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Case Report

Triple Trouble with Triple Solutions: A Unique Case Report of a Severe Exudative Retinal Detachment Accompanied by Retinal Traction and Two Retinal Holes in Coats Disease

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Keywords
Coats disease · Retinal detachment · Scleral buckle

Abstract
Introduction: Coats disease is a rare vasculature pathology that usually presents as retinal telangiectasia with possible progression to exudative retinal detachment (RD). Intravitreal anti-VEGF injections, cryotherapy, laser photocoagulation, and surgery are commonly used to control the disease and prevent its progression. Although iatrogenic tractional RDs secondary to anti-VEGF injections have been reported in patients with Coats disease, RDs in Coats disease are exudative, secondary to retinal exudation and vascular abnormalities. In this article, we present the first reported case of a severe exudative RD accompanied by retinal traction and two retinal holes in a patient with Coats disease. ***Case Presentation:*** A 32-year-old male initially presented with Coats disease stage 2A, which then progressed to 3A1 within a month of close follow-ups, finally leading to exudative RD. Following cryotherapy and bevacizumab injections, the exudative RD progressed, with the emergence of retinal traction and two retinal holes. This complex case was successfully treated with a scleral buckle vitrectomy accompanied by radial elements to support the RD, pars plana vitrectomy, silicon oil tamponade, and post-surgical bevacizumab injections. Six months following the surgical intervention, the patient's vision is restored at 20/30, and retinal imaging shows a totally flattened retina.

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Conclusion: We present the first reported case of a severe exudative RD accompanied by retinal traction and two retinal holes in a patient with Coats disease. The combination of surgical treatment and bevacizumab injections is thought to have collectively contributed to our patient's favorable outcome.

Introduction

Coats disease was initially described by George Coats in 1908 as a unilateral retinal vascular anomaly characterized by the presence of exudation, primarily affecting boys in their early stages of life [1]. Throughout the years, our understanding of this condition led to the modern definition of this disease, characterized by retinal vascular telangiectasia and microvascular anomalies, frequently followed by intraretinal or subretinal exudation and retinal detachment (RD) [2]. Symptoms of Coats disease are mostly unapparent in the early stages of the disease until poor visual acuity, eye pain, glaucoma, squinting, and leukocoria occur, usually in the first 10 years of the patient's life [2]. Despite exhibiting a higher prevalence amongst young male patients, the origin of Coats disease remains uncertain as no definitive genetic or hereditary transmission has been established in relation to this condition [2, 3].

Visual prognosis and therapeutic options can be oriented by a Coats staging classification established by Shields et al. [4]; this classification offers guidance for the appropriate treatment plan and estimates the visual prognosis according to the evolution of the disease. Furthermore, subcategories within these stages enable a more specific classification based on the degree of detachment and foveal involvement [2]. Upon disease progression to stage 3, meaning the presence of RD, visual prognosis values are not available due to the possibility of more severe causes of vision loss developing during the course of the illness, such as tractional RD, affecting the prognosis [4].

In addition to typical ocular findings of Coats disease, such as retinal telangiectasia, intraretinal exudation, and retinal hemorrhage, up to 50% of the patients with Coats disease will suffer from RD, specifically exudative RD [5]. Although tractional RD has been found in patients with Coats disease, this type of RD is rarely part of the natural course of the disease; it is more frequently attributed to complications from intravitreal bevacizumab injections used in the treatment of Coats disease leading to iatrogenic vitreoretinal fibrosis, thus causing tractional RD [6]. Rhegmatogenous RD has also been reported in a few patients with Coats disease, underlying the variability in this disease [7]. Combined RDs have interestingly yet to have been linked to Coats disease other than exudative RDs combined with iatrogenic tractional RD linked to bevacizumab injections [8].

Different therapeutic tactics for treating RD in Coats disease have been described, including laser photocoagulation, cryotherapy, surgical intervention, and intravitreal anti-VEGF injections [2]. Cryotherapy is an effective initial treatment for extensive exudation and RD associated with Coats disease and is commonly used in patients with stage 2–3A Coats disease [2]. However, risks of increased retinal traction on the retina leading to tractional RD are associated with cryotherapy [2]. Recent advancements in combined treatment plans for Coats disease, including a combination of laser photocoagulation and intravitreal anti-VEGF injections, have shown promising results in Coats disease-associated exudative RD, even leading to the resolution of Coats-associated RD in all 24 patients in a small patient series by Villegas et al. [9]. Although these results may seem promising, intravitreal anti-VEGF injections with bevacizumab as a treatment for Coats-related exudative RD should be used cautiously, as

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some cases of vitreous fibrosis and traction RD have been linked to the use of bevacizumab injections [6]. Extensive RD and RD refractive to initial treatments in advanced Coats disease are treated by vitreoretinal surgery, specifically vitrectomy, which is considered to play a vital role in the management of complications in advanced cases of the disease; however, visual outcomes of surgical treatments vary significantly [1, 2]. In the late phases of Coats disease, vitreoretinal surgery can be utilized as a means to mitigate anterior segment issues and uphold the anatomical integrity of the eye in the optics, avoiding complications leading to enucleation [2, 10]. Finally, complicated Coats disease presenting with neovascular glaucoma and/or other refractory symptoms such as vomiting, pain, and nausea may lead to the necessity of enucleation [11]. In this case report, we describe a case of severe exudative RD accompanied by retinal traction and two retinal holes in a patient with Coats disease who underwent combined surgery of pars plana vitrectomy and encircling scleral buckle, leading to RD resolution. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary Material (for all online suppl. material, see https://doi.org/10.1159/000535821).

Case Presentation

A 32-year-old generally healthy male patient with no history of eye disease nor significant family history presented with a complaint of floaters in his right eye for 3 months with preserved visual acuity and −0.50 refraction in both eyes. Upon examination, the anterior segment in both eyes and the left fundus were within normal limits. Dilated fundus evaluation of the right eye revealed telangiectasia of the retinal vessels, with subretinal exudation in the superonasal quadrant, not involving the macula, as shown in Figure 1. At this point, the patient was diagnosed with Coats disease stage 2A according to the Shields classification, supported by the presence of telangiectasia and extrafoveal exudation.

After thoughtful consideration of our patient's initial presentation, it was decided to follow up closely with our patient; this decision was made per recommendations from the Shields guidelines established by Sen et al. [1], stating that patients with little or no exudation not threatening the vision can be followed up closely, and treatments such as photocoagulation can be initiated upon progression. In the following month, our patient developed exudative RD in the superonasal quadrant. Consequently, his staging went up from 2A to 3A1, signifying that an intervention should be done to stabilize the retina and prevent further RD to preserve the patient's vision. The area of the retinal lesion was initially treated with cryotherapy and bevacizumab injections, and close follow-ups were scheduled. Three weeks later, the vision deteriorated to 20/200 BCVA, and the exudative RD progressed toward the macula, accompanied by a new traction on the superonasal region of the retina. In a meticulous examination with a scleral depression, we saw two holes near the traction area. We realized that we were facing a complex case of severe exudative RD accompanied by retinal traction and two retinal holes. Dilated fundus evaluation in LE showed telangiectasia of the retinal vessels with subretinal exudation in the superonasal quadrants and the presence of subretinal fluid in the superior area extending into the fovea. There was also the presence of fibrosis with traction on the superonasal part with two holes under it around the 11 o'clock position.

Consequently, the patient underwent a scleral buckle with 360° broader bands (e.g., 42 band, 4 mm width). A radial element was cut from the band and placed under the band on the superior nasal quadrant to support the tractional RD. A pars plana vitrectomy with membrane peeling was combined with the scleral buckle to alleviate the traction and allow the retina to reattach. Silicon oil tamponade was then performed to stabilize the retina, and the patient received one injection of bevacizumab post-vitrectomy to address the retinal exudation.

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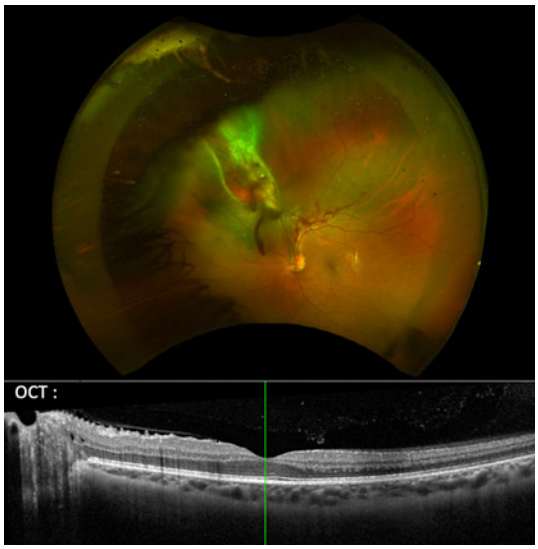

Fig. 1. Telangiectasias supero-temportal and nasal to the macula, with exudation not involving the macula In a patient with 2A Coats disease.

One week post-vitrectomy, his visual acuity improved in the affected eye from 20/200 BCVA to 20/60 BCVA with a refraction of +4.00 + 0.50cyl@85 due to the combination of silicone oil tamponade and scleral buckle. The recovery was uneventful following the surgery. Fundus imaging 1 week post-operation showed that the retina is attached with the presence of residual exudation in the superonasal quadrant, and OCT imaging illustrated that the fovea was successfully reattached, also showing the residual subretinal fluid on the temporal side (Fig. 2). In the following months, the patient received bevacizumab injections every 8 weeks since exudation was still present and leaking in the pathological area was suspected. One month following the initial bevacizumab injection, the retina was seen as entirely flat, the bleeding was almost completely absorbed, and no residual subretinal fluid remained in the macula. Finally, at the 6-month post-vitrectomy follow-up, the retina was totally flattened, showing no signs of traction and exudative RD, as seen in Figure 3, and his vision improved to 20/30 BCVA with a refraction of +4.00 + 0.50cyl@85. The silicon oil tamponade is still present in the vitreous chamber.

Discussion

Our patient experienced the first reported case of a severe exudative RD accompanied by retinal traction and two retinal holes. The surgical treatment explained above allowed the recuperation of our patient's vision and the recovery of this rare type of triple-combined RD. His visual acuity currently stands at 20/30 BCVA with a refraction of +4.00 + 0.50cyl@85 with the presence of silicon tamponade in the vitreal chamber.

Exudative RD is part of the usual disease progression of Coats disease. In fact, a comprehensive study conducted by Shields et al. [5] involving a significant sample size of 158 eyes affected by Coats disease showed that a mere 19% of the assessed eyes exhibited no signs of RD, exposing the importance of RD in this disease. Common treatment for Coats-related exudative RD with Shields staging 3A is similar to what was executed in our patient's case, including cryotherapy and the more recent addition of anti-VEGF injections of bevacizumab [9]. The addition of bevacizumab to Coats-related RD has shown promising results; however, it does not come without inevitable risks, as seen in a retrospective study by Bhat et al. [8] showing that the combination of cryotherapy with bevacizumab injections resulted in the

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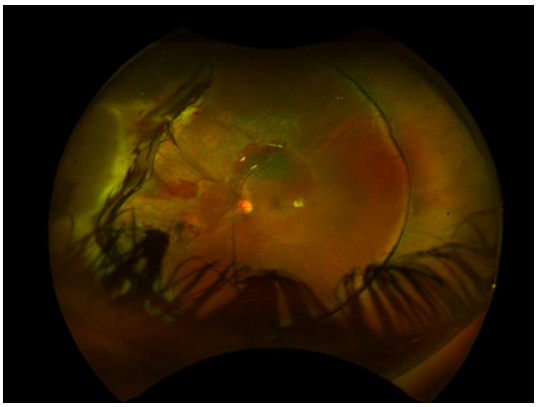

Fig. 2. One week after the combined surgery of 360° scleral buckle with radial element, pars plana vitrectomy with silicone oil. The retina is attached with bleeding and residual exudation in the superonasal quadrant.

attachment of the retina in 85.7% of their patients with exudative RD, although 75% of the patients in this study had iatrogenic tractional RD following bevacizumab injections [6, 9]. These findings could explain the initial progression of our patient's condition from exudative RD with Coats disease to tractional RD with Coats disease. However, the appearance of the rhegmatogenous RD simultaneously with the tractional RD has yet to be reported in a similar situation, underlining the importance of our article in understanding the vast variability of Coats disease-related RD.

Pars plana vitrectomy for advanced Coats disease remains the surgical treatment of choice, although this surgery has always been challenging in patients with this disease [2]. Scleral buckle combined with vitrectomy has been shown to be effective in the resolution of the RD and in preventing further complications of Coats disease, as seen in a study by Li et al. [12] These findings correlate with our patient's positive response to pars plana vitrectomy with scleral buckle, leading us to believe that the addition of external support by the placement of radial elements to the scleral buckle, combined with the epiretinal membrane peeling, the silicone oil tamponade, and the pars plana vitrectomy, simultaneously contribute to our patient's positive surgical outcome in this case of a very complex RD. In addition to vitrectomy, epiretinal membrane stripping in patients with advanced Coats disease seems to have favorable results for visual prognosis [2]. Hence, this procedure was performed on our patient with an original Coats-related RD who now has recovered from a 20/200 BCVA visual acuity to a 20/30 BCVA vision, supporting the effectiveness of epiretinal membrane peeling during surgery in patients with Coats disease.

It is interesting to note that only a small number of cases of rhegmatogenous RD in Coats disease have been documented. In one of these cases, the patient's vision was preserved after performing an encircling scleral band buckle and vitrectomy with the placement of silicone tamponade [9]. A similar procedure was performed for our patient. This underlines the effectiveness of vitrectomy with scleral band buckles for RD in Coats disease, showing promising results in a case of a severe exudative RD accompanied by retinal traction and two retinal holes. Our findings, along with others, aim to pave the way for further surgeons facing unfamiliar situations with patients with this rare disease.

Combined RDs are more frequent in conditions such as proliferative diabetic retinopathy, where combined rhegmatogenous and tractional RDs can be found [13]. To treat this condition, intensive surgical procedures like pars plana vitrectomy with intravitreal silicone oil have been shown to improve postoperative visual acuity [13]. The addition of an encircling scleral buckle to lessen residual traction on the retina and promote retinal reattachment can also be combined with this procedure to offer more stability to the retina [13]. Although diabetic retinopathy and Coats disease are two different entities, it is essential to note that

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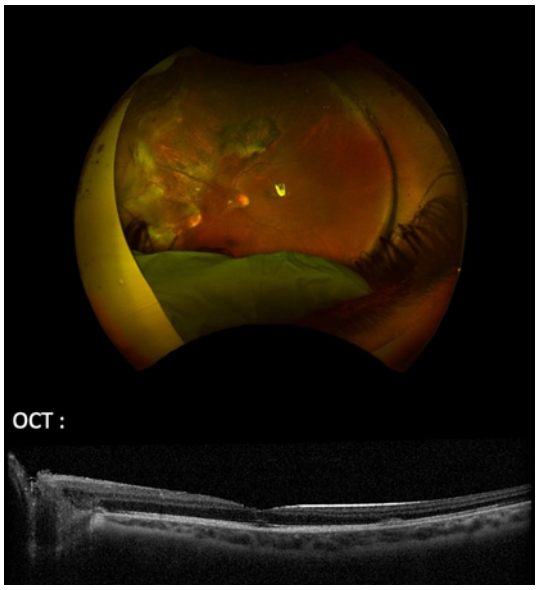

Fig. 3. Six months following pars plana vitrectomy, scleral buckle, and silicon oil tamponade of the left eye with the vision improved to 20/30. No subretinal fluid or exudation in the OCT scan.

combined RDs of other etiologies have been treated successfully in a similar manner as our surgical interventions, supporting the effectiveness of this procedure in our patient's case.

The steady improvement of our patient's vision after surgical treatment is consistent with results from other cases of Coats disease with subretinal exudation treated with bevacizumab injections [9]. As reported by Zhao et al. [14], VEGF concentrations seem to be directly correlated with the severity of Coats disease, and this explains directly how an anti-VEGF injection such as bevacizumab results in the significant improvement of exudation and RD in cases such as our patients.

A notable limitation of this study arises from the absence of similar reported cases in the literature, allowing us to compare ocular findings to precisely diagnose this case as a triple combined rhegmatogenous, exudative, and tractional RD. The lack of similar cases also limits the interpretation of the patient's outcome since no other surgical or medical treatments can be compared to our surgical management in this particular situation. Moreover, as with every case report, the primary limitation is the inability to generalize the findings and establish a cause-and-effect relationship.

We are herein reporting the first case of a severe exudative RD accompanied by retinal traction and two retinal holes and an effective surgical and medical management method for this rare presentation. This case report contributes to the field by proposing an effective management method for a rare condition that could be reproduced in similar situations to achieve optimal patient care.

Conclusion

Due to the rare nature of the condition and the wide range in severity at the time of manifestation, Coats disease therapy should be personalized. The combination of surgical treatments, including scleral buckle with radial elements to support the traction, pars plana vitrectomy, silicon oil tamponade, and the use of post-surgical bevacizumab injections, contributed to our patient's positive outcome in his case of severe exudative RD accompanied by retinal traction and two retinal holes in a patient with Coats disease. Recent advancements in retinal imaging, enhanced diagnostic precision, and refined surgical methodologies have all

played a pivotal role in yielding improved patient results. The timely identification of a condition and diligent monitoring are crucial in order to achieve optimal outcomes. Finally, this study highlights the diverse manifestations observed in one of the most uncommon ocular conditions and proposes a potential therapeutic approach that can be implemented in clinical practice.

Statement of Ethics

Ethical approval was not required for this study in accordance with local/national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors declare that the research was conducted without any commercial or financial relationship that could be constructed as a potential conflict of interest.

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Author Contributions

Simon Elsliger and Efraim Berco contributed to the writing of the manuscript draft. Efraim Berco, Tamir Weinberg, Waleed Ghannam, and Nir Shoham-Hazon contributed to the initial conception and methodology of the study. All authors contributed to the revision and approved the submitted version.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material files. Further inquiries can be directed to the corresponding author.

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