Multiple Evanescent White Dot Syndrome in a Case of Idiopathic Retinal Vascular Tortuosity

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Multiple evanescent white dot syndrome (MEWDS) is a unilateral chorioretinitis characterized by retinal multiple white dots caused by acute inflammation at the level of the outer retina and retinal pigment epithelium (RPE). It is a self-limiting disease which has a propensity to affect young, myopic females.^{1,2} Idiopathic retinal vascular tortuosity is an uncommon disease of retinal arteries and/or veins which may occasionally lead to hemorrhagic and occlusive problems.^{3,4} Here in, we report the coexistence, for the first time to our knowledge, of these two clinical entities.

A 26-year-old white male presented with blurry vision in the left eye for 2 weeks. Right eye examination revealed no abnormality besides retinal venous tortuosity (Figure 1, A), Left eye examination showed mild vitritis and retinal venous tortuosity. Small, subtle, discrete, white-yellow lesions were visible in the deep retina at the posterior pole to the midperiphery. Granular appearance in the central macular region and optic disc hyperemia were also observed (Figure 1, B-D). Optical coherence tomography (OCT) of the left eye revealed the discontinuity of the ellipsoid zone (Figure 1, F, arrows). Fundus autofluorescence (FAF) showed retinal venous tortuosity bilaterally, and hyper/hypoautofluorescent spots in the left eye (Figure 2, A,B). Fluorescein angiography (FA) showed no leakage from tortuous veins (Figure 2, C,D). Perifoveal, punctate hyperfluorescence and optic disc staining were seen in the left eye FA (Figure 2, D). ICG of the right eye was normal (Figure 2, E). On latephase indocyanine green angiography (ICG), multiple spots of hypofluorescence at the posterior pole and midperiphery were observed in the left eye (Figure 2, F).

Ocular and systemic conditions which may cause retinal vascular tortuosity were ruled out after thorough laboratory tests and imaging. The patient was diagnosed as bilateral idiopathic retinal vascular tortuosity and MEWDS in the left eye, and was followed up with no treatment. In follow up visits, white lesions resolved (Figure 3, A). Imaging confirmed complete resolution. (Figure 3, B-E).

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Figure 1a-e: Initial color fundus photos and optical coherence tomography (OCT) horizontal scans through foveola. A: Right eye with no abnormality but "corkscrew-like" appearance of retinal veins. B-D: Left eye examination revealed small, subtle, discrete, white-yellow lesions in the deep retina at the posterior pole to the midperiphery, granular appearance in the central macular region and optic disc hyperemia. Retinal venous tortuosity was also evident. E: Normal OD OCT. F: OCT of OS revealed the discontinuity of the ellipsoid zone at macular region (arrows).



Figure 2a-e: Initial visit fundus autofluorescence (FAF), fluorescein angiography (FA) and indocyanine green angiography (ICG) images. A,B: FAF of left eye showing speckled appearance with hyper/hypoautofluorescent spots. C,D: FA shows no leakage from tortuous veins in both eyes. In the left eye, perifoveal, wreath-like punctate hyperfluorescence due to leakage and staining of the optic disc can be seen. E,F: Right eye ICG is normal. Multiple spots of hypofluorescence at the posterior pole and midperiphery are visible at the late-phase ICG of the left eye.



Figure 3a-e: Follow up visit color fundus photo montage, horizontal enhanced depth imaging optical coherence tomography (EDI-OCT) scan through foveola, fundus autofluorescence (FAF), fluorescein angiography (FA) and indocyanine green angiography (ICG) of the left eye. A-D: FAF, FA and ICG confirmed almost complete resolution except subtle residual findings such as optic disc late staining and mild granularity on FAF and ICG. E: Ellipsoid zone also recovered as seen in EDI-OCT.