ARTYKUŁ PRZEGLĄDOWY REVIEW ARTICLE

DOI: 10.24292/01.OT.060726

Article ID: A0009

Coats disease: diagnosis and treatment

Natalia Wolińska[1], Julia Kaczmarek[1], Monika Papież[1], Alan Pigoński[1], Natalie Papachristoforou[2], Anthony Ueno[2], Aneta Pyza[1], Jakub Piórek[1], Ismael Alsoubie[3], Maciej Kozak[1, 4]

[1] Andrzej Frycz Modrzewski University in Kraków, Faculty of Medicine and Science, Collegium Medicum
Dean: prof. UAFM Janusz Ligęza, PhD
[2] SP ZOZ MSWiA, Krakow
Head: Michał Zabojszcz, MD, PhD
[3] 4th Military Clinical Hospital with Polyclinic, Independent Public Health Care Facility in Wroclaw
Commander: gen. bryg., Wojciech Tański, MD, PhD
[4] Provincial Ophthalmological Hospital in Kraków, Clinical Ophthalmology Department, Section „A" Head of Department: Maciej Kozak, MD, PhD

HIGHLIGHTS

The key to successful treatment of Coats' disease is early diagnosis and prompt initiation of treatment, primarily based on minimally invasive methods such as laser therapy and cryotherapy.

ABSTRACT

Coats disease is a rare, non-hereditary retinopathy. It is characterised by progressive destruction of the retinal blood vessels with exudates and aneurysms usually occurring unilaterally. Despite several studies, the etiology of this disease remains unclear. Treatment varies depending on the stage of the disease. Currently, laser therapy and cryocoagulation are commonly used. The disease is progressive; therefore, early diagnosis and treatment reduce the risk of severe complications.

Key words: Coats disease, telangiectasias , exudation, retina

INTRODUCTION

Coats' disease as an idiopathic eye disease is characterised by telangiectasias, intraretinal and subretinal exudates, and aneurysms [1].

If untreated, it can result in neovascular glaucoma. According to a population-based study, its incidence is 0.09 per 100 000 [2]. The disease affects boys and girls in a 3:1 ratio, with an average age of onset between 8 and 16 years. Nevertheless, de novo incidence can occur in adults up to the eighth decade of life [3]. First described in 1908 by ophthalmologist George Coats, it was characterised as a monocular retinal vascular abnormality with exudation occurring in young male patients [1, 4].

Subsequently, Theodor von Leber described a condition in which there were retinal telangiectasias and multiple aneurysms without subretinal exudate [1].

In 1955, Reese presented the similarities of Coats' disease with Leber's military retinopathy. Their primary pathology is retinal telangiectasia leading to progressive retinal exudation and detachment over time [4].

A wide spectrum of clinical manifestations of which reduced visual acuity is the first to manifest and strabismus, nystagmus, are its subjective manifestations. Corneal oedema, a shallow anterior chamber, iris neovascularisation and secondary resulting iris heterochromia and cataracts occur despite it being primarily a retinal vascular abnormality. In some cases, the condition may remain asymptomatic and is incidentally diagnosed during fundus examination, especially in older patients [4].

DIAGNOSIS

The basis of diagnosis is fundoscopic examination using indirect ophthalmoscopy. In all cases, retinal telangiectasias can be observed. Due to their shape and extensive exudation, they are described as "lightbulb telangiectasias".

These abnormal vessels usually occur in the peripheral zones taking on a spindle shape. The inferior and temporal retinal quadrants are the most commonly affected [4]. According to Shields' classification, there are 5 stages of Coats' disease. This classification describes the severity of the disease based on the presence of telangiectasias, exudation leading to retinal detachment and secondary complications [5]:

- Stage 1: Retinal telangiectasias only.
- Stage 2a: Telangiectasias and extrafoveal exudation.
- Stage 2b: Telangiectasias and foveal exudation.
- Stage 3a: Subtotal exudative retinal detachment.
- Stage 3b: Total exudative retinal detachment.
- Stage 4: Total detachment with secondary glaucoma.
- Stage 5: Advanced end-stage disease.

TREATMENT

Cryotherapy

Cryotherapy is a method of treatment for Coats' disease, primarily indicated for patients with peripheral telangiectasia accompanied by extensive exudation and retinal detachment. Cryotherapy uses a double or triple freezing and de-freezing technique, which allows it to be performed 2 or 3 times during the same session under indirect ophthalmoscopy [6].

A therapeutic interval of 4–6 weeks is used to assess the response to treatment. In case of incomplete efficacy of cryotherapy, treatment is repeated [7]. There is some evidence that cryotherapy may promote the formation of the epiretinal membrane and increase the risk of tractional retinal detachment secondarily [8]. An significant complication of severe destruction of the blood–retinal barrier after cryotherapy may also be an increase in subretinal fluid shortly after surgery. Despite these risks, cryotherapy remains an effective treatment for Coats' disease [6].

Laser photocoagulation

Laser photocoagulation in Coats' disease was first described by Meyer-Schwickerath and has historically been the most effective treatment [9]. It is indicated in the early-stage of Coats' disease (stages 1–3a) with exudation or flat retinal detachment [4].

Laser photocoagulation involves destruction of the abnormal vasculature and minimisation of the exudate. A conventional green or yellow laser with a duration of (0.1 to 0.5 s) is preferred for effective vascular obliteration [10]. Laser photocoagulation converts monochromatic light energy into thermal energy, resulting in an increase in the temperature of the absorbing tissue. This results in denaturation of proteins and coagulation of the tissue [11]. The absorptive capacity of the absorbing tissue is determined by its absorption spectrum in relation to the incident laser wavelength. In the case of Coats' disease, green light is sufficient to directly photocoagulate vascular abnormalities even in the presence of subretinal fluid [12]. The effectiveness of treatment is influenced by the number of retinal quadrants with vascular abnormalities. More than 2 vascular abnormality-laden quadrants of the retina reduce the chance of treatment success, so the period after ablative therapy requires close monitoring [5].

Anti-VEGF therapy

Studies on the use of anti-VEGF therapies in the treatment of Coats' disease point to their potential as an adjunctive therapy, especially in combination with other treatments such as cryotherapy or laser photocoagulation. The effect of anti-VEGF is to reduce vascular permeability, leading to a reduction in macular oedema and subretinal exudates. This offers the possibility of more effective laser therapy [13, 14]. Further studies are needed to establish the efficacy and safety of anti-VEGF therapy in Coats' disease [15].

Intravitreal corticosteroid therapy

Intravitreal corticosteroid therapy involves the administration of steroids, particularly by injection into the vitreous body of the eye. The treatment appears to have a moderate effect by reducing lipid exudate, which is a characteristic symptom of Coats' disease [16]. Research has shown that triamcinolone acetate can significantly reduce exudates, however, its use is associated with the risk of complications such as increased intraocular pressure and the risk of developing cataracts. An alternative to the use of triamcinolone in Coats' disease is intravitreal dexamethasone used particularly in cases resistant to other treatments. Studies show that the use of dexamethasone can have a positive therapeutic effect in the treatment of macular oedema, in addition, this steroid has a higher safety profile than triamcinolone, but the risk of complications, including an increase in intraocular pressure, also exists [15].

Surgical treatment

Surgical intervention in Coats' disease is recognized in advanced stages of the disease, particularly when satisfactory outcomes have not been achieved with laser photocoagulation or cryotherapy. The primary surgical procedure employed in the treatment of Coats' is vitrectomy, particularly in cases of retinal detachment or persistent subretinal exudation.

In addition, surgical treatment is also employed prior to other therapeutic methods, such as before laser therapy, to remove accumulated subretinal fluid. Surgical interventions, like other therapeutic approaches for Coats' disease, carry inherent risks and potential complications, they may lead to e.g. vitreoretinal proliferation [15, 17]. Vitrectomy surgery has a fundamental role in the late stages of the disease (especially stage 3b and higher according to the Shields classification). In extreme cases, when the disease leads to neovascular glaucoma, pain and loss of vision, enucleation, i.e. removal of the eyeball, may be necessary, which is the end of treatment in approximately 16% of patients [5, 13, 18, 19].

PROGNOSIS

The prognosis of Coats' disease is determined by several factors, including the age of the patient, the stage of the disease at the time of diagnosis as well as the effectiveness of the treatment implemented. Younger age at the time of diagnosis is associated with a more severe course of the disease, ultimately leading to vision loss over time. To prevent complications and ensure better therapeutic outcomes, it is essential to initiate treatment in the early stages of the disease, which underscores the importance of prompt diagnosis [2]. A randomised study by Shields et al. showed a correlation between the advanced stage of Coats' disease and poorer visual acuity after treatment. In stages 1 and 2, the telangiectatic vessels disappeared completely or partially several months after treatment. Patients in stages 4 and 5 of the disease, due to the course of the disease, may require enucleation surgery [5].

TREATMENT COMPLICATIONS

The course of Coats' disease is progressive. The occurrence of spontaneous remissions of the disease is rare. Untreated Coats' disease can lead to complications such as neovascularization, followed by intraocular hemorrhage, cataract, iris rubeosis, and neovascular glaucoma [1]. Judisch et al. report that orbital connective tissue inflammation can also be a complication of Coats' disease [20].

CONCLUSION

Coats' disease is a condition that leads to significant vision impairment, especially if diagnosed at a later stage. Early diagnosis and prompt initiation of treatment, primarily based on minimally invasive methods such as laser therapy and cryotherapy, can significantly improve the patient's prognosis.

CORRESPONDENCE
med. stud. Natalia Wolińska
Andrzej Frycz Modrzewski University in Kraków, Faculty of Medicine and Science, Collegium Medicum
30-705 Kraków, ul. Gustawa Herlinga-Grudzińskiego 1
e-mail: 1natawol1@gmail.com

ORCID
Natalie Papachristoforou – ID – https://orcid.org/0009-0006-8417-3794
Ismael Alsoubie – ID – https://orcid.org/0000-0002-0974-3025
Aneta Pyza – ID – https://orcid.org/0009-0000-5318-1368
Julia Kaczmarek – ID – https://orcid.org/0009-0001-2268-6547
Jakub Piórek – ID – https://orcid.org/0009-0001-4744-1933
Natalia Wolińska – ID – https://orcid.org/0000-0003-0975-9163
Anthony Ueno – ID – https://orcid.org/0009-0008-6946-4096
Maciej Kozak – ID – https://orcid.org/0000-0001-7993-2588
Alan Pigoński – ID – https://orcid.org/0009-0003-9955-0888
Monika Papież – ID – https://orcid.org/0009-0008-0397-7825

Cite as: Wolińska N, Kaczmarek J, Papież M et al. Coats disease: diagnosis and treatment. Ophthatherapy. 2026; 13(2): A0009.

References

1. Gupta A, Paulbuddhe VS, Shukla UV et al. Exudative Retinitis (Coats Disease). In: StatPearls. StatPearls Publishing, Treasure Island (FL) 2024.
2. Morris B, Foot B, Mulvihill A. A population-based study of Coats disease in the United Kingdom I: epidemiology and clinical features at diagnosis. Eye (Lond). 2010; 24(12): 1797-801. https://doi.org/10.1038/eye.2010.126.
3. Adeniran JF, Duff SM, Mimouni M et al. Treatment of Coats' disease: an analysis of pooled results. Int J Ophthalmol. 2019; 12(4): 668-74. https://doi.org/10.18240/ijo.2019.04.23.
4. Sen M, Shields CL, Honavar SG et al. Coats disease: An overview of classification, management and outcomes. Indian J Ophthalmol. 2019; 67(6): 763-71. https://doi.org/10.4103/ijo.IJO_841_19.
5. Shields JA, Shields CL, Honavar SG et al. Classification and management of Coats disease: the 2000 Proctor Lecture. Am J Ophthalmol. 2001; 131: 572-83.
6. Adam RS, Kertes PJ, Lam WC. Observations on the management of Coats' disease: less is more. Br J Ophthalmol. 2007; 91(3): 303-6. https://doi.org/10.1136/bjo.2006.103382.
7. Haik BG. Advanced Coats' disease. Trans Am Ophthalmol Soc. 1991; 89: 371-476.
8. Glaser BM, Vidaurri-Leal J, Michels RG et al. Cryotherapy during surgery for giant retinal tears and intravitreal dispersion of viable retinal pigment epithelial cells. Ophthalmology. 1993; 100(4): 466-70. https://doi.org/10.1016/s0161-6420(93)31620-9.
9. Spitznas M, Joussen F, Wessing A. Treatment of Coats' disease with photocoagulation. Albrecht Von Graefes Arch Klin Exp Ophthalmol. 1976; 199(1): 31-7. https://doi.org/10.1007/BF00660813.
10. Ridley ME, Shields JA, Brown GC et al. Coats' disease. Evaluation of management. Ophthalmology. 1982; 89(12): 1381-7. https://doi.org/10.1016/s0161-6420(82)34634-5.
11. McGrand JC. Photocoagulation in Coats' disease. Trans Ophthalmol Soc UK. 1970; 90: 47-56
12. Nucci P, Bandello F, Serafino M et al. Selective photocoagulation in coats' disease: ten-year follow-up. Eur J Ophthalmol. 2002; 12: 501-5.
13. Ghorbanian S, Jaulim A, Chatziralli IP. Diagnosis and treatment of coats' disease: a review of the literature. Ophthalmologica. 2012; 227(4): 175-82. https://doi.org/10.1159/000336906.
14. Bai J, Song Z, Li G et al. Efficacy and Safety of Anti-Vascular Endothelial Growth Factor Drugs for Coats' Disease Treatment: A Systematic Review. J Ocul Pharmacol Ther. 2023; 39(7): 418-29. https://doi.org/10.1089/jop.2023.0028.
15. Yang X, Wang C, Su G. Recent advances in the diagnosis and treatment of Coats' disease. Int Ophthalmol. 2019; 39(4): 957-70. https://doi.org/10.1007/s10792-019-01095-8.
16. Oli A, Balakrishnan D, Jalali S. Coats' disease: trends and long-term treatment outcomes in a tertiary referral centre. Ther Adv Ophthalmol. 2021; 13: 25158414211055957. https://doi.org/10.1177/25158414211055957.
17. Kodama A, Sugioka K, Kusaka S et al. Combined treatment for Coats' disease: retinal laser photocoagulation combined with intravitreal bevacizumab injection was effective in two cases. BMC Ophthalmol. 2014; 14: 36. https://doi.org/10.1186/1471-2415-14-36.
18. Berco E, Elsliger S, Weinberg T et al. 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. Case Rep Ophthalmol. 2024; 15(1): 84091. https://doi.org/10.1159/000535821
19. Sigler EJ, Randolph JC, Calzada JI et al. Current management of Coats disease. Surv Ophthalmol. 2014; 59(1): 30-46. https://doi.org/10.1016/j.survophthal.2013.03.007.
20. Judisch GF, Apple DJ. Orbital cellulitis in an infant secondary to Coats' disease. Arch Ophthalmol. 1980; 98(11): 2004-6. https://doi.org/10.1001/archopht.1980.01020040856014.

Authors' contributions:
All authors have equal contribution to the paper.

Conflict of interest:
None.

Financial support:
None.

Ethics:
The content presented in the article complies with the principles of the Helsinki Declaration, EU directives and harmonized requirements for biomedical journals.