Response to comment on "Multimodal imaging in dominant cystoid macular dystrophy"

Sir,

We thank authors[1] for their interest in our article "Multimodal imaging in dominant cystoid macular dystrophy (DCMD)."[2]

Differential diagnosis of bilateral cystoid macular edema in a young adult with no systemic disorder or inflammation includes Goldman-Favre syndrome, retinitis pigmentosa, juvenile X-linked retinoschisis (XLRS), stellate nonhereditary idiopathic foveomacular retinoschisis, autosomal dominant cystoid macular edema, and various medication-induced maculopathies, such as niacin and paclitaxel.[3]

XLRS usually presents in the first or second decade of life with variable visual loss; on the contrary, patients with DCMD become symptomatic in the third decade like our patient who presented quite late at the age of 30 years.[4]

Both XLRS and DCMD can present with hyporeflective spaces in optical coherence tomography (OCT). However, their OCT features have subtle but definite differences. OCT in XLRS classically shows cavitations in neurosensory retina with thin vertical interconnecting septa and often become confluent, which is usually not the case with DCMD who have preponderance of cystoid spaces as in our case.[3]

Fundus fluorescein angiography (FFA) in XLRS shows no leakage. FFA in DCMD shows leakage of variable degree depending on the stage of the disease. Our case [Fig. 6 of publication] shows leakage albeit faint which can be appreciated in higher magnification. Fundus autofluorescence imaging in our case shows multispot hyperautofluorescence in fovea which can also be seen in XLRS. However, in addition to this, we find a broader area of hyperautofluorescence [Figs. 3 and 4 of publication] suggestive of diffuse retinal pigment epithelial dysfunction. This pattern of hyperautofluorescence is not seen in XLRS, which is predominantly an inner retinal disease to start with. These findings point more toward DCMD.[4,5]

We agree completely with authors regarding the utility of genetic testing, evaluation of family members and electrophysiological tests and accept the lack of them as a drawback of our report.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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