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A CASE OF ATYPICAL TOLOSA–HUNT SYNDROME WITH ISOLATED THIRD NERVE PALSY AND ANGLE CLOSURE

Ophthalomology

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ABSTRACT

Tolosa–Hunt Syndrome (THS) is an uncommon granulomatous inflammatory disorder of the cavernous sinus and orbital apex, classically presenting with unilateral pain and ophthalmoplegia. Diagnosis may be challenging when neuroimaging is normal, especially in early or benign variants. Coexistence of THS with acute angle-closure glaucoma is exceedingly rare and can obscure the underlying pathology due to overlapping symptoms of ocular pain, pupillary dysfunction, and motility impairment. We report a 50-year-old woman who presented with recurrent headache, right eye pain, ptosis, and markedly elevated intraocular pressure, initially suggestive of acute angle-closure glaucoma. The presence of a pupil-involving third nerve palsy and dramatic steroid responsiveness supported benign THS despite normal MRI findings. Persistent narrow angles required bilateral Nd:YAG iridotomy. The patient showed complete neurological recovery and stable intraocular pressures on follow-up. This case highlights a rare dual presentation and emphasizes the need for prompt recognition, multidisciplinary evaluation, and early intervention to prevent visual morbidity.

KEYWORDS

Tolosa–Hunt Syndrome; Painful Ophthalmoplegia; Third Nerve Palsy; Acute Angle-Closure Glaucoma; Neuro-Ophthalmology; Steroid-Responsive Neuropathy

INTRODUCTION

Tolosa–Hunt Syndrome (THS) is an uncommon inflammatory disorder involving the cavernous sinus and orbital apex, producing unilateral orbital pain with ipsilateral ocular motor nerve dysfunction. Although MRI abnormalities are considered part of the diagnostic criteria, several studies describe early or benign forms of THS that present with normal imaging, complicating recognition. The syndrome is well known for its dramatic response to corticosteroids, a feature often relied upon when neuroimaging is inconclusive.

Acute angle-closure glaucoma is a distinct ophthalmic emergency characterized by sudden elevation of intraocular pressure (IOP), corneal edema, a fixed dilated pupil, and severe ocular pain. The simultaneous occurrence of THS and angle closure is extremely rare, and the overlapping symptoms of pain, pupillary dysfunction, and neurological deficits can delay identification of the underlying neuro-ophthalmic pathology.

The purpose of reporting this case is to highlight an unusual clinical scenario in which benign THS masqueraded as acute angle-closure glaucoma, emphasizing the importance of maintaining diagnostic vigilance when neurological and glaucomatous features coexist.

Case Report

A 50-year-old woman presented with a one-month history of intermittent right-sided headaches, followed by eight days of progressive right eye pain and drooping of the upper eyelid. She denied systemic illness, prior ocular trauma, or previous ocular interventions.

On examination, right eye visual acuity was counting fingers at 3 meters. The eyelid showed complete ptosis. The pupil was mid-dilated and unreactive to both direct and consensual light. The cornea demonstrated microcystic epithelial edema, and the conjunctiva was congested. The anterior chamber was shallow (Van Herick Grade 1), and posterior synechiae extended from 6 to 9 o'clock. Gonioscopy revealed Shaffer Grade 0, indicating complete angle closure. Extraocular movements were markedly restricted in adduction, elevation, and depression, with the eye resting in a "down-and-out" position, consistent with a pupil-involving third nerve palsy. Intraocular pressure was 44 mmHg. Fundus examination was hazy; however, a 0.45 CDR with a healthy neuroretinal rim was noted.

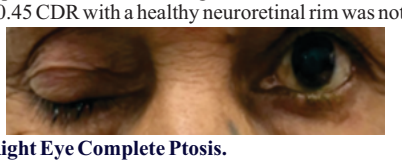

Figure 1: Right Eye Complete Ptosis.

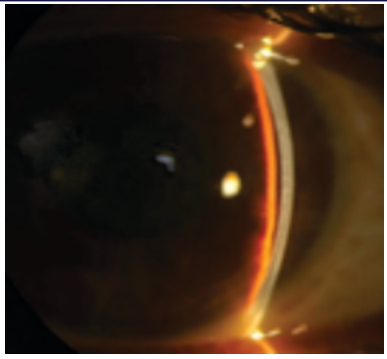

Figure 2: RE Anterior Chamber Van Herick Grade 1

The left eye had 6/6 vision, a shallow anterior chamber (Van Herick Grade 2), an immature cataract, Shaffer Grade 1 on gonioscopy, and an intraocular pressure of 24 mmHg. Ocular motility was full and painless. Fundus examination revealed a 0.4 CDR with a healthy neuroretinal rim and a positive foveal reflex.

A provisional diagnosis of right eye acute angle-closure glaucoma with concurrent pupil-involving third nerve palsy and left eye primary angle-closure suspect was made. The patient was treated with oral acetazolamide 250 mg twice daily, topical timolol–brimonidine, and topical fluorometholone. MRI brain and orbit with contrast revealed no cavernous sinus abnormality. Laboratory evaluation showed elevated ESR with otherwise normal parameters. Intravenous methyl-prednisolone 500 mg daily was administered for five days, followed by an oral taper.

By Day 10, right eye vision improved to 6/6, corneal clarity was restored, extraocular movements normalized, and IOP decreased to 11 mmHg. The pupil became sluggishly reactive. Despite neurological recovery, angles remained occludable. The fellow eye IOP also stabilized at 11 mmHg.

Pilocarpine was initiated in both eyes. On Day 12, with IOP measuring 8 mmHg in the right eye and 12 mmHg in the left, bilateral Nd:YAG peripheral iridotomy was performed. Brinzolamide was started in the right eye.

Subsequent follow-up demonstrated stable IOP between 12–14 mmHg, patent iridotomies, and healthy optic discs. Early posterior

subcapsular cataract developed in the right eye. Based on clinical features, exclusion of differentials, and steroid responsiveness, a final diagnosis of Benign Tolosa–Hunt Syndrome with concurrent acute angle-closure glaucoma in the right eye and primary angle closure suspect in the left eye was established.

DISCUSSION

THS is classically characterized by unilateral orbital pain and ophthalmoplegia due to involvement of cranial nerves III, IV, or VI. Although MRI often demonstrates granulomatous inflammation within the cavernous sinus, up to one-third of patients may have normal imaging, particularly in early stages. With increasing recognition of alternative causes of painful ophthalmoplegia, including neoplastic, infectious, and systemic inflammatory conditions, THS remains a diagnosis of exclusion. The rapid and marked clinical improvement following corticosteroid therapy strongly supports the diagnosis of benign THS in such cases.

Isolated cranial nerve involvement in THS is uncommon, as multiple cranial nerves lie in close proximity within the cavernous sinus. However, rare cases of single nerve involvement have been reported. A case by Alharbi et al. described an isolated, pupil-involving third nerve palsy as the presenting feature of THS, mimicking compressive pathology, with complete resolution following steroid therapy . These findings suggest that early or limited variants of THS may present with isolated cranial neuropathy, increasing diagnostic difficulty.

The coexistence of THS and acute angle-closure glaucoma is extremely rare. Neurogenic mydriasis due to third nerve palsy can precipitate pupillary block in anatomically narrow angles. In this patient, pre-existing narrow angles likely predisposed to acute angle closure triggered by parasympathetic dysfunction, resulting in elevated IOP and corneal edema.

Differential diagnoses of painful ophthalmoplegia include posterior communicating artery aneurysm, cavernous sinus thrombosis, invasive fungal sinusitis, idiopathic orbital inflammation, sarcoidosis, tuberculosis, and carotid–cavernous fistula. These were excluded based on imaging and systemic evaluation.

Management requires addressing both inflammatory neuropathy and angle closure. Corticosteroids provide rapid relief of inflammation, while Nd:YAG iridotomy prevents recurrent angle closure. The favorable outcome in this case highlights the importance of a multidisciplinary approach.

CONCLUSION

This case illustrates a rare presentation of benign Tolosa–Hunt Syndrome mimicking acute angle-closure glaucoma. A fixed dilated pupil with ophthalmoplegia warrants urgent neuroimaging; however, normal MRI does not exclude THS. Early corticosteroid therapy and definitive angle management are essential to prevent irreversible visual loss.

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