**Vitrectomy for epiretinal membrane in adult-onset Coats’ disease**

Pradeep Kumar, Vinod Kumar

Coats’ disease is characterized by retinal vascular telangiectasia and subretinal and intraretinal exudation. A relatively benign form of the disease that occurs in adults is referred to as adult-onset Coats’ disease. Involvement of macula in the form of macular edema and exudation are the common presenting features in both forms of the disease. We describe a rare case of adult-onset Coats’ disease that presented with epiretinal membrane (ERM). Laser photocoagulation of retinal vascular telangiectasia resulted in worsening of patient’s symptoms and ERM. Early pars plana vitrectomy resulted in resolution of the patient’s symptoms. Utility of ultra-wide-field imaging and rationale of early vitrectomy in such cases are discussed.

**Key words:** 25-gauge, adult onset, Coats’ disease, epiretinal membrane, vitrectomy

Coats’ disease is an idiopathic disorder characterized by telangiectatic and aneurysmal retinal vessels with intraretinal and subretinal exudation and fluid accumulation.\[1\] The disease is characteristically seen in children, but a variant can be seen in adults and is referred to as adult-onset Coats’ disease.\[2\] Adult-onset Coats’ disease is characterized by less extensive involvement, more benign natural course, and more favorable treatment outcome as compared to Coats’ disease in the young. Although the presence of telangiectasia at macula is seen in only 1% of cases, macular exudation and edema can occur in up to 75% and 37% of cases with Coats’ disease, respectively, and are common presenting features.\[1\] The occurrence of epiretinal membrane (ERM) in Coats’ disease is rare, and in a large series by Rishi et al.,\[3\] the incidence of ERM was 4.4%. ERM in Coats’ disease can also be seen as a result of laser photocoagulation or cryopexy.\[4\]

In view of the poor long-term outcome, the main focus of treatment in Coats’ disease in the past has been the management of telangiectatic vessels in the form of laser photocoagulation or transscleral cryotherapy. Vitreoretinal surgery is commonly used to manage the retinal detachment secondary to exudation and allow successful laser or cryoablation of abnormal retina. However, vitrectomy for the macular involvement in Coats’ disease has been limited to a few case reports.\[5,6\]

Pars plana vitrectomy (PPV) for ERM management in juvenile\[7\] and adult-onset Coats’ disease\[8\] has been described scarcely. We report a case of adult-onset Coats’ disease who presented with metamorphopsia and decreased visual acuity due to ERM. Symptoms of the patient worsened after laser ablation of vascular telangiectasia. The patient was managed successfully with early intervention by 25-gauge PPV.

**Case Report**

A 32-year-old man presented with complaints of metamorphopsia in his right eye (RE) for 1-month duration. There was no prior history of intraocular inflammation, surgery, or trauma. His best-corrected visual acuity (BCVA) was 20/25 in the RE and 20/20 in left eye (LE). Anterior segment examination of both the eyes was unremarkable. Fundus examination of the RE showed an ERM involving the supertemporal arcade with inner retinal folds radiating toward the fovea [Fig. 1a] and the presence of multiple peripheral bulb-like dilatations of vessels in the temporal periphery [Fig. 1b]. Ultra-wide-field (UWF) fluorescein angiography (Optos Inc.,) demonstrated typical light bulb-like dilatations of telangiectatic vessels characteristic of Coats’ disease along with interspersed capillary nonperfusion (CNP) areas [Fig. 1c]. Spectral domain optical coherence tomography (SD-OCT, Carl Zeiss Inc.,) confirmed the presence of ERM, an intact ellipsoid zone, and inner retinal folds in the area nasal to fovea [Fig. 1d]. The LE fundus was normal. A diagnosis of adult-onset Coat’s disease with ERM was made.

Dr. R. P. Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Correspondence to: Dr. Vinod Kumar, Dr. R. P. Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi - 110 095, India. E-mail: eyepradeep@yahoo.com

Manuscript received: 08.05.17; Revision accepted: 03.08.17

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

Cite this article as: Kumar P, Kumar V. Vitrectomy for epiretinal membrane in adult-onset Coats’ disease. Indian J Ophthalmol 2017;65:1046-8.
After obtaining informed consent, the supertemporal lesion along with the CNP areas as depicted on the UWF autofluorescence (UWFA) was subjected to laser ablation (532 nm green laser). No surgical intervention was recommended as the patient was comfortable with his visual status and a regular follow-up was advised. At 8 weeks' follow-up, the patient reported worsening of metamorphopsia and diminution of vision in his RE. BCVA RE was 20/40. SD-OCT showed worsening changes in the inner retina as well as external limiting membrane Fig 1e.

After informed consent, the patient underwent 25-gauge PPV, posterior vitreous detachment induction, ERM and internal limiting membrane peeling, fluid-air exchange, and endolaser augmentation of supertemporal Coats’ lesion. The patient was advised standard postoperative regimen with topical steroids and cycloplegic drops.

At 4 weeks' follow-up, the patient reported the improvement in metamorphopsia and BCVA in the RE improved to 20/30. Dilated fundus examination showed normal appearing macula [Fig, 2a]. Laser photocoagulation scars were seen in the temporal periphery on UWFA [Fig, 2b]. SD-OCT revealed macula devoid of ERM along with flattened foveal contour [Fig, 2d]. At 6 months, the patient had no complaints of metamorphopsia and BCVA (RE) improved to 20/25.

Discussion

ERMs have been described in Coats’ disease of pediatric as well as adult onset.[4,6,7] Pathogenesis of ERM formation in Coats’ disease involves chronic macular edema,[1] vitreomacular traction,[8] and/or reactive gliosis, leading to ERM formation.[9] Laser photocoagulation done for the retinal vascular telangiectasia and resultant inflammation also contribute toward occurrence of ERM. In the present case, metamorphopsia as a result of ERM was the presenting feature of Coats’ disease. This has not been reported in the literature to
the best of our knowledge. ERM gradually progressed over a period of 8 weeks to alter the foveal structure and worsen the visual symptoms. This progression was attributable to the laser photocoagulation done for the retinal vascular telangiectasia.[8]

UWF imaging over the past decade has revolutionized the diagnosis and management of peripheral retinal disorders. We previously reported the utility of UWF imaging in the diagnosis and management of adult-onset Coats’ disease, including the present case.[10] It helped in the exact localization of telangiectasia and CNP and follow-up of patients with Coats’ disease.

Vitrectomy for macular diseases in Coats’ disease has been reported very rarely.[4-7] The authors reported good visual outcomes following PPV for such cases. With the availability of small gauge sutureless vitrectomy systems, safety and outcome of vitreoretinal surgeries have improved significantly. We decided on early intervention in this case because of the worsening symptoms of the patient. This resulted in early improvement and resolution of the patient’s symptoms. Early intervention is especially important since permanent macular changes can occur in patients with longstanding ERMs.

Conclusion
Adult-onset Coats’ disease may present with ERM and resultant metamorphopsia. A careful retinal peripheral examination is thus warranted in all young patients with ERM. Laser photocoagulation for the treatment of primary pathology (retinal vascular telangiectasia) may worsen the symptoms associated with such ERMs. Early PPV and removal of ERM provide good visual and anatomic outcomes in such cases.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. Shields JA, Shields CL, Honavar SG, Demirci H. Clinical variations and complications of coats disease in 150 cases: The 2000 Sanford Gifford memorial lecture. Am J Ophthalmol 2001;131:561-71.
2. Rishi E, Rishi P, Appukuttan B, Uparkar M, Sharma T, Gopal L, et al. Coats’ disease of adult-onset in 48 eyes. Indian J Ophthalmol 2016;64:518-23.
3. Rishi P, Rishi E, Uparkar M, Sharma T, Gopal L, Bhende P, et al. Coats’ disease: An Indian perspective. Indian J Ophthalmol 2010;58:119-24.
4. Yadav NK, Vasudha K, Gupta K, Shetty KB. Vitrectomy for epiretinal membrane secondary to treatment for juvenile Coats’ disease. Eye (Lond) 2013;27:278-80.
5. Wong SC, Neuwelt MD, Trese MT. Delayed closure of paediatric macular hole in Coats’ disease. Acta Ophthalmol 2012;90:e326-7.
6. Shukla D, Chakraborty S, Behera UC, Kim R. Vitrectomy for epimacular membrane secondary to adult-onset Coats’ disease. Ophthalmic Surg Lasers Imaging 2008;39:239-41.
7. Mino A, Mitamura Y, Katome T, Sembra K, Egawa M, Naito T, et al. Case of adult-onset coats’ disease treated with 25-gauge pars plana vitrectomy. J Med Invest 2013;62:85-8.
8. Gass JD. Stereoscopic Atlas of Macular Diseases: Diagnosis and Treatment. 3rd ed. St. Louis: Mosby; 1987.
9. Soman M, Ganekal S, Nair U, Nair K. Effect of panretinal photocoagulation on macular morphology and thickness in eyes with proliferative diabetic retinopathy without clinically significant macular edema. Clin Ophthalmol 2012;6:2013-7.
10. Kumar V, Chandra P, Kumar A. Ultra-wide field imaging in the diagnosis and management of adult-onset coats’ disease. Clin Exp Optom 2017;100:79-82.