Purpose: To evaluate clinical, angiographic and optical coherence tomographical characteristics of eyes with butterfly-shaped pattern dystrophy.

Methods: Twenty-one eyes of 11 patients were diagnosed as butterfly-shaped pattern dystrophy. Clinical examination, spectral domain optical tomography (OCT) and fundus fluorescein angiography (FA) were applied. Patients were evaluated for need of any therapeutic intervention.

Results: Mean age was 67 (48-93). Seven patients were female. Follow-up period ranged between 1 to 24 months. Median best corrected visual acuity (BCVA) at presentation was 20/40 (counting fingers at 2 meters to 20/20). Four eyes were pseudophakic. Fundoscopy revealed retinal pigment epithelial (RPE) irregularities as linear and punctate hyper- and hypopigmentation giving several patterned appearance at the macula. FA revealed mostly speckled hyper and hypofluorescence of the lesion, serous pigment epithelial detachment, large window defect, hyperfluorescence secondary to active choroidal neovascular membrane (CNVM). OCT findings included; normal foveal contour except for PED and CNVM cases, irregular reflectence of RPE, ondulation, defect, thickening and stratification at the RPE-choriocapillaris hyperreflective band, foveal thinning, irregularity at IS/OS band, pigment migration and hyperreflectance at the subfoveal area secondary to CNVM scar. Low visual acuities were due to CNVM, large RPE atrophy and RPE irregularities. Intravitreal anti-VEGF injection was performed/indicated for 2 eyes for activation of CNVM /serous PED decreasing vision.

Conclusion: Pattern dystrophy causes RPE irregularities detected clinically and angiographically. OCT adds details to these findings as well as it is effective in evaluating any differences of retina and RPE integrity during the follow-up.