



# Butterfly-shaped Pattern Dystrophy: case series

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## INTRODUCTION

Butterfly-shaped pattern dystrophy (BPD) is part of a group of hereditary autosomal dominant macular diseases, related to mutations in the retinal degeneration slow (RDS)/peripherin gene <sup>(1, 2)</sup>. Due to its initial good visual prognosis, it is usually diagnosed in younger age and asymptomatic patients. However, central vision loss can occur by middle age <sup>(2)</sup>. Clinically, it is characterized by a bilateral accumulation of pigment at the level of retinal pigment epithelium (RPE), in the shape of the wings of a butterfly. Fundoscopically, yellow patches can be seen in the posterior pole <sup>(1)</sup>. In addition to fundoscopy, fluorescein angiography and autofluorescence can be made to help diagnosis and to distinguish this condition from other pattern dystrophies of the macula and other prevalent diseases such as age-related macular degeneration <sup>(3)</sup>. Besides that, optical coherence tomography (OCT) can also demonstrate the yellowish material under the sensory retina and above RPE.

## METHODS

Medical records review.

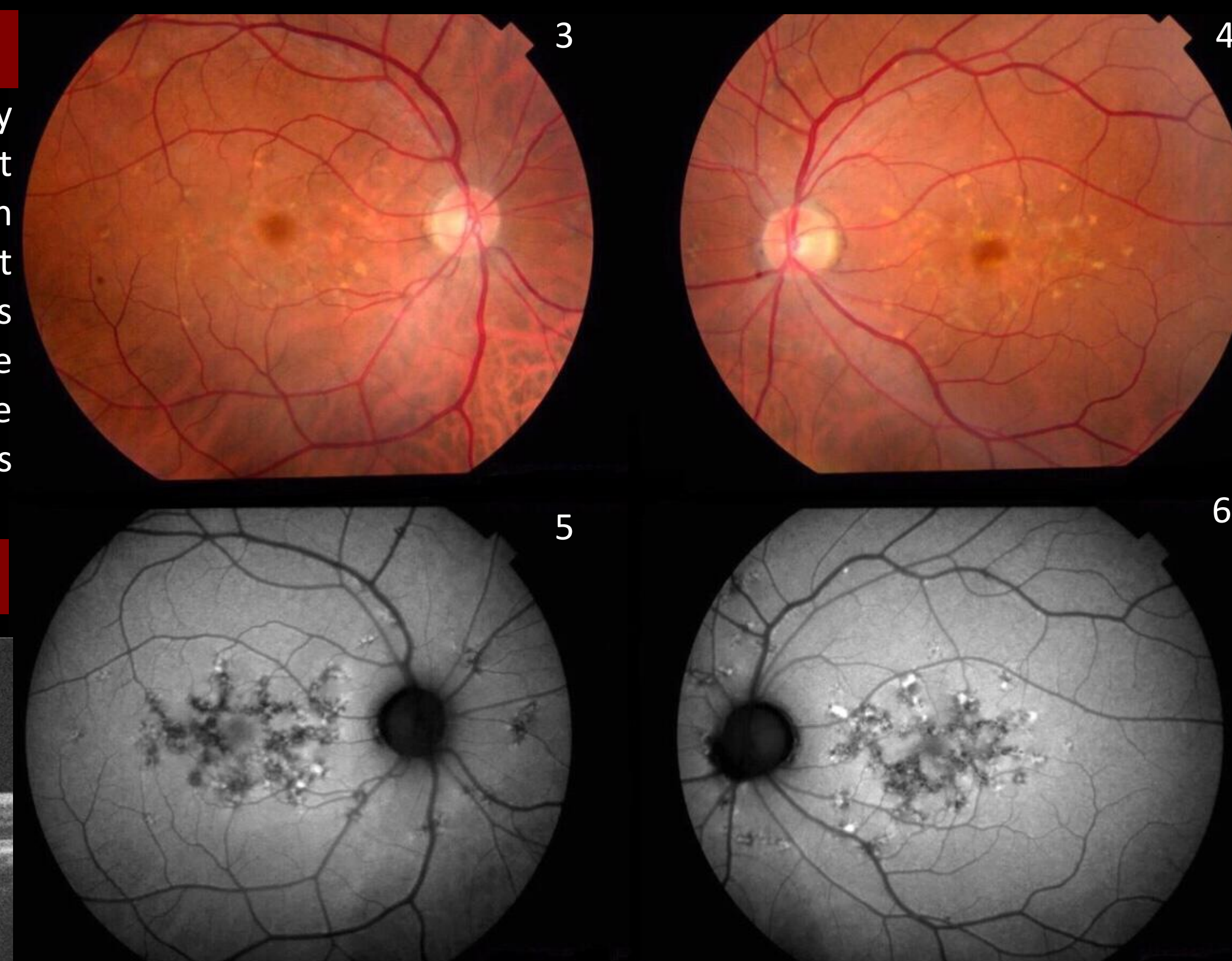
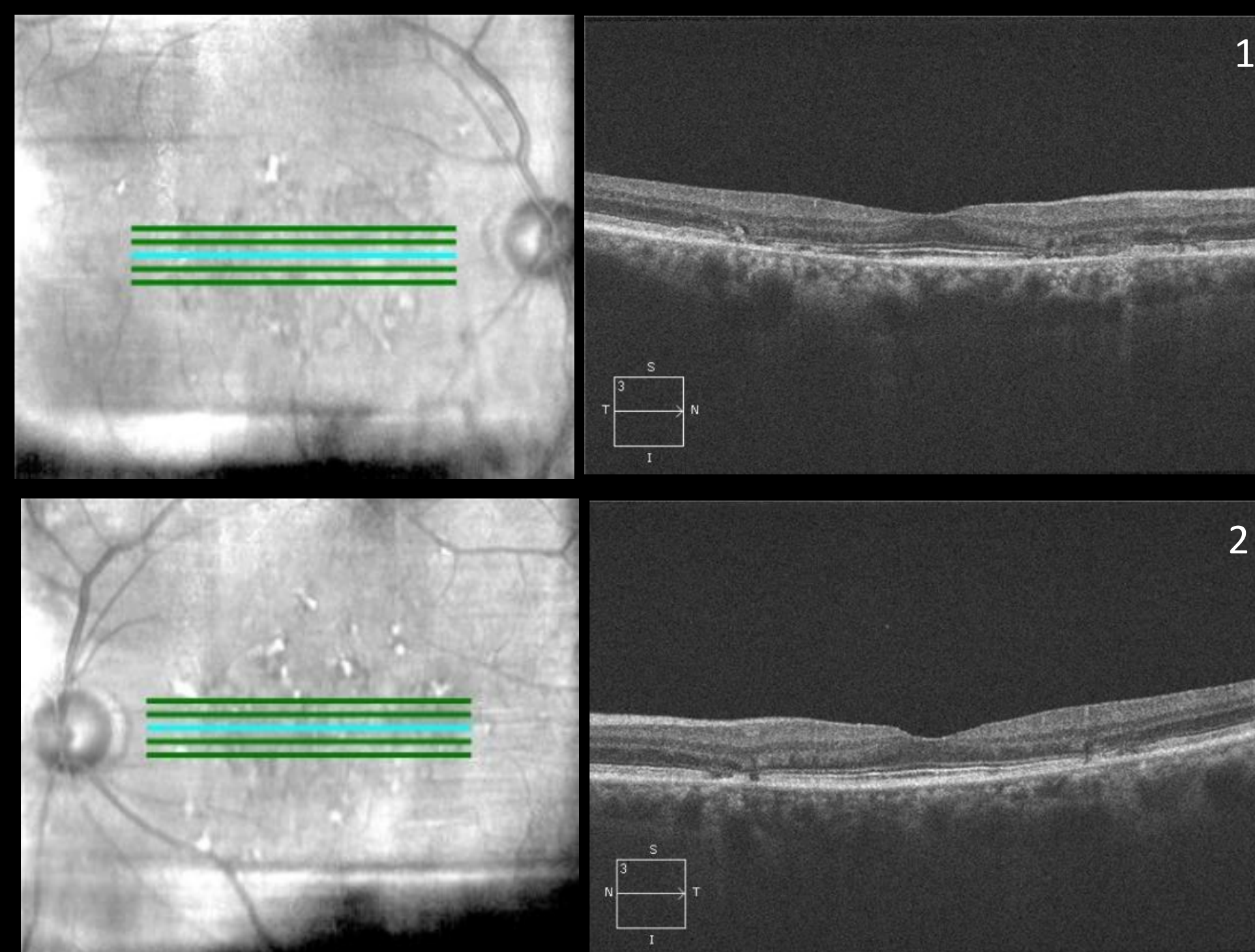
## RESULTS

We report three cases of BPD in patients with 64, 75 and 86 years old. All of them were complaining of progressive visual acuity loss. They were all pseudophakics, had no pathological antecedents and best visual acuity (BVA) was 20/40. Retinal fundoscopic exam showed bilateral pigmented material at the macula in two of them. The third one had yellow flecks in the posterior pole. Fluorescein angiography demonstrated the hypofluorescent classic butterfly-shaped macular lesion, in which the flecks observed fundoscopically were blocking fluorescence. OCT showed areas of ellipsoid layer and RPE atrophies.

## DISCUSSION

BPD is a chronic slow progressive disease, usually randomly diagnosed during routine examination. For this reason, most patients are asymptomatic at diagnosis but over the years it can progress with visual acuity impairment <sup>(2)</sup>. Although it was first suggested to be a benign condition, it is important to be aware of its complications such as macula failure due to RPE atrophy into the peripapillary region and/or choroidal neovascularization <sup>(4)</sup>. Despite the absence of a specific treatment for that condition, complications can be managed properly in order to reduce visual loss.

## IMAGES



Figures 1 and 2: OCT showing material above RPE; Figures 3 and 4: Retinography showing macular yellow patches; Figures 5 and 6: Fluorescein angiography demonstrating hypofluorescent butterfly-shaped macular lesion.

## REFERENCES

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