A Case of Posner-Schlossman Syndrome
Christine Anggun Putri¹, Karl Mercieca²

Abstract
Posner-Schlossman syndrome (PSS), the eponymous name for glaucomatocyclitic crisis, is a rare cause of recurrent unilateral ocular hypertension associated with mild ciliary body inflammation (cyclitis). Due to the mild nature of inflammation, the diagnosis of PSS can be challenging. Medical and surgical treatments are indicated to reduce inflammation and to prevent long-term glaucomatous optic nerve damage related to the high intraocular pressure (IOP). This report describes a patient with PSS who had recurrent unilateral ocular hypertension associated with mild anterior uveitis over a four-year period. He subsequently developed glaucomatous optic head damage with an early visual field defect.

Keywords: posner-schlossmann syndrome, intraocular inflammation, glaucomatocyclitic crisis, glaucoma, trabeculitis, uveitis

Introduction
Posner-Schlossman syndrome (PSS), also known as glaucomatocyclitic crisis is described as recurrent attacks of unilateral increased intraocular pressure and mild inflammation of the ciliary body. This is a rare clinical entity with uncertain aetiology. There is a risk of progression of PSS to secondary chronic open angle glaucoma.¹

Case Presentation
A 48 year-old Caucasian man presented with a three day history of left sided headache, red eye, blurry vision and epiphora. He had a history of type II diabetes and hypertension. His current medications included aspirin, simvastatin, ramipril, glimepiride and metformin. He had no known allergies and no significant personal or family history of ocular disease. On examination, he had reduced visual acuity (VA) in the left eye, 6/12-1 compared to VA of 6/9 in the right eye. Examination of his left eye demonstrated mild circumlimbal injection, corneal oedema, and a deep and quiet anterior chamber with an unresponsive, semi-dilated pupil. Intraocular pressure (IOP) was 17 mmHg in the right and 52 mmHg in the left eye. Examination of his right eye was normal. Fundal examination was not possible due to corneal oedema. Gonioscopy revealed open angles in both eyes.
Investigations
Following slit lamp examination, the patient was also evaluated for systemic causes of red eye. Investigations carried out included routine blood tests and ESR, the results of which were all within normal limits. Once a diagnosis of Posner-Schlossman syndrome (PSS) was made, Humphrey visual field testing and optic disc colour photography was used to monitor disease progression.

Differential Diagnoses
The differential diagnoses include ocular hypertension, acute angle-closure glaucoma (AACG), primary open-angle glaucoma (POAG) and uveitic glaucoma, such as PSS and Fuchs’ heterochromic uveitis. Even though the symptoms and signs correlate with AACG, gonioscopy showed an open angle, which excluded this diagnosis. POAG is classically asymptomatic with bilateral high IOP, which was not the case here. Due to the mild inflammatory signs, a uveitic cause should be considered. Fuchs’ heterochromic uveitis was ruled out as a result of the absence of heterochromia and stellate keratic precipitates.

Outcome & Follow-up
Oral and intravenous Diamox (acetazolamide), G. Pred Forte 1% (prednisolone), G. Timoptol 0.25% (timolol), G. Iodipine 1% (apraclonidine) were administered in the first instant. However, the next pressure check revealed inadequate pressure control and therefore G. Xalatan (latanoprost) and intravenous Mannitol were given in addition. The following day the patient developed a more severe left circumlimbal injection, a mild degree of anterior chamber cells and flare. The left eye IOP was 30mmHg. Right eye examination was normal. After continuous treatment with the regimens mentioned above, his IOP was controlled at 19 mmHg but signs of anterior uveitis persisted. At this point in time, the working diagnosis was an inflammatory response, presumed to be trabeculitis, causing raised IOP, also known as Posner-Schlossman syndrome.

In the following 20 month period, the patient had three further episodes of unilateral IOP spikes associated with cyclitis. His left optic disc remained healthy with normal visual fields following these flare-ups. However, eighteen months later, he had yet another episode of high IOP (35mmHg left eye) with circumlimbal injection and on this occasion the left optic disc showed subtle increased cupping with nasal...
step changes on visual field testing. He was treated with further IOP-lowering and steroid drops. Two months later, IOP in the left eye was still 24mmHg with a slightly thinned neuro-retinal rim inferiorly. Since then, the IOP in the left eye has remained in the low twenties despite different combinations of IOP-lowering drops. His left optic disc and visual field have shown glaucoma-related changes (Figure 1 and Figure 2); therefore, surgical treatment with trabeculectomy was considered.

Discussion

Posner-Schlossman syndrome (PSS) or glaucomatocyclitic crisis is a rare unilateral recurrent ocular hypertensive disease which is associated with mild non-granulomatous cyclitis. The patient described here fits into the typical age group as it usually affects patients between the second and fourth decade, with a male predominance. The classic feature of PSS is that the high IOP does not correlate with the severity of inflammation. Patients usually present with unilateral mild ocular discomfort, blurred vision, mild cell/flare in the anterior chamber, corneal oedema and high IOP. Sometimes, non-pigmented keratic precipitates (KPs) accumulate in the lower third of the cornea. Gonioscopy shows open angles with an absence of abnormal pigmentation. Most of these symptoms and signs were present in this case, except the presence of KPs. Because of the mild nature of inflammatory features of this disease, making a diagnosis of PSS can be very challenging.

The aetiology and pathophysiology of PSS remain unclear. The acute pressure rise in PSS is thought to be related to trabeculitis and subsequent trabecular meshwork congestion by monocellular cell infiltration. Several aetiologies have been postulated, including an abnormal vascular process, immune mechanism, autonomic defects, allergic conditions, variation of developmental glaucoma, cytomegalovirus and herpes simplex virus infections, aqueous hypersecretion, and immunoglobulin E–mediated reaction.

Our patient had recurrent episodes of high intraocular pressure with associated cyclitis which required medical treatment to reduce the IOP and control inflammation. The treatment of PSS includes IOP-lowering topical agents such as beta-blockers, alpha-2-agonists, prostaglandin analogues, and carbonic anhydrase inhibitors. Systemic treatment such as carbonic anhydrase inhibitor and osmotic agents are also used to lower IOP if topical treatment is insufficient. Most authors also recommend topical steroid drops such as prednisolone drops, systemic anti-inflammatory agents and topical NSAIDs to control the inflammation. Weaker cycloplegic drops have also been suggested to prevent formation of posterior synechiae. Topical miotic agents such as pilocarpine are contraindicated since they can cause ciliary spasm and lead to formation of peripheral anterior and posterior synechiae. In steroid-responders it is important not to sacrifice the control of inflammation solely to prevent an IOP rise.

PSS was originally described as a benign disease with normal optic disc and visual fields. Later, studies have shown that there is an association between POAG and PSS. One study showed that 45% of PSS patients had concomitant POAG. Besides this association, some studies have also shown that patients with PSS can develop secondary
chronic open angle glaucoma. In a study of 53 cases of PSS, 26.4% of the patients developed glaucomatous changes. After 3.5 years of recurrent PSS attacks, our patient similarly developed a glaucomatous optic disc and visual field changes. Because of this risk of progression to chronic open angle glaucoma, patients with PSS have to be followed-up and monitored closely. Surgical treatment, such as trabeculectomy, may be necessary to prevent further glaucomatous damage if topical treatment is insufficient. Some authors postulate that trabeculectomy with antimetabolites might actually prevent IOP spikes and reduce the severity and recurrence rate of uveitis. Studies by Stravou and colleagues and Jap et al found that in 72%-80% of eyes that had trabeculectomy with uveitis and high IOP, the course of disease actually improved.12,13

Acknowledgements
The authors wish to thank Mr John Suharwardy and the staff in the ophthalmology department at the Royal Oldham Hospital for their kind help and support in writing this report.

References

LEARNING POINTS
- PSS is a rare cause of intermittent spikes in unilateral intraocular pressure due to cyclitis and trabeculitis, commonly presenting with ocular discomfort, blurred vision, mild flare in the anterior chamber, corneal oedema, with open angles seen on gonioscopy.
- Due to the risk of progression to secondary chronic open angle glaucoma, patients with PSS must be monitored closely.
- Trabeculectomy should be considered early in the management of PSS with persistently high IOP in order to control IOP and prevent glaucomatous changes.