Vitrectomy for full-thickness macular hole in adult-onset Coats' disease

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The occurrence of full thickness macular hole in Coats' disease is extremely rare. The purpose of this case report is to report pars plana vitrectomy for the treatment of full thickness macular hole in a patient with adult onset Coats disease. A young male presented with decreased vision in his right eye because of full thickness macular hole. The macular hole was found to be associated with adult onset Coats' disease that was evident on ultra-wide field imaging. The patient underwent laser photocoagulation to the vascular telangiectasia followed by pars plana vitrectomy, large internal limiting membrane peeling and gas tamponade. This resulted in regression of exudation, closure of macular hole and improvement in vision. Coats disease of adult onset can present with decreased vision because of full thickness macular hole. Vitrectomy with internal limiting membrane peeling can result in excellent visual outcome.

Key words: Adult-onset Coats' disease, macular hole, pars plana vitrectomy, ultra-wide field imaging

Coats' disease is a retinal vascular disease of unknown cause that mostly affects males in the first or second decade of life.[1] The clinical manifestations include retinal vascular telangiectasia, intraretinal and subretinal exudation, capillary nonperfusion (CNP), and exudative retinal detachment. A variant occurring in adults is known as adult-onset Coats' disease and tends to have a limited area of involvement, slower progression, and more posterior involvement.[2,3] The common macular changes include macular exudation, macular edema, epiretinal membrane (ERM), and fibroglial nodule.[1,2,4] Macular hole is a rare occurrence in Coats' disease, with only a handful of reported cases.[5-9] Surgical closure of the macular hole was attempted in only one report.[9] We report a case of macular hole in a patient with adult-onset Coats' disease that was managed successfully with pars plana vitrectomy (PPV) and internal limiting membrane (ILM) peeling.

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Case Report

A 35-year-old male presented with diminution of vision in the right eye of 5-month duration. There was no history of prior ocular disease, trauma, or systemic complaints. The best-corrected visual acuity (BCVA) was 20/400 in the right eye and 20/20 in the left eye with +1.5 D sphere in both the eyes. Anterior segment examination of both eyes was unremarkable. The left eye fundus was normal. Fundus examination of the right eye showed a full-thickness macular hole (FTMH) with ERM [Fig. 1a]. The inferior retina showed a large area of yellowish subretinal exudation with overlying telangiectatic retinal vessels [Fig. 1b]. Swept-source optical coherence tomography (SS-OCT) through the fovea (DRI, Triton Topcon Inc.,) confirmed an FTMH with overlying ERM [Fig. 1c]. The minimum diameter of hole was 790 microns while base diameter was 1375 microns. Ultra-wide field imaging (UWF, Optos Inc.,) captured the macular hole and peripheral lesion simultaneously [Fig. 1d]. UWF fundus fluorescein angiography (FFA) [Fig. 1e] revealed diffuse peripheral vascular leakage, window defect corresponding to the macular hole, and staining corresponding to the subretinal exudation. An inferiorly steered UWF FFA showed light bulb dilatations of the retinal blood vessels interspersed with CNP, overlying the area of exudation in peripheral retina [Fig. 1f]. A diagnosis of adult-onset Coat's disease with FTMH was made.

FFA-guided laser ablation of inferotemporal lesion and CNP areas was done (532 nm Green laser). After 6 weeks, the patient underwent 25-gauge PPV. The posterior vitreous detachment (PVD) was already present. After ERM peeling, a large area of ILM (1.5–2 disc diameters) around the hole was peeled with the help of Brilliant Blue-G dye (Ocublue Plus™, Aurolab, Madurai, India). This was followed by tamponade with 20% Sulfur hexafluoride gas. The patient was advised to maintain a face down position for 48 h along with the standard postoperative regimen of topical steroids and cycloplegic drops.

At 2-week follow-up, the BCVA had improved to 20/100. Dilated fundus examination showed complete apposition of hole edges with the retinal tissue [Fig. 2]. OCT revealed Type 1 macular hole closure.[10] Pigmented laser marks were seen on the telangiectasia in the inferior retina. Clinical picture remain unchanged at 26-weeks follow-up, with BCVA of 20/80.

Discussion

FTMH has been reported rarely with Coats' disease (pediatric or adult onset).[5-9] The pathogenesis of FTMH in such

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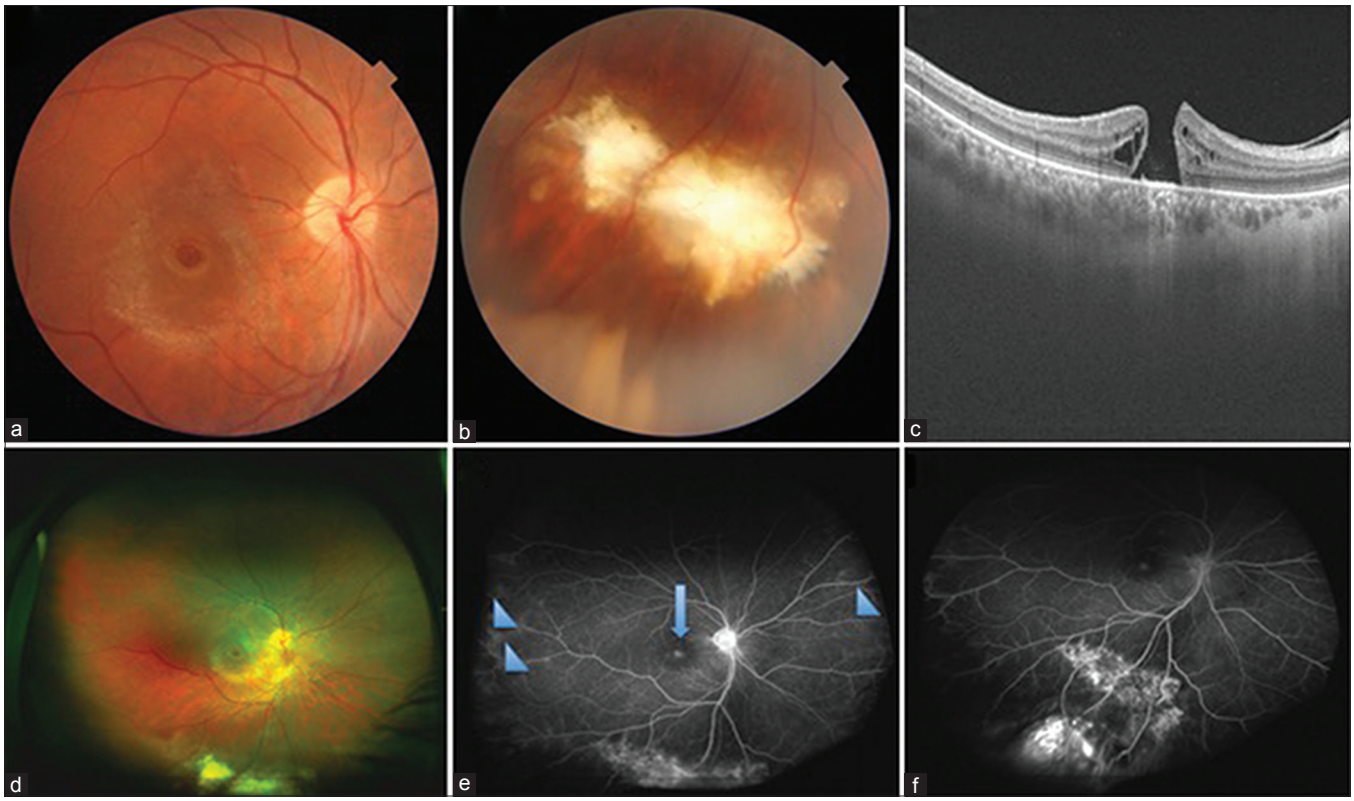

Figure 1: (a) Color fundus picture of the right eye showing full-thickness macular hole with epiretinal membrane. (b) Inferotemporal fundus showed subretinal exudation with overlying retinal telangiectasia. (c) Swept-source optical coherence tomography through fovea depicting full-thickness macular hole with the presence of epiretinal membrane. (d) Ultra-wide field color photograph could document full-thickness macular hole along with inferotemporal exudation and telangiectasia. (e) Ultra-wide field fluorescein angiogram revealed central window defect due to hole (arrow), peripheral vascular leakage (arrowheads), and retinal telangiectasia inferotemporally. (f) Inferior steered fundus fluorescein angiography showed light bulb dilatations characteristic of Coats' disease

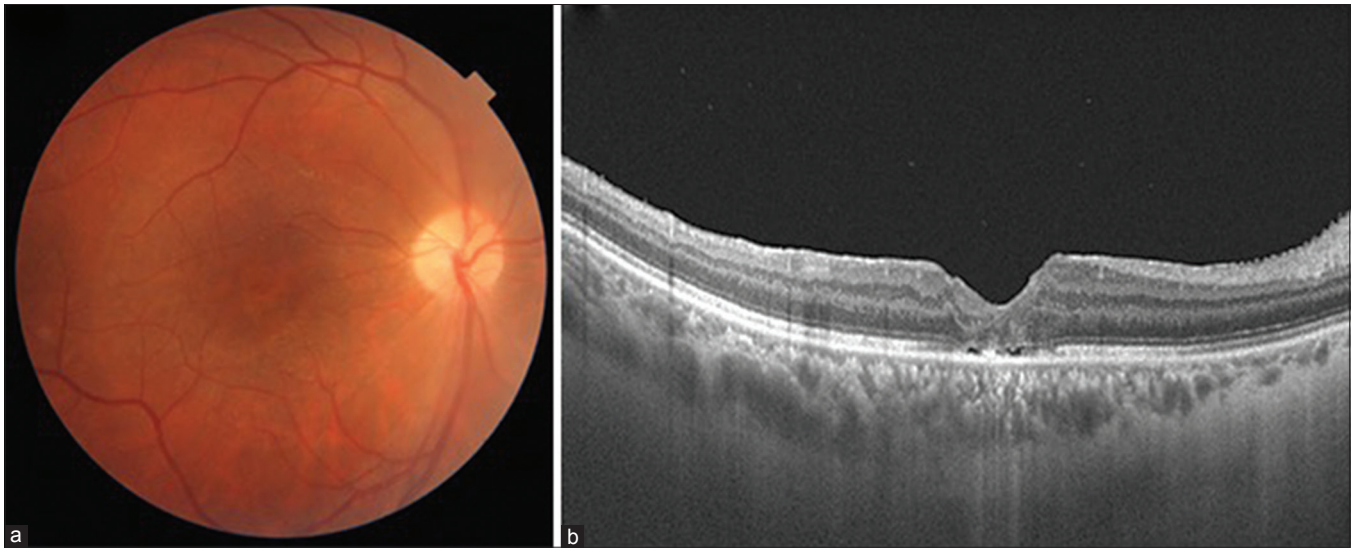

Figure 2: (a) Color fundus photograph at 2 weeks after vitrectomy showing apposition of macular hole edges. (b) Swept-source optical coherence tomography through fovea showed Type 1 macular hole closure

cases depends on several factors. These include chronic macular leakage leading to macular edema and consequent deroofing,[7] vitreous changes leading to vitreomacular traction,[9] and/or reactive gliosis leading to ERM formation.[11] In our case, vitreomacular traction was not present. SS-OCT and the presence of PVD noted intraoperatively substantiated

this. Tangential traction caused by ERM (seen clinically and on SS-OCT) may have a role in the causation of macular hole. Interestingly, widespread peripheral vascular leakage was seen on UWF FFA in this case [Fig. 1e]. The peripheral vascular leakage may lead to macular edema in spite of the vascular telangiectasia being located in the retinal periphery.[12] The presence of chronic macular edema could have contributed to the formation of a macular hole in this case.

With availability of UWF imaging, it is possible to image a large area of the retina. UWF imaging is useful for the management of patients with Coats' disease.[12] It is extremely valuable for documentation, which is important for patient counseling as well as medicolegal records.

Surgery in the form of PPV has been reported for the macular involvement in patients with Coats' disease. Most of these reports are for the treatment of ERMs and report good outcomes.[13-15] However, only a single case report exists that describe PPV for the treatment of FTMH in Coats' disease.[8] The authors noticed macular hole closure in this case 6 years after PPV, which is unlikely because of the surgery itself. Ours is the first report of successful closure of FTMH with 25-gauge PPV and ILM peeling, to the best of our knowledge. In this case, we planned PPV for FTMH 6 weeks after thermal laser to the vascular telangiectasia in periphery. During PPV, after ERM peeling, a large area of ILM was peeled (with a radius of 1.5-2 disc diameter size) to increase chances of hole closure.[16] This treatment plan took care of both the factors in causation of the macular hole (chronic leakage and tangential traction by the ERM/ILM). The patient showed excellent visual and anatomic recovery following PPV.

Conclusion

This report highlights that Coats' disease may present with FTMH hole. UWF imaging may be beneficial in the management of such cases. PPV with ILM peeling along with the treatment of the underlying cause (vascular telangiectasia) provided good visual and anatomic outcomes.

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Conflicts of interest

There are no conflicts of interest.

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