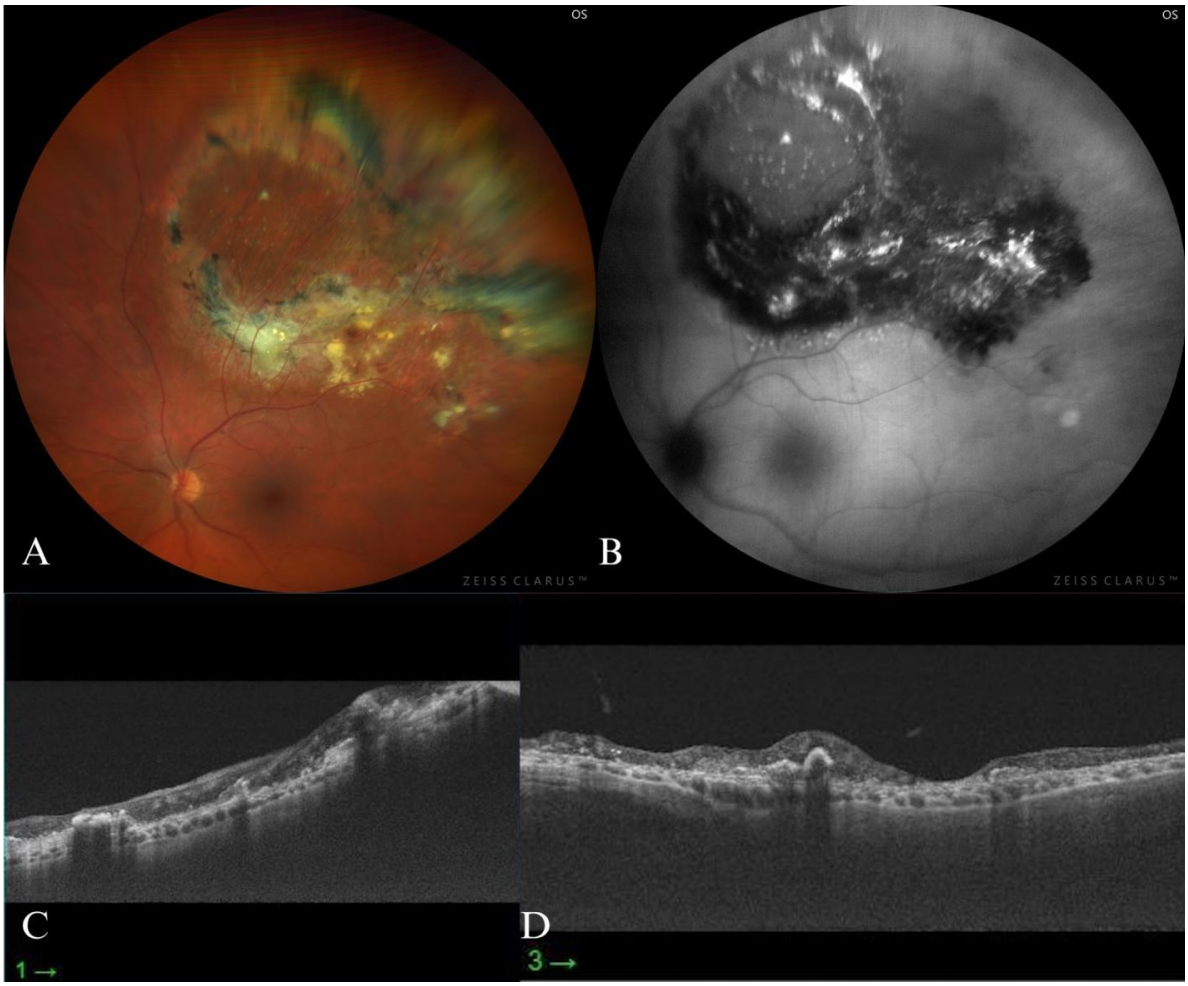
Patient 1: colour (A) and autofluorescence (B) fundus photography after the treatment (November 2024). OCT scans through the lesions showing sub- and intraretinal fluid (C, D).



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FIGURE 3

Patient 2: colour (A) and autofluorescence (B) fundus photography before the treatment (October 2024). OCT made through the lesions presenting intraretinal exudates (C, D).



photocoagulation treatment. Two months after the procedure, his visual acuity was 0.6 logMAR in the right eye and 0.5 logMAR in the left eye. The patient reported no new symptoms. Comparative fundus photography showed stable retinal exudation without progression.

Case 3

A 25-year-old man was referred to our clinic in October 2017 with a suspicion of an intraocular vascular tumour in the left eye. He noticed a decrease in visual acuity since August 2017. His medical history was unremarkable for prior ocular procedures or ongoing ophthalmic treatment.

On the initial examination, his visual acuity was 0.0 logMAR and 0,4 logMAR in the right and left eye, respectively. The intraocular pressure was within normal limits. Anterior segment examination was unremarkable. Fundoscopic evaluation of the left eye revealed the macular edema and a large area of exudation in the upper quadrant (fig. 5 A, B). The fluorescence angiography confirmed the diagnosis of adult-onset Coats' disease, showing non-perfusion and hyperfluorescence areas in the upper quadrant of the left eye (fig. 5 C, D). Initial laser and transpupillary thermotherapy (TTT) treatments were unsuccessful and adjacent anti-VEGF injections were initiated. Despite initial reduction of exudates, lesions recurred and cryotherapy was applied, followed by another two anti-VEGF injections.

In total, 4 laser photocoagulation treatments, 5 TTT sessions, 6 anti-VEGF injections, and one cryotherapy were applied in the span of 3 years, and lesions' stabilisation was achieved (fig. 6). However, the final vision acuity in the left eye remained impaired due to subfoveal scarring, measuring 0.3 logMAR on the last control visit in October 2020.

DISCUSSION

This is a short case series presenting three patients with adult-onset Coats' disease. The diagnosis was made on the basis of clinical presentation and fluorescein angiography in each

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FIGURE 4

Patient 2: early (A) and late (B) arteriovenous phase of fluorescein angiography revealing hyperfluorescence due to the leakage and telangiectasis (October 2024).

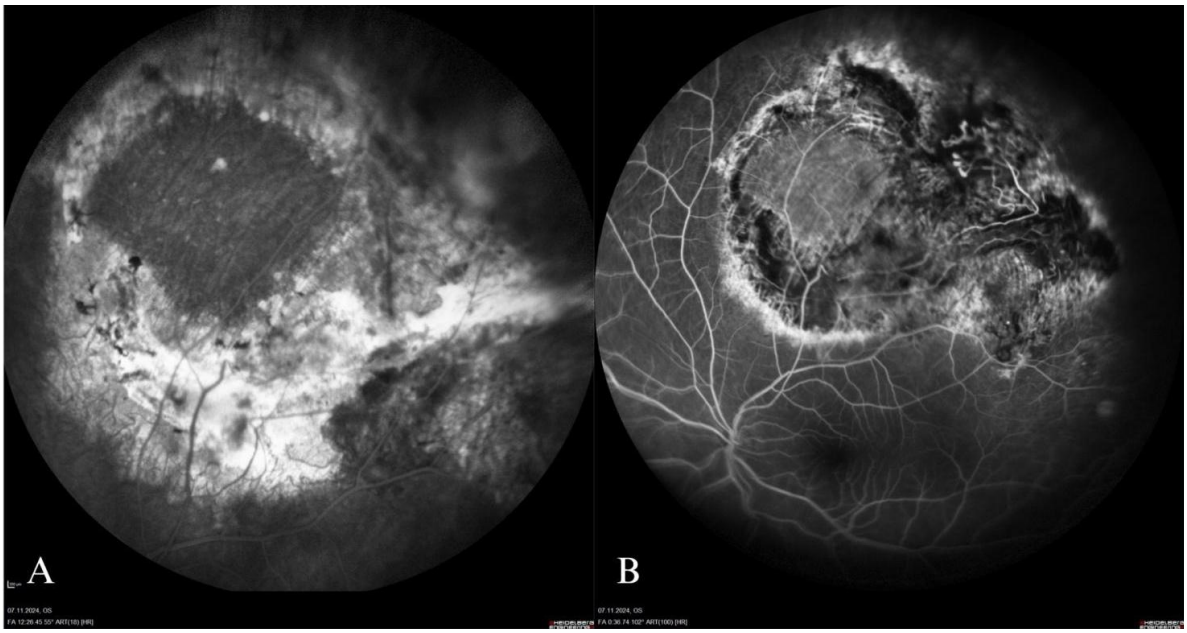
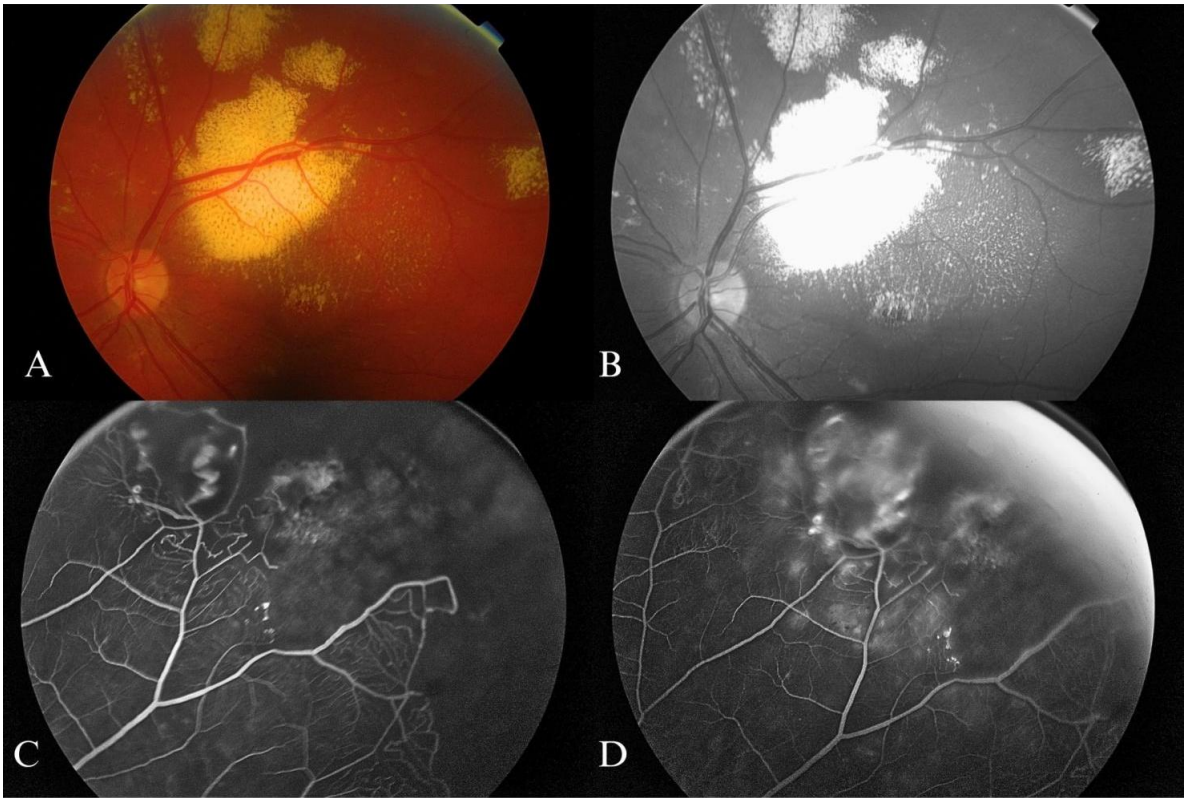


FIGURE 5

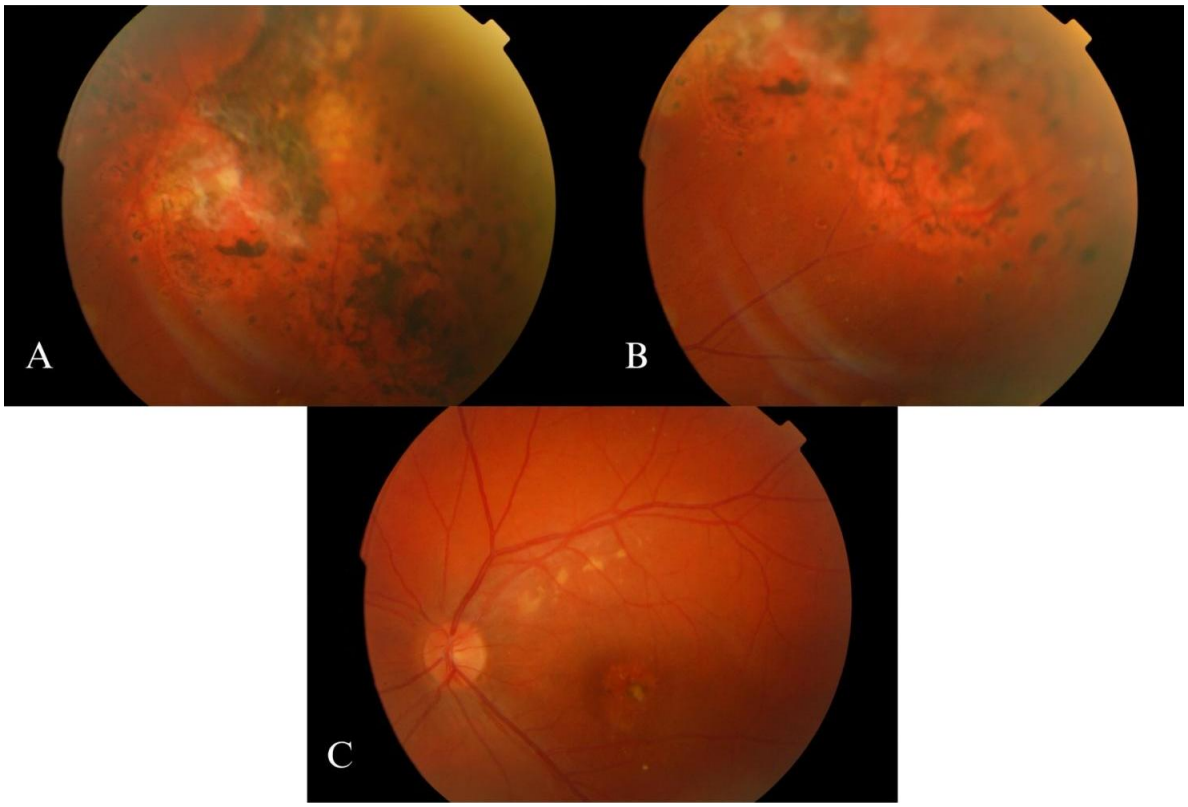
Patient 3: fundus colour (A) and autofluorescence (B) photography of the massive exudates in the upper quadrant of the left eye. Areas of non-perfusion on the fluorescence angiography scans (C, D).



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FIGURE 6

Patient 3: smaller exudates in the upper quadrant of the left eye, fibrotic scar after cryotherapy (A, B). Pigment epithelial atrophy area around the macula (C).



case. The differential diagnosis for these cases would be dominant (familial) exudative vitreoretinopathy, facioscapulohumeral muscular dystrophy, retinopathy of prematurity (ROP) and retinal hemangioblastomas [7].

A search of the PubMed database was conducted using the terms „Coats disease" AND „adult onset". A total of 41 articles were identified, including 3 review papers, 19 single-patient case reports, 16 case series involving at least 2 patients, and 3 commentaries on previously published papers. From the 16 case series, only those with extractable data specific to adult patients were included for literature review and detailed analysis. Variables such as age of onset, sex, laterality, stage at the presentation, visual acuity before and after treatment, and treatment modalities were evaluated and summarised in table 1.

The reported mean age of disease onset ranged from 30.5 years up to 73.5 years. The youngest adult patient was 21 years old [12], while the oldest one 76 [13]. Our patients were diagnosed at 25, 31 and 49 years of age, falling within the typical adult-onset range.

A comprehensive review of Coats disease in 351 eyes for 45 years by Shields et al. [14] reports a mean onset age of approximately 12 years without distinguishing between paediatric and adult populations, indicating a predominance of paediatric cases. Although historically described as a condition predominantly affecting young male [15], the literature shows that adult-onset disease occurs across a wide age range and continuous to demonstrate a strong male predominance – males account for ≥50% of all adult cases and up to 84% in combined pediatric and adult populations [14]. The disease is classically unilateral in both children and adults, with no significant laterality preference (right eye: 46% vs. left eye: 54%, p = 0.31) [14]. However, a recently published study involving 19 adult patients reported bilateral findings in 8 cases, with characteristics of telangiectasia and other defining features of Coats disease [10]. This uncommon presentation may be attributed to the use of ultra-wide field fluorescein angiography and possibly a genetic predisposition within that specific population.

Although the ethology remains unclear, modern molecular studies have provided insight into the underlying pathophysiology. Banerjee et al. [16] highlighted the role of VEGF in disease progression. Non-perfused retinal areas trigger VEGF production, leading to the development of weakened, permeable vessels, which in turn cause accumulation of exudation and subretinal fluid [16]. Somatic mutations have also been implicated, with studies reporting a higher prevalence of NDP gene mutations – particularly missense vari-

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TABLE 1

Characteristic of the adult-onset Coats' disease cases.

Author	Year	No. eyes	Mean age at disease onset; years (±SD)	Sex (%male)	Laterality (%) unilateral	Clinical manifestation	Initial stage (most prevalent)	Eye fundus examination	Vision prior to treatment (mean, logMAR)	Vision after treatment (mean, logMAR)	Treatment
Amoroso et al. [8]	2024	7	53 (±13.29)	100	100	-	2B	aneurysms, macular edema	0.77 (±0.27)	0.39 (±0.38)	Navilas navigated laser photocoagulation
Hansraj et al. [9]	2024	74	50	72.5	92.8	-	2A	hard exudates (94.6%), microaneurysms (81.1%), macroaneurysms	1.0	0.92	laser photocoagulation (43.4%); intravitreal agents + laser photocoagulation (37.7%); laser + cryotherapy (3.8%); cryotherapy (1.9%)
Jung et al. [10]	2024	19	39.5 (±15.8)	57.9	63.2	decreased vision (36.7%), routine examination (33.3%)	2A (63.2%)	telangiectasia, exudate, capillary dropout, macular edema	0.19 (±0.51)	0.44 (±0.83)	laser treatment (94.7%); prophylactic laser treatment and intravitreal anti-VEGF injection for the fellow eye
Zhou et al. [11]	2024	15	57.33 (±12.61)	66.67	100	-	2B (73.3%)	telangiectasia (86.7%), patchy hemorrhage (33%)	4.0 (±1.1) (BCVA)	4.6 (±0.5) (BCVA)	combined therapy: laser + injections (60%); laser treatment (20%); anti-VEGF injections: conbercept 0.5 mg (13%)
Dave et al. [12]	2022	3	55 (±7.12)	66.67	100	decreased vision	2B	macroaneurysm, lipid exudates, hemorrhage, edema	0.53 (±0.33)	0.9 (±0.25)	laser; sub-Tenons triamcinolone injections; intravitreal injections using bevacizumab, aflibercept; dexamethasone intravitreal implant
Gawęcki et al. [6]	2021	6	36.67 (±17)	83.3	100	decreased vision (66.67%), asymptomatic	3	hard exudates, macular cystic edema	0.49 (±0.53)	0.27 (±0.28)	laser treatment; anti-VEGF injections; observation
Kang et al. [13]	2021	39	39.8 (±16.1)	73	95	decreased vision (41%), floaters (24%), blurred vision (16%)	2A, 2B	temporal exudates (62%), retinal hemorrhage (67%), macular edema (46%)	0.80 (±0.73)	0.85 (±0.75)	combined therapy 36%; laser treatment 28%; anti-VEGF injections 13%
Xu et al. [14]	2021	2	30.5 (±7.5)	50	100	blurred vision, metamorphop-sias	2B	microaneurysms, lipid depositions, edema	0.65 (±0.35)	0.1 (±0.1)	intravitreal anti-VEGF injections; thermal laser; laser photocoagulation
Wang et al. [15]	2020	2	32.5 (±7.5)	100	100	decreased vision	2A	lipid exudates, telangiectasia, haemorrhages, cystic macular edema, epiretinal membrane	0.61 (±0.09)	0.26 (±0.04)	laser treatment; anti-VEGF injections

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Zhang et al. [16]	2018	12	44.08 (±10.61)	66.67	100	-	3A	subretinal hard exudate, cystoid macular edema, subretinal fluid, telangiectasia	1.27 (±0.69)	1.05 (±0.73)	anti-VEGF injections combined with laser treatment
Kumar et al. [17]	2016	3	35.33 (±8.5)	100	100	decreased vision, blurred vision, metamorphopsias	2B	telangiectasias, edema, exudation (temporal), macroaneurysm, epiretinal membrane,	0.65 (±0.35)	0.46 (±0.12)	laser treatment; anti-VEGF injections (bevacizumab); triamcynolon sub-Tenon injections
Rishi et al. [5]	2016	48	47.2 (±8.1)	73	93	decreased vision (83.3%), floaters (6.2%); routine examination (8%)	2B	telangiectasis (mainly peripheral), exudates, haemorrhages, retinal detachment	1.13 (±0.91)	improved – 34.7% stable – 47.8% Worsened – 17.3%	laser – 60.4%; observation –27.08%; surgery –6.2%; cryotherapy –4%; laser + cryotherapy –2%
Zheng et al. [18]	2014	5	33.6	80	100	-	3B	retinal detachment, telangiectasia, vitreoretinal fibrosis (40%)	1.42 (±0.71)	1.25 (±0.62)	bevacizumab injections –100%; laser photocoagulation – 60%; triamcynolon – 20%
Otani et al. [19]	2011	2	73.5 (±2.5)	100	100	decreased vision	3	subretinal exudates, telangiectasis, non-perfusion areas, microaneurysms	0.31 (±0.09)	0.0; 1.0	laser photocoagulation; cryotherapy
Wang et al. [20]	2011	3	21; 51; 68	100	100	blurred vision, metamorphop-sias	2B	telangiectasia, aneurysms, intraretinal haemorrhage, exudates, subretinal fluid	0.6 (±0.35)	0.2 (±0.23)	intravitreal bevacizumab, argon yellow laser photocoagulation
Goel et al. [21]	2011	3	42 (±2.06)	66.67	100	decreased vision	2B	telangiectasia, macula exudation, aneurysms, epiretinal membrane	1.3 (±0.15)	0.91 (±0.3)	intravitreal bevacizumab, laser photocoagulation
Smithen et al. [22]	2005	13	50.4 (±11.6)	92	100	decreased vision (46.1%), floaters (30.7%)	-	telangiectasia, exudates, haemorrhages (preretinal), retinal detachment	-	-	laser – 84.6%; observation – 15.3%

ants – in affected retinal tissue. Additionally, the telomerase RNA component gene (*TERC*) has been associated with disease in some cases, although further research is needed to validate the significance [17].

Diagnosis is primarily clinical, based on fundoscopic findings. Hallmark features – present in all reported cases – include retinal telangiectasia (often located in the temporal quadrant) and lipid-rich exudation [4, 5, 10, 16]. In adults, retinal hemorrhages are more prevalent, with reported incidence ranging from 22.9% [5] to 67% [18]. Other commonly observed findings include microaneurysms, cystoid macular edema, and subretinal fluid [9]. Compared to adults, chil-

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dren typically present with more extensive retinal involvement – affecting a greater number of quadrants and clock hours [10, 18]. Additional signs include peripheral nonperfusion and capillary dropout [10].

While telangiectasia and microaneurysms are generally found between the equator and the ora serrata, exudation and subretinal fluid tend to extend from the macula to the equator [14]. Retinal detachment, though more common in pediatric patients [5, 18], has been reported in 20–100% of adult cases [5, 19], possibly reflecting differences in genetic background of study design. Notably, adult patients are more likely to present with macular involvement, which may explain the early decline in visual acuity [16]. Initial visual acuity varies significantly, ranging from 0.19 logMAR [10] to finger counting [20], depending on disease severity and macular involvement at diagnosis.

Fluorescein angiography is the diagnostic standard for Coats disease [2]. While it remains the cornerstone of the diagnostic process, some authors suggest that the optimal approach is to perform ultra-wide field fluorescein angiography to better document lesions in the far periphery of the retina [10, 21, 22]. Ultra-wide field imaging has proven to be more precise in detecting retinal abnormalities, particularly in cases with an epiretinal membrane [22]. In selected cases, indocyanine green angiography (ICGA) may also be utilized, as it provides visualization of the choroidal vasculature and aids in ruling out differential diagnoses [23]. Additionally, for new photocoagulation techniques using computer-guided laser therapy, ICGA is one of the recommended tests for precise retinal mapping [8]. Another essential diagnostic tool is optical coherence tomography (OCT), which is now widely accessible and routinely used to monitor macular changes, including central macular thickness [11], edema [6, 18, 24], sub- and intraretinal fluid accumulation [9], and retinal detachment [13]. Although optical coherence tomography angiography (OCTA) cannot fully replace fluorescein angiography [16], it provides valuable insights into microvascular alterations, such as disturbances in the superficial capillary plexus, reduced blood flow density in the deep capillary plexus, and deformation of the foveal avascular zone [11, 21]. Interestingly, for research purposes, Wang et al. [24] expanded standard diagnostic methods by performing fine-needle aspiration to analyse intraocular levels of VEGF and inflammatory cytokines (IL-6, IL-8). While they observed increased levels of IL-6 and IL-8, VEGF levels remained within the normal range or were even reduced. The initial ratios of interleukins to VEGF may serve as potential biomarkers for predicting treatment responses to anti-VEGF or corticosteroid injections [24].

Laser therapy has been the most commonly employed treatment for Coats' disease in the pediatric population over the years, with an average of 3.6 treatment sessions per patient [15]. The primary objective of laser photocoagulation is to ablate telangiectatic vessels, promote exudate resorption, and stop disease progression [15]. Photocoagulation achieves exudate resolution by occluding the lumens of macroaneurysms, reducing fluid leakage, and directly ablating retinal cells that produce inflammatory markers [21]. Green or yellow laser wavelengths are typically used [16], with yellow laser being particularly selective for hemoglobin, thereby minimizing collateral damage to surrounding structures, including the retinal pigment epithelium [12]. Although laser photocoagulation is generally considered a safe treatment modality, potential complications, such as disruption of the blood–retina barrier, increased exudation, or vitreoretinal traction, should be taken into account [12, 25]. In cases where vitreoretinal traction develops, surgical intervention may become necessary [5]. To address the formation of exudates following laser therapy, a combined treatment approach with anti-VEGF injections has been proposed. Xu et al. [21] observed in their cohort that fibrovascular scar formation, a potential consequence of laser therapy, was mitigated by concurrent anti-VEGF treatment. A recent advancement in laser therapy is computer-navigated laser photocoagulation, which utilizes pre-treatment lesion mapping to enable precise laser delivery while preserving healthy tissue [8]. Amoroso et al. [8] reported a significant improvement in BCVA following this approach, with no documented adverse effects, including scar formation. Over recent years, improvements in final BCVA and patient satisfaction have been noted, which may be attributed to the transition from cryotherapy to laser treatment, thereby reducing the risk of proliferative vitreoretinopathy [18]. Additionally, selecting cryotherapy as a first-line treatment has been associated with a higher likelihood of requiring subsequent interventions [18].

Given the potential role of VEGF in the pathophysiology of Coats' disease, targeted anti-VEGF therapies have emerged as a treatment option aimed at preventing disease progression to irreversible blindness [3]. Several studies have reported positive outcomes following treatment with bevacizumab monotherapy [6, 9, 10, 19, 23, 24], combination therapy with laser photocoagulation [9, 11, 18–21], ranibizumab [9], conbercept [11], and aflibercept [21, 23, 24]. The number of bevacizumab injections administered has been shown to correlate significantly with final BCVA outcomes, particularly in patients whose initial visual acuity exceeded 20/66 [10], suggesting that baseline BCVA at diagnosis is the most important prognostic factor [18].

Corticosteroid therapy represents another potential treatment strategy. The mechanism of action involves stabilizing vascular integrity by repairing tight junctions between endothelial cells, thereby reducing exudation [16]. Corticosteroids can be administered via sub-Tenon injections or long-acting intravitreal implants. While some patients

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demonstrate a favourable response, the use of corticosteroids is limited due to associated side effects [23]. Indications for corticosteroid therapy include coexisting central macular edema [12, 22], macular lipid deposition [23], and combination therapy with other treatment modalities [19]. Surgical intervention, while frequently required in pediatric patients (25%) [5, 14], is rarely necessary in adults [16]. In cases of advanced disease with retinal detachment, vitrectomy with silicone oil or gas tamponade may be indicated [15]. Additionally, surgical intervention may be required for complications such as macular hole formation or epiretinal membrane [3]. Enucleation remains a last-resort option in cases of intractable complications, such as treatment-resistant intraocular pressure elevation or iris neovascularization leading to a painful blind eye [15].

CONCLUSIONS

Adult-onset Coats' disease is a rare variant of retinal idiopathic teleangiectasia in adults. The laser photocoagulation combined with anti-VEGF injections represents the most effective treatment option.

Figures: The figures included in the manuscript consist of photographs obtained from the database of the Ocular Oncology Department at Poznan University of Medical Sciences.

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