

Characteristics of Pentosan Polysulfate Sodium-Associated Maculopathy and Similarities with Other Maculopathies Commonly Managed in a Retina Practice

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Summary

- Pentosan Polysulfate Sodium (PPS) is one of only a few FDA-approved treatments for Interstitial Cystitis (IC)
- Chronic PPS use recently associated with a progressive pigmentary maculopathy and choroidal neovascularization
- Here we present a retrospective analysis of 31 patients (62 eyes) taking PPS who were graded into three maculopathy groups
- Our data revealed a higher incidence of maculopathy in older patients and adds to evidence of an association with CNV
- This study highlights the similarities between PPS-associated maculopathy and ARMD - especially advanced cases - and the difficulties of managing these cases in a large retina practice

Interstitial Cystitis and Pentosan Polysulfate Sodium

- Interstitial cystitis is a chronic and debilitating retropubic pain syndrome
- PPS is one of only a few FDA-approved treatments for IC
- Chronic PPS use was recently associated with a progressive pigmentary maculopathy
- Progression of the maculopathy despite cessation of the drug and development of choroidal neovascularization have also been shown

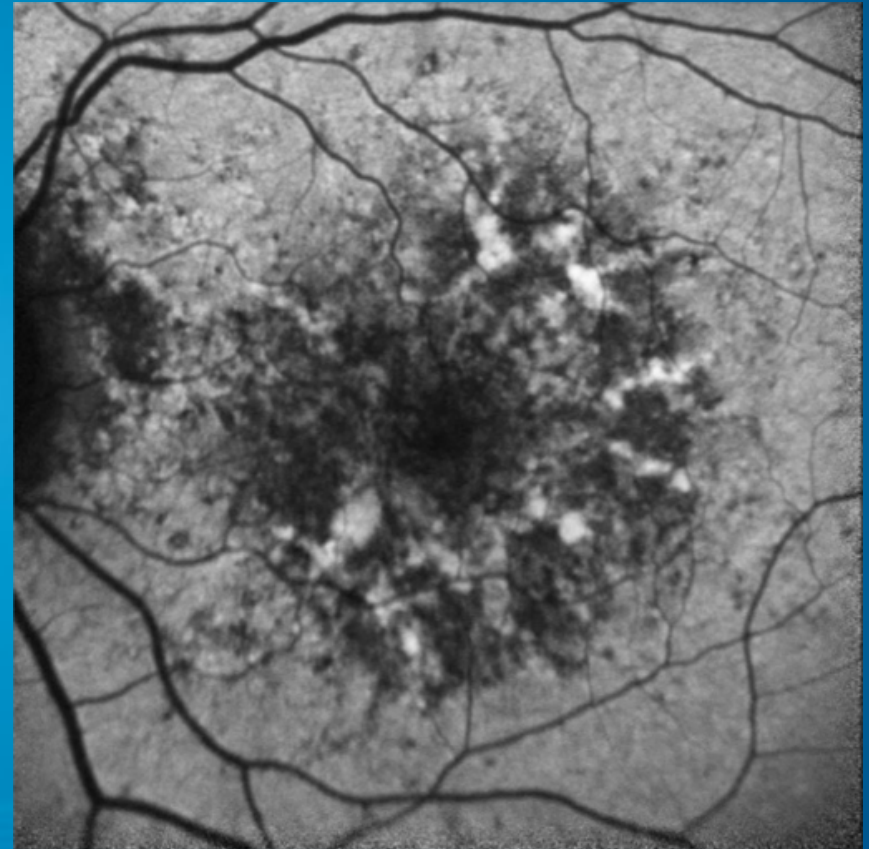
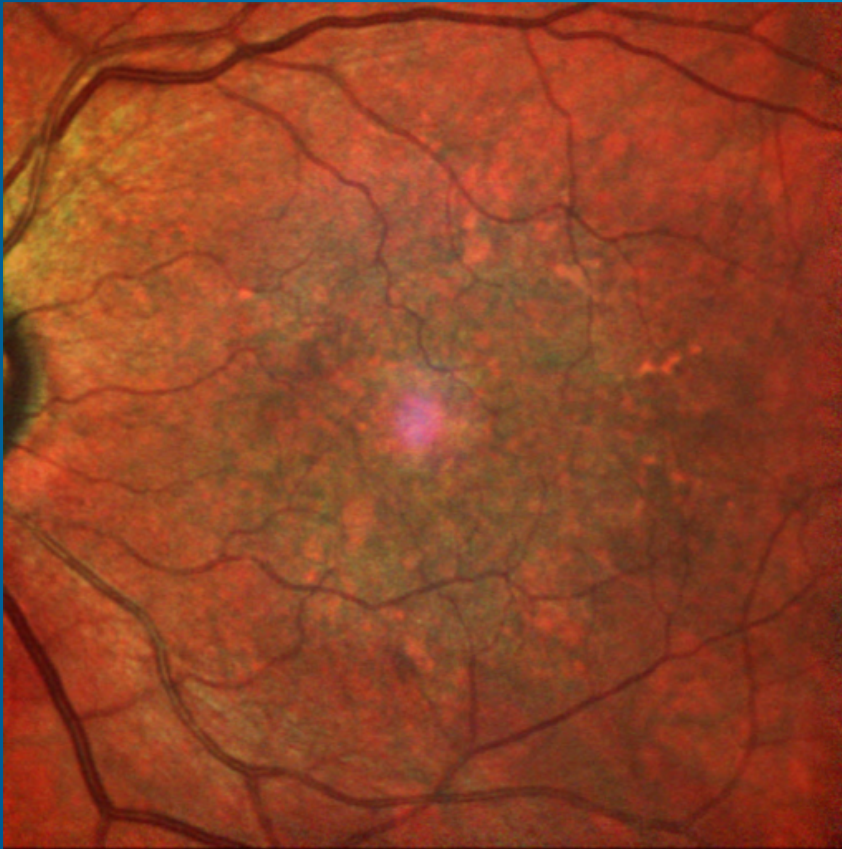
Purpose and Methods

- To describe characteristics of PPS-associated maculopathy and the similarities with other common maculopathies in a large retina practice
- IRB-approved retrospective analysis
- 62 eyes of 32 patients taking PPS
- Patients classified in one of 'Unlikely', 'Likely', or 'Possible' maculopathy groups

Maculopathy Grading

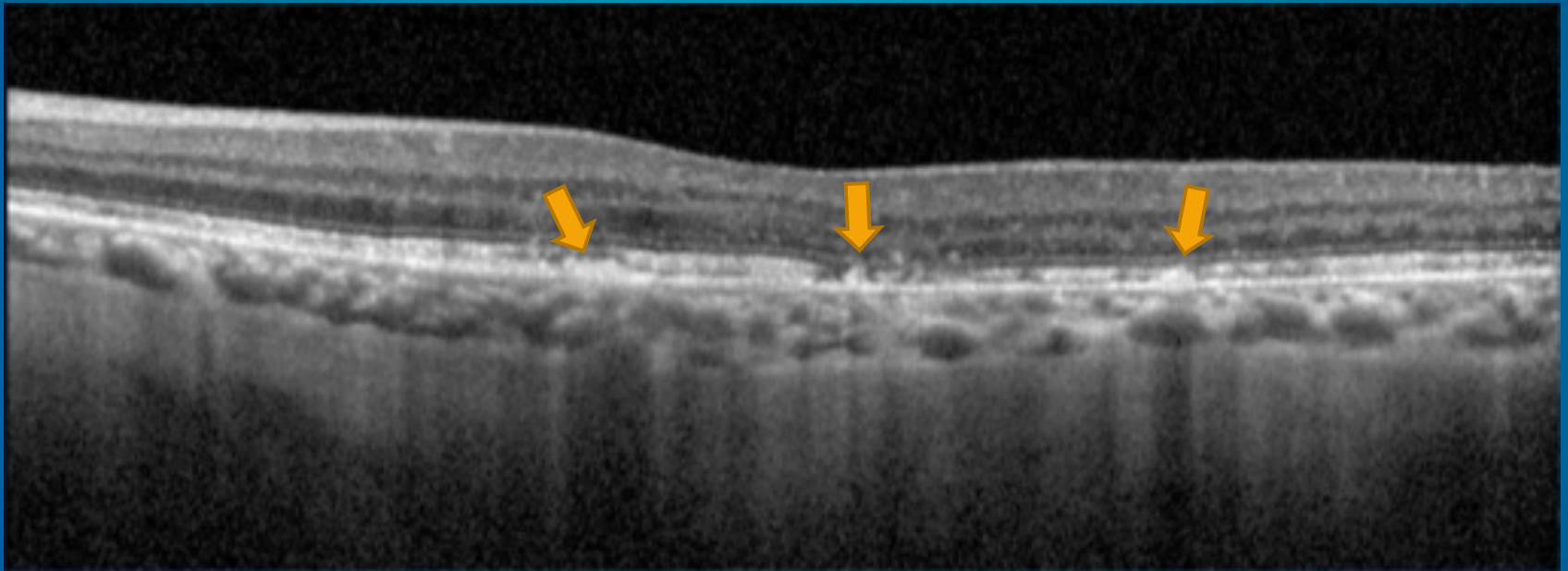
- Fundus imaging graded using characteristic features of PPS-associated maculopathy described by Pearce et al.
 - Macular hyperpigmented spots, yellow-orange deposits, and/or patchy RPE atrophy on color fundus photography
 - Dense array of macular and peripapillary alternating hyper- and hypo-autofluorescent spots and reticular changes on FAF
 - Nodular retinal pigment epithelium thickening on OCT corresponding to hyper-reflectance on near infrared reflectance imaging
 - Bilaterality

PPS-Associated Maculopathy



Color fundus photograph (left) and autofluorescence (right) of a Likely maculopathy patient demonstrating the aforementioned characteristics of PPS-associated maculopathy

PPS-Associated Maculopathy



Macular optical coherence tomography image portraying of a Likely maculopathy patient demonstrating nodular RPE thickening

Patient Characteristics

- Median age: 70 (range 24-104)
- Majority female (87%)
- Most common reason for referral was Age-Related Macular Degeneration (ARMD) (29%)
- 78% of patients diagnosed with ARMD after initial evaluation
- PPS-associated maculopathy grades were assigned as Likely (29%), Unlikely (45%), and Possible (26%)

Inter-Group Analysis

- Higher age at presentation in Likely group compared to Unlikely group ($p < 0.003$)
- No significant difference in race, gender, or smoking status between any of the maculopathy groups ($p > 0.05$)
- 78% of patients in the Likely maculopathy group received a diagnosis of ARMD after initial evaluation
- CNV was noted in 16% of all eyes
- CNV was noted in 11% of Likely maculopathy eyes

Limitations

- Absence of patient PPS cumulative dosing
- Inherent selection bias as many patients are referred to due to the presence of fundusoscopic or visual changes
- Retrospective nature
- Small patient cohort
- Lack of electrophysiologic testing and genetic analysis

Conclusions

- Our data represents that of one of the largest retina-only practices in the United States
- Adds to the growing evidence of a pigmentary maculopathy associated with PPS use
- Shows a higher incidence of maculopathy in older patients
- Suggests a more common incidence of CNV
- We encourage other practices to similarly review their records to identify at-risk patients

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