

A case of Leber's miliary aneurysms with diffuse peripheral retinal vascular sheathing

Anna Lee, Jiwon Baek, Ho Ra

A 30-year-old female presented with macular edema and discoid exudation at the posterior pole. Diffuse vascular sheathing was observed at the peripheral retina. Fluorescein angiography revealed multiple microaneurysms at the posterior pole and leakage from the peripheral vessels. Two monthly intravitreal bevacizumab led to minimal improvement, and resolution of macular edema was achieved by an additional intravitreal triamcinolone. The findings at the posterior pole resembled those of Leber's miliary aneurysm. However, this case also demonstrated a peculiar vascular sheathing at the periphery and showed response to triamcinolone, which are evidences for an inflammatory condition.

Key words: Coats' disease, inflammation, Leber's miliary aneurysms, vascular sheathing

Coats' disease is an idiopathic condition characterized by telangiectatic and aneurysmal retinal vessels with intraretinal and subretinal exudation and fluid.^[1] The term encompasses idiopathic retinal telangiectasias and exudation of varying severity from a mild form with no symptoms to a severe form with massive exudation and bullous retinal detachment. Leber's miliary aneurysm is a rare form of retinal telangiectasia characterized by the formation of multiple retinal vascular aneurysms, varying degrees of leakage, and deposition of lipid exudates.^[1,2] This entity is believed to belong to the same spectrum of diseases with Coats' disease but in a milder form.^[3] In this report, we present a patient with Leber's miliary aneurysms at the posterior pole and an uncommon finding of diffuse vascular sheathing at the peripheral retina.

Case Report

A 30-year-old female presented with decreased vision for 2 weeks in the right eye. Her visual acuity was 20/25 in the

right eye and 20/20 in the left eye. The anterior segments of both eyes were normal. The right eye displayed a two-disc diameter retinal thickening surrounded by hard exudates at the superotemporal posterior pole involving the fovea. Diffuse retinal vascular sheathing around the mid-to-far periphery was observed [Fig. 1a and b]. Ultrawide-field fluorescein angiography (FA) demonstrated multiple microaneurysms in the area of retinal thickening, and mild leakage and minimal nonperfusion with no definitive neovascularization at the periphery [Fig. 1c-f]. Spectral-domain optical coherence tomography (OCT) of the right eye showed subretinal and intraretinal fluid involving the fovea [Fig. 2a]. Her blood pressure was within normal limits. Laboratory tests, full blood counts, chest X-ray, tuberculin skin test, and serology for syphilis, toxoplasmosis, macroglobulinemia, and autoimmune antibodies (ANA and HLA B54) revealed no abnormalities. The patient was first treated with intravitreal bevacizumab (IVB) injection. A partial yet subtle response was observed after two monthly IVB injections [Fig. 2b and c]. Additional intravitreal triamcinolone injection (IVTA) was performed. At one month after IVTA injection, resolution of the macular edema was observed [Fig. 2d] and visual acuity returned to 20/20. Although exudates at the posterior pole decreased after IVTA, peripheral vascular sheathing was consistently observed during the monthly follow-ups afterwards [Fig. 3a]. Seven months later, recurrence of macular edema was observed [Fig. 3b]. Macular edema was resolved again with IVTA [Fig. 3c]. FA at the time of recurrence revealed more prominent vascular leakage from the sheathed vessels [Fig. 3d and e].

Discussion

Leber's miliary aneurysms are grouped as idiopathic retinal telangiectasias, along with idiopathic juxtapapillary retinal telangiectasias and Coats' disease. The disease is usually male-dominant, unilateral, adult-onset, and slow-progressing. Aneurysms are surrounded by hard whitish exudates and are most frequently found in the mid-to-remote peripheral temporal retina.^[4] Our patient was diagnosed with Leber's miliary aneurysms due to the pathognomonic characteristics at the temporal posterior retina.

For the etiology of Coats' disease, an infectious theory was once supported by several reports, but subsequent investigations and the failure of antiinflammatory and steroid therapies have refuted that early theory.^[1] Histologic studies of the retinal vasculature in Coats' disease have confirmed abnormal vascular integrity as the disease pathogenesis by demonstrating a thickened wall replaced by a laminated fibrous coating of membrane-like materials, patchy thinning, or the absence of the vessel wall, and some vessels completely devoid of endothelium and pericytes.^[5]

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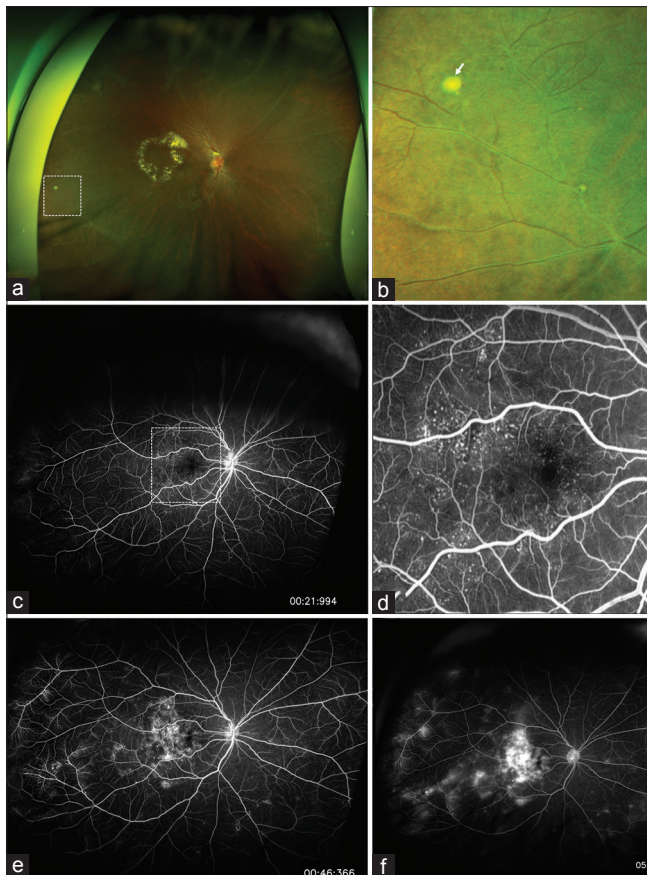


Figure 1: Initial ultrawide-field funduscopy and FA findings. (a) Discoid hard exudate is observed at the superotemporal posterior pole involving the fovea. At the periphery, diffuse vascular sheathing is present. (b) Magnified image of peripheral subset reveals diffuse sheathing and an aneurysmal dilatation (arrow). (c and d) Microaneurysms are observed in the posterior pole in the early-phase FA. (e) The intermediate-phase FA shows multiple leakages from aneurysms in the posterior pole as well as in the periphery. (f) The late-phase FA shows diffuse leakage from the temporal aneurysms and mild leakage with nonperfusion area at the nasal periphery

Diffuse vascular sheathing is not a common finding in Coats' disease or Leber's miliary aneurysms. Segmental vascular sheathing caused by yellow cholesterol deposits can be observed in Coats' disease, but diffuse white sheathing with leakage on FA is a finding suggestive of an inflammatory condition. Diffuse sheathing with vascular leakage on FA with no evidence of neovascularization at the peripheral retina was observed in the current case. Also, the macula edema responded better to treatment with IVTA than IVB. We suppose that there was an inflammatory component stronger than the usual Coats' disease. Nonetheless, we cannot ignore the fact that the protective role of steroids in Coats' disease includes not only the antiinflammatory effects, but also their role in the stabilization of the blood-retinal barrier, resulting in a decrease of vascular leakage from abnormal telangiectasia.

Differential diagnosis of unocular exudative retinopathy, including tuberculosis, toxoplasmosis, syphilis, leptospirosis, macroglobulinemia, and autoimmune diseases, should be considered. Systemic conditions such as hypertension, diabetes

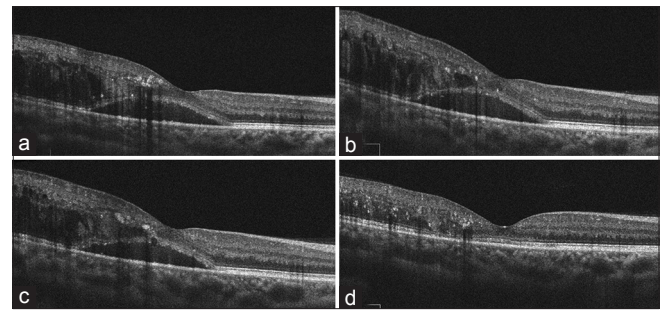


Figure 2: OCT findings of the patient at baseline and after treatment. (a) Baseline OCT image demonstrates subretinal and intraretinal fluid involving the fovea, with hard exudate seen as intraretinal hyperreflective dots. (b) OCT image 1 month after IVB injection shows no improvement in macular edema. (c) OCT image 1 month after the second IVB injection shows mild regression of the macular edema. (d) The treatment was switched to IVTA. OCT image 1 month after IVTA shows prominent improvement of macular edema

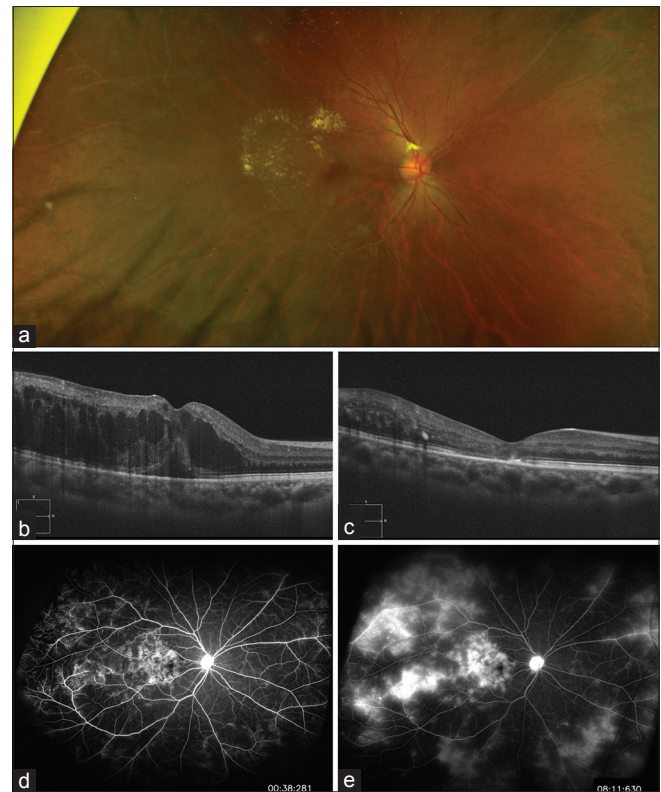


Figure 3: Follow-up funduscopy, FA, and coherence tomography (OCT) findings. (a) Ultrawide-field fundus color photography at 7 months after IVTA shows decreased exudates at the posterior pole with consistent peripheral vascular sheathing. (b) OCT at 7 months after initial intravitreal triamcinolone injection shows recurrence of macular edema. (c) Resolution of macular edema is observed on OCT image taken 1 month after IVTA. (d and e) Intermediate- and late-phase FA reveals diffuse vascular leakage from the posterior pole as well as from the peripheral sheathed vessels

mellitus, and hypercholesterolemia have also been reported with adult-onset Coats' disease.^[6,7] In the current case, a clear history and lab findings supported the idiopathic form of retinal exudation with telangiectasias that characterize

Leber's miliary aneurysms. Nonetheless, the possibility of a new disease entity in which vasculitis results in aneurysms cannot be ruled out.

Conclusion

In summary, we have presented a rare case of Leber's miliary aneurysms that coexisted with diffuse peripheral vascular sheathing. Although the disease is known to be caused by abnormal vessel integrity, it can also present with an inflammatory component. Steroid might be the most helpful treatment in such conditions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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