

## Evolution of Asymptomatic Pentosan Polysulfate Maculopathy Following Medication Discontinuation: A Case Report

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**Introduction:** Pentosan Polysulfate Sodium maculopathy (PPSM), first reported in 2018, is a newly characterized retinal disease associated with long-term use of PSS, a semisynthetic sulfated polysaccharide, which is the only Food and Drug Administration approved oral entity for interstitial cystitis. Most patients with experience progressive nyctalopia, central scotoma, and potentially severe vision loss. Asymptomatic cases have been seldom reported in the literature. Herein, we describe the clinical course of a patient receiving pentosan polysulfate (PPS) for interstitial cystitis, who developed asymptomatic PPS maculopathy (PPSM) that nonetheless persisted and evolved for four years after medication discontinuation.

**Methods:** Case report of one patient at a tertiary retina clinic. The charts of the given patient were reviewed for their clinic visits between 2016 and 2023.

**Results:** A 48-year-old female with a history of interstitial cystitis was referred to our retina clinic for query findings of age-related macular degeneration, despite endorsing no vision complaints. She had no relevant past ocular history. She had been taking PPS for 10 years for interstitial cystitis, first consuming 600mg/day for eight years, followed by 200mg/day for two years. At first presentation in 2016, examination revealed macular pigmentary clumping in both eyes. In subsequent years, pigmentary changes were confirmed by optical coherence tomography (OCT) which showed progressive macular thinning in both eyes. In 2019, the patient discontinued PPS; however, she continued to exhibit PPSM progression for years thereafter. At four-year follow-up after discontinuation of PPS, the patient returned with stable vision, but showed changes in macular lipofuscin deposits with outer retinal and retinal pigment epithelial alterations. Across the entire span of follow-up, she did not report a consistent constellation of visual symptoms, nor did she endorse the typical progressive symptoms of PPSM.

**Conclusions:** This case demonstrates sustained and evolving PPSM even after drug discontinuation, suggesting that active PPS may have a prolonged half-life in the retina and retinal pigment epithelium. Further, we report a rarer case of asymptomatic PPSM, despite clinically progressive findings.