Clinical & Refractive Optometry CRO
ONLINE CE A CE JOURNAL FOR OPTOMETRY

CE Credit - Case Report(s) & Topic Review

Posner Schlossman's Syndrome (PSS): A Case Refractory to Medical Treatment with Glaucoma Progression

Eulogio Besada OD, FAAO1, Barry J. Frauens OD, FAAO[1]

CRO (Clinical & Refractive Optometry) Journal

ABSTRACT

Introduction: A case of unattended Posner Schlossman's syndrome (PSS) and its conversion to glaucoma is reviewed. It illustrates the risk of progression and becoming refractory to prevailing medical treatment. It debates whether initial surgical intervention should be considered due to minor symptoms during recurrences and poor patient compliance.

Case Presentation: A 52-year-old male presents with blurry vision and minor headache discomfort. Records from the last eye exam 6 years prior indicated a diagnosis of ocular hypertension OD. No symptoms other than blurry vision were reported. Intra-ocular pressures (IOP) were 32 OD, 20 OS mm Hg, with cup disc (C/D) ratios of 0.35 with healthy rim tissue. No presence of anterior chamber cells, nor a relative afferent pupillary defect (RAPD) was documented. He was scheduled for a glaucoma workup but did not return for follow-up. He returned 6 years later. Corrected visual acuities were 20/20 OD, OS. A RAPD OD, IOP of 35 OD and 17 OS mm Hg, C/D ratios of 0.8 glaucomatous cupping OD, 0.45 with healthy rim tissue OS and less than grade 1 anterior chamber cells (AC) were noted. Gonioscopy revealed open angles to the ciliary body OU. Optical coherence tomography and Visual field revealed glaucomatous damage OD. A diagnosis of PSS-related glaucoma OD was specified, and prednisolone acetate 1% qid OD was prescribed. Subsequently, dorzolamide and timolol bid OD were added. The elevated IOP did not respond to treatment. He was referred for surgery as PSS was considered refractory.

Conclusion: PSS may be challenging to diagnose and manage since AC cells may be difficult to detect, and patients may have minimal symptoms during recurrences. With poor compliance, it may become refractory and progress to glaucoma. Patient education and the importance of strict compliance should be stressed. Primary surgical intervention should be considered in cases of regular recurrences and poor compliance.

INTRODUCTION

Posner Schlossman's syndrome (PSS) also known as glaucomatocyclitic crisis is generally characterized by unilateral elevation of intra-ocular pressures (IOP) that may increase to 40 mm Hg or higher, presence of minimal symptoms during recurrences, and mild inflammation associated with presence of anterior chamber (AC) cells.[1-4] Although there is significant understanding of PSS and its management, it is probable that some cases will be initially misdiagnosed or unattended due to the presence of minimal symptoms during recurrences and patient neglect. In some instances, PSS may become refractory and progress to secondary glaucoma. This article discusses such a patient's case.

CASE REPORT

A 52-year-old male presented with complaints of blurry vision OD and minor nonspecific headache. At an initial examination 6 years earlier, the patient reported no symptoms other than uncorrected blurry vision. Best corrected visual acuities (BVA) were 20/20 OD, OS. During this visit IOP was recorded to be 32 mm Hg, 20 mm Hg OD/OS respectively. C/D ratios were 0.35 with healthy rim tissue OU. Presence of AC cells was not noted in the right eye. Gonioscopy was documented to be open to ciliary body (CB) 360 degrees without evidence of angle recession OU. The patient was diagnosed with ocular hypertension OD and recommended to return to clinic in 2-3 days in order to undergo additional testing including VF, OCT and to repeat IOP measurement. The patient did not comply with the recommended visit but returned to clinic 6 years later complaining of blurry vision OD and a minor nonspecific headache of 1 month onset. BVA were 20/20-2 OD 20/20 OS. VF were full to confrontation finger counting OU however a 2+ RAPD was present OD. IOP's were 35 mm Hg, 17 mm Hg OD, OS respectively. Scrutinized examination of the AC revealed less than grade 1 cell OD

[1]*Professor, Nova-Southeastern University College of Optometry, Florida, USA*

only. Dilated fundus examination revealed C/D ratios of 0.8 with inferior/superior rim thinning OD, and 0.45 OS with healthy rim tissue (Figure1 a-b). Gonioscopy was open to ciliary body (CB) 360 degrees without evidence of angle recession OU. Iris atrophy, or corneal fine keratic precipitates were not present OU. Corneal pachymetry was 521 um and 532 um OD/OS respectively. Subsequent OCT (Figure 2) and VF (Figure 3a-b) confirmed glaucomatous damage OD. A diagnosis of secondary glaucoma, likely the result of PSS OD was made. Prednisolone acetate 1% drops 4 times a day was prescribed OD. On follow up visits, no significant reduction in IOP OD occurred, subsequently dorzolamide/timolol bid was added. During a period of observation with the combined treatment the IOP could not be significantly lowered and when brimonidine was added the patient reported he did not tolerate it. At the last visit 3 months later the IOP was 41 and 22 mm Hg respectively. The patient's condition was determined to be refractory to medical treatment and a referral to a glaucoma specialist was issued to consider surgical management option.

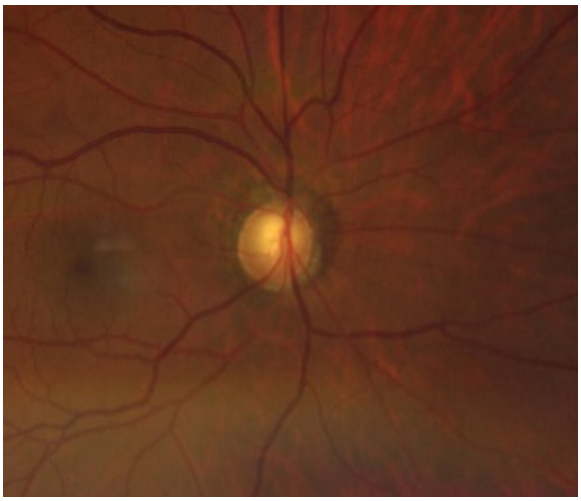

Figure 1a. Optic nerve photos OD. Evident cupping with cup/disc ratios of 0.8 with inferior/superior rim thinning.

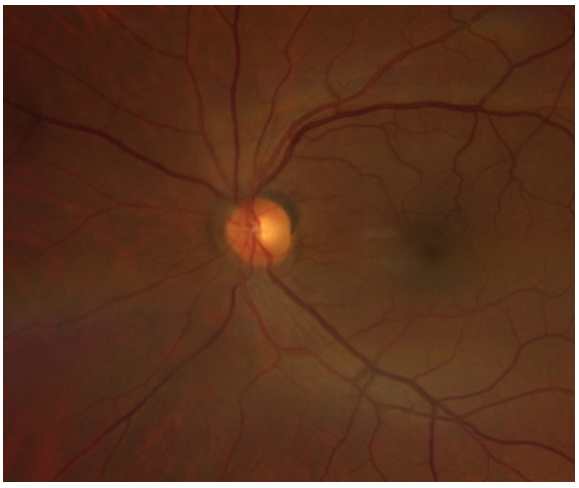

Figure 1b. Optic nerve photos OS. Observed cup disc ratio of 0.45 OS with healthy rim tissue.

Summary Parameters

RNFL Analysis	OD	OS	Inter Eye (OD-OS)
Average RNFL (µm)	57	93	-36
Superior RNFL (µm)	54	100	-46
Inferior RNFL (µm)	60	86	-26
Intra Eye (S-I) (µm)	-6	14	N/A

ONH Analysis	OD	OS	Inter Eye (OD-OS)
Cup/Disc Area Ratio	0.80	0.31	0.49
Cup/Disc V. Ratio	0.86	0.57	0.29
Cup/Disc H. Ratio	0.97	0.55	0.42
Rim Area (mm^2)	0.40	1.17	-0.77
Disc Area (mm^2)	1.98	1.70	0.28
Cup Volume (mm^3)	0.578	0.075	0.503

GCC Analysis	OD	OS	Inter Eye (OD-OS)
Average GCC (µm)	67	90	-23
Superior GCC (µm)	67	90	-23
Inferior GCC (µm)	66	89	-23
Intra Eye (S-I) (µm)	1	1	N/A
FLV (%)	3.62	0.07	3.55
GLV (%)	28.76	5.14	23.62

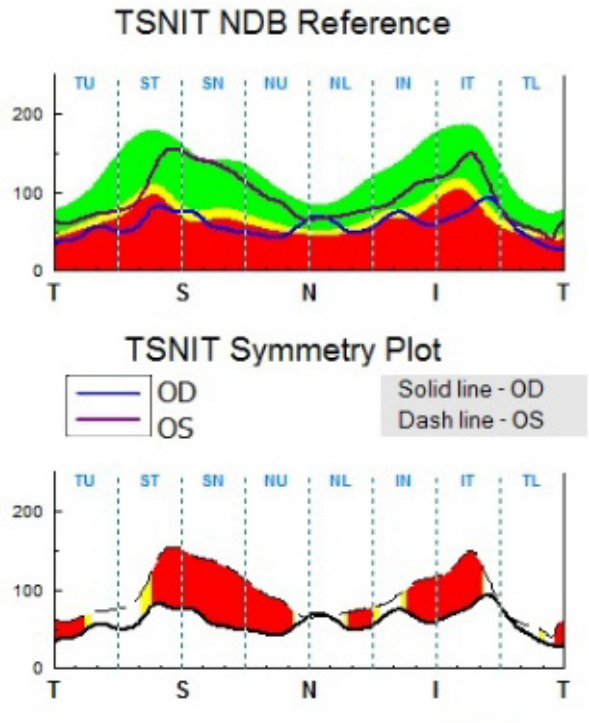

Figure 2. Optical coherence tomography RTVue confirmed glaucomatous damage OD.

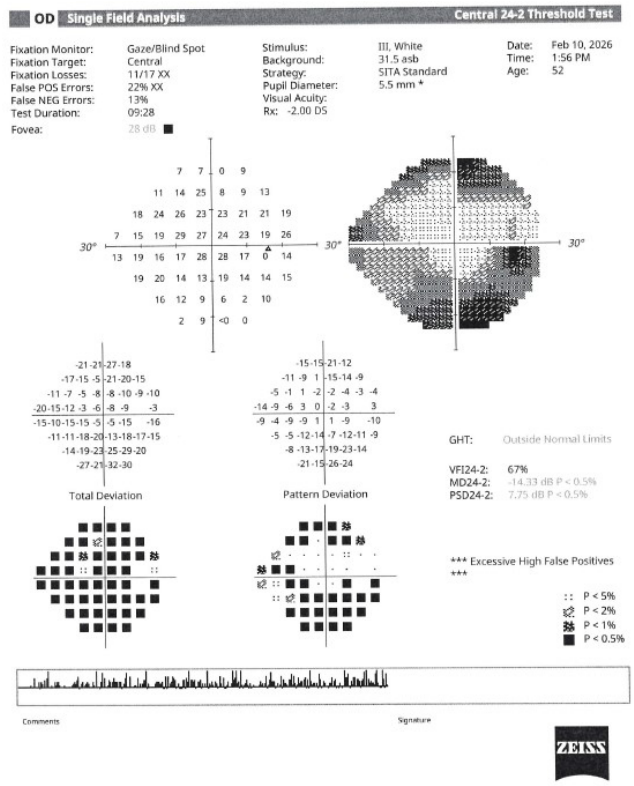

Figure 3a. Humphrey's Visual Field demonstrates glaucomatous damage OD. Fixation losses, significant. Glaucomatous loss nevertheless corresponded to optic nerve appearance and OCT OD.

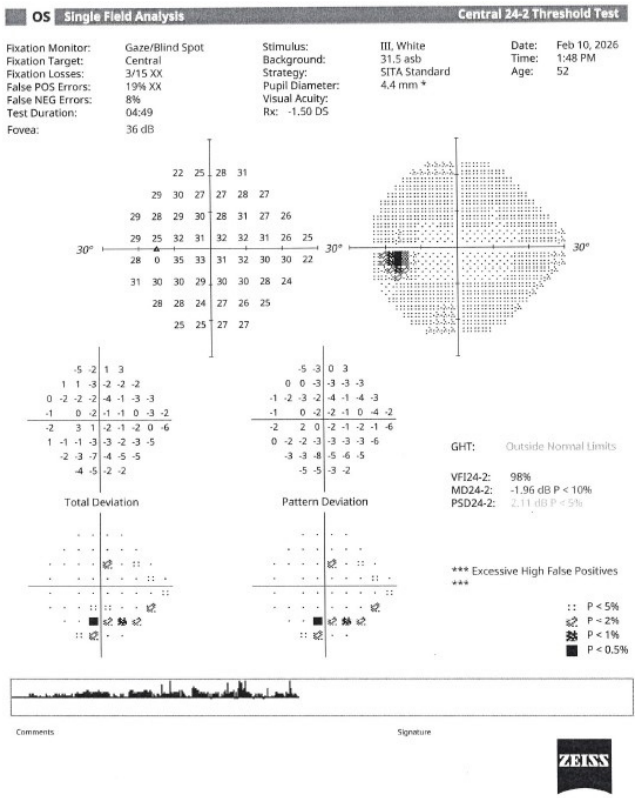

Figure 3b. Humphrey's Visual Field OS. Inferior peripheral defects (non-repeatable) considered to be artifacts. Fixation losses significant. OU. Nonetheless not corresponding to optic nerve appearance or OCT OS.

DISCUSSION

This case was not characteristic of the possible differential diagnoses of PSS. AC inflammation was minimal and unlike in uveitis glaucoma, the cornea was not affected as in herpetic kerato-uveitis, iris atrophy was not present as in Fuch's Heterochromic Iridocyclitis, and angles were open to CB 360 degrees.[4] This case exemplifies the risk of progression to secondary glaucoma due to PSS. Progressive optic nerve damage and ultimately secondary glaucoma have been documented to occur between 5 to 10 years from initial presentation.[1,5,6] In some cases of PSS, it may be challenging to observe cells in the anterior chamber. In a case with unilateral elevated IOP and no optic nerve cupping, a presumptive diagnosis of ocular hypertension (OHT) may be reached. Patient education regarding risk of development of glaucoma and requirement of follow up evaluations in both OHT and PSS is of utmost importance. In PSS, it must be explained that although the inflammatory cause of elevated IOP may be resolved with temporary medical management, recurrences with non-perceptible symptoms may occur. If not attended properly, these cases may advance to glaucoma. In this case during the initial examination, the IOP was elevated OD, and angles were open to CB OU. However, presence of cells were not documented in the right eye. Although the patient was scheduled for a follow-up appointment for additional tests and management, he did not comply. Upon his return to the clinic 6 years later, he developed glaucoma OD. We speculate that although not documented, AC cells were probably present at the initial visit with elevated IOP. During the interim period recurrences with minimal symptoms possibly lead to periods of sustained IOP. Since the patient did not respond to medical treatment, it is also likely that chronic TM damage would have resulted in persistently elevated IOP.

Human leucocyte antigen (HLA) haplotypes, presence of prostaglandin E and autotaxin (ATX) has been suggested to be associated in the development of TM changes in PSS.[1,4,7-9] Herpes simplex family virus and specifically CMV have been documented to be associated with development of PSS.[4,7,10-13] In PSS, patients that tested positive to CMV are most likely to become refractory to medical treatment and need surgery compared to CMV negative patients.[7,14] High viral load numbers in CMV positive PSS individuals have been documented to provide a significant risk of IOP elevation that becomes refractory to medical treatment.[4,7,14-17] Surgery may be needed in refractory PSS cases with progressive glaucoma.[3,4] TM and Schlemm's canal (SC) impairment is thought to contribute to elevated IOP in CMV positive PSS patients.[7] This dysfunction may result from aqueous humor outflow restrictions due to development of thickness and edema in the TM tissue.[1,7,18] Xiaoquin and associates reported that TM edema was reduced following anti-inflammatory treatment in a cohort subgroup. They indicated that this may explain why steroids with or without IOP lowering medications may reduce IOP in some PSS patients.[7]

CMV upregulation of ATX has also been described to lead to fibrotic changes in the TM and reduce permeability in SC in PSS patients. In vitro analysis of CMV infected human TM cells identified upregulated expression of ATX and TGF-ß1 genes that resulted in significant fibrosis of TM. This TM fibrosis was reduced using Rock inhibitors. Levels of ATX in the AC similarly have shown to correlate with IOP.7 It has been suggested that extracellular material deposition and fibrosis in TM cells seem to be conducted via the Rho/Rock pathway. This affects the biomechanical contractile quality of the TM and SC and leads to IOP elevation.[7,19,20] Similar positive correlation has been described to occur between the aqueous humor levels of ATX and elevated IOP. It has been suggested that in CMV induced ATX upregulation produces a cascade of biochemical pathways that may cause sustained and refractive IOP elevation in PSS positive patients. Since Rock inhibitors can help modulate cellular adhesion, relocation and contraction of smooth muscle, it has been suggested as potential treatment in refractory IOP elevation in CMV positive PSS patients[7,21]

In this case, CMV titers were not obtained, and consequently its relation to the PSS could not be determined in this patient. As a result of being refractory to medical treatment, and the advanced stage of glaucoma in the affected eye, a surgical consult was considered more appropriate before commencing Rock inhibitor management. A topic of further discussion is whether initial surgical intervention should be contemplated as adequate prophylactic treatment in cases of poor patient compliance.

REFERENCES

1. Yan X, Li M, Wang J, et al. Morphology of the trabecular meshwork and Schlemm's canal in Posner-Schlossman syndrome. *Invest Ophthalmol Vis Sci*.2022;63(1):1-6.
2. Posner A, Schlossman A. Syndrome of unilateral recurrent attacks of glaucoma with cyclitic symptoms. *Arch Ophthal*. 1948;39(4):517–35.
3. Posner A, Schlossman A. Further observations on the syndrome of glaucomatocyclitic crises. *Trans Am Acad Ophthalmol Otolaryngol*. 1953;57(4):531–6.
4. Gottehrer J. Posner-Schlossman syndrome. *J Med Optom*.2024;2(3):1-9.
5. Jap A, Sivakumar M, Chee SP. Is Posner Schlossman syndrome benign? *Ophthalmology*.2001; 108(5):913–8.
6. Su CC, Hu FR, Wang TH, et al. Clinical outcomes in cytomegalovirus-positive Posner-Schlossman syndrome patients treated with topical ganciclovir therapy. *Am J Ophthalmol*.2014; 158(5):1024–31.
7. Igarashi N, Honjo M, Yamagishi R, et al. Involvement of autotaxin in the pathophysiology of elevated intraocular pressure in Posner-Schlossman syndrome. *Sci Rep*.2020;10(1):6265.
8. Hirose S, Ohno S, Matsuda H. HLA-Bw54 and glaucomatocyclitic crisis. *Arch Ophthalmol*. 1985;103(12):1837–9.
9. Hess LK, Lee GA, Shah P. Bilateral simultaneous presentation of Posner-Schlossman syndrome. *Clin Exp Ophthalmol*.2017;45(9):925-7.
10. Chee SP, Jap A. Presumed fuchs heterochromic iridocyclitis and Posner-Schlossman syndrome: comparison of cytomegalovirus-positive and negative eyes. *Am J Ophthalmol*.2008;146(6):883–9.
11. Shazly TA, Aljajeh M. Latina MA. Posner-Schlossman glaucomatocyclitic crisis. *Semin Ophthalmol*.2011;26(4–5):282–4.
12. Cao G, Tan C, Zhang Y, et al. Digital droplet polymerase chain reaction analysis of common viruses in the aqueous humour of patients with Posner-Schlossman syndrome in Chinese population. *Clin Exp Ophthalmol*.2019;47(4):513-20.
13. Rodier-Bonifas C, Cornut PL, Billaud G, et al. Cytomegalovirus research using polymerase chain reaction in Posner-Schlossman syndrome. *J Fr Ophtalmol*.2011;34(1):24-9.
14. Su C, Hu FR, Wang TH, et al. Clinical outcomes in cytomegalovirus-positive Posner-Schlossman syndrome patients treated with topical ganciclovir therapy. *Am J Ophthalmol*.2014;158(5):1024–31.
15. Accorinti M, Gilardi M, Pirraglia MP, et al. Cytomegalovirus anterior uveitis: long-term follow-up of immunocompetent patients. *Graefes Arch Clin Exp Ophthalmol*.2014;252(11): 1817–24.
16. Zhong Y, Cheng Y, Liu X, Feng P. Trabeculectomy in the management of glaucomatocyclitic crisis with visual field defect. *Ocul Immunol Inflamm*. 2010 Jun;18(3):233-6.
17. Darchuk V, Sampaolesi J, Mato L, et al. Optic nerve head behavior in Posner-Schlossman syndrome. *Int Ophthalmol*. 2001;23:373–9.
18. Megaw R, Agarwal PK. Posner-Schlossman syndrome. *Surv Ophthalmol*. 2017;62(3):277–85.
19. Torrejon K Y, Papke EL, Halman JR, et al. TGFß2-induced outflow alterations in a bioengineered trabecular meshwork are offset by a rho-associated kinase inhibitor. *Sci Rep*. 2016;6(10):1038.
20. Vranka JA, Kelley MJ, Acott TS, Keller KE. Extracellular matrix in the trabecular meshwork: intraocular pressure regulation and dysregulation in glaucoma. *Exp Eye Res*. 2015;133: 112–25.
21. Tan JK, Khaw PT, Henein C. Rho Kinase (ROCK) inhibitors in the treatment of glaucoma and glaucoma surgery: A systematic review of early to late phase clinical trials. *Pharmaceuticals*.2025;18(4):523.