Pentosan polysulphate sodium associated maculopathy: a case in the United Kingdom

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INTRODUCTION

Interstitial cystisis (also known as bladder pain syndrome) is a chronic bladder condition characterise by pain, urinary urgency, frequency and nocturia (1,2). Intertitial cystitis (IC) may affect up to 400,000 people in the UK(3).

Pentosan polysulphate sodium is used for the treatment of interstitial cystitis (IC). It was approved for the treatment of IC by National Institute of Clinical Excellence (NICE) in 2019 (3). It was previously used off-license in the UK for the treatment of IC. Recent studies have linked Pentosan Polysulphate sodium (PPS) to a pigmentary retinopathy (1,2). The maculopathy is irreversible and linked to chronic exposure to PPS (4)

AIM

To report a case of a patient with Pentosan polysulphate sodium associated maculopathy in the United Kingdom.

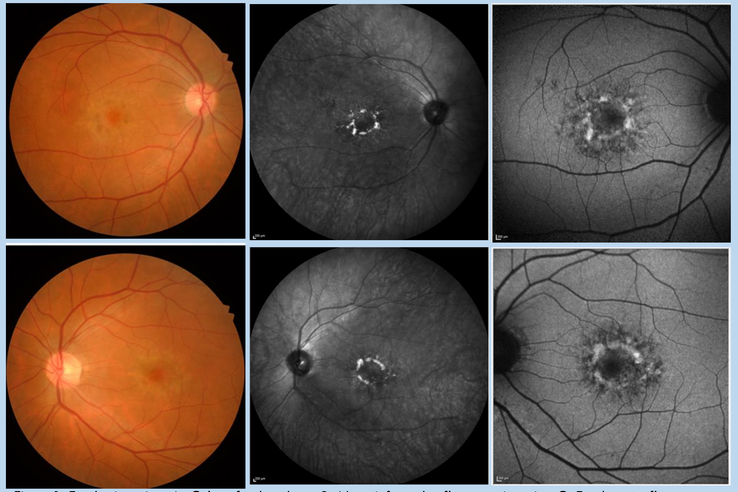


Figure1. Fundus imaging. A- Colour fundus photo. B- Near infrared reflectance imaging C- Fundus autofluorescence imaging of the right eye. Top row- right eye. Bottom row shows the left eye.

CASE REPORT

A 51 year-old Caucasian female presented to the Medical retina clinic with a suspected macular dystrophy and was referred to Retinal genetics clinic.

She complained of distortion in her vision, difficulty with reading and poor night vision. Her past medical history included reflux disease, hypercholesterolaemia and interstitial cystitis. Her medications included Pentosan polysulphate (PPS), Imipramine, Atorvastin, Cimentidine, Lansoprazole and Hydroxyzine. She had no family history of inherited retinal disease. She had been on PPS for 26years.

Best corrected visual acuity was 75 letters OD 76 letters OS. Examination of the anterior segment was normal.

RESULTS

Fundal examination	Bilateral parafoveal pigmented deposits at the level of the RPE.
Near infrared reflectance	Irregular reflectance pattern with hyperreflectant areas in the same area as the hyperpigmented spots noted on fundal examination.
Fundal autofluorescence imaging	Irregular autofluorescence pattern surrounded by normal fluorescence. Hyperautofluorescent spots align with the RPE deposits of the fundal examination.
OCT images	Small elevations at the level of the RPE which correspond to hyperreflectant lesions noted in the near infrared images
Functional studies	Visual field on standard automated perimetry was normal. Full field electroretinogram (ERG), pattern electroretinogram and electrooculogram were normal.

Table 1. Results

DISCUSSION

This is the first case of PPS maculopathy described in the United Kingdom. Pearse et al (1) first reported a case series of 6 patient in the United States with PPS maculopathy. Symptoms described by the patients in that study included dim vision, difficulty with near vision, paracentral scotoma and metamorphopsia. 4 eyes had paracentral RPE atrophy. 1 patient developed cystoid macula oedema (CMO) that responded to treatment with carbonic anhydrase inhibitors.

Deep scotoma on visual field testing associated with paracentral atrophy was seen in 3 eyes of 2 patients. Full field ERG showed mild attenuated delay with rod and cone responses in one patient.

PPS has been used off-license in the United Kingdom for the treatment of IC. NICE approval as a second line treatment for IC might spark an increase in the use of PPS and a possible increase in the associated maculopathy in the UK over time.

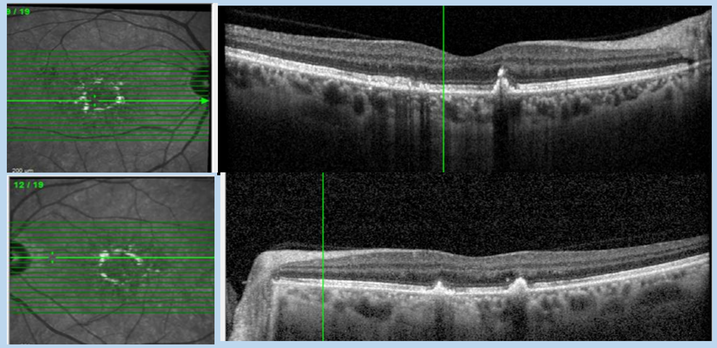


Figure 2 Cross section on optical coherence tomography

CONCLUSION

PPS associated maculopathy is related to chronic exposure to PPS. It seems to primary affect the RPE (1,2). The predominant symptoms are difficulty with reading and dark adaptation. Clinicians should be aware of the condition as it can be mistake for a macular dystrophy or age-related macular degeneration.

References

1. Pearce WA, Chen R, Jain N. Pigmentary Maculopathy Associated with Chronic Exposure to Pentosan Polysulfate Sodium. Ophthalmology. 2018;125(11):1793-1802.
2. Hanif AM, Armenti ST, Taylor SC, et al. Phenotypic Spectrum of Pentosan Polysulfate Sodium-Associated Maculopathy: A Multicenter Study [published online ahead of print, 2019 Sep 5]. JAMA Ophthalmology. 2019;137(11):1275-1282.
3. Pentosan polysulfate sodium for treating bladder pain syndrome (TA610)
4. Hanif A M, Shah R, Yan J, et al Strength of Association between Pentosan Polysulfate and a Novel Maculopathy Ophthalmology 2019 Oct;126(10):1464-1466.

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