Glaucomatocyclitic Crisis in a Nigerian Child: A Case Report

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Abstract

Background: Glaucomatocyclitic crisis is a condition that elicits significant elevated intraocular pressure (IOP) with minimal non-granulomatous anterior uveitis. It is usually unilateral, occasionally recurrent, and easily treatable in most cases. It is seen mainly in young adults but is rarely reported in children. This is to report an uncommon case of glaucomatous cyclitic crisis (Posner-Schlossman syndrome) in a Nigerian child. Case report: A 15-year-old boy presented with pain, haloes around light, photophobia, and decreased vision in the left eye over the course of the day. He has a similar history from 5 years ago, with a brief resolution on steroid eye drops. Ocular examination in the left eye revealed minimal non-granulomatous anterior uveitis with significantly elevated intraocular pressure. He was treated with topical steroid and ocular hypotensive and his IOP was 14 mmHg at 14 hours, down from 44 mmHg at the start. The anterior chamber was quiet by the fourth day of treatment. The clinical picture in this child was consistent with Posner-Schlossman syndrome. Following resolution of the inflammation and discontinuation of all medications, the patient is being followed up as a glaucoma suspect because of the subtle disc finding and the slight risk of glaucoma development later. Conclusion: Although glaucomatocyclitic crisis is uncommon in children, it should be considered in the differential diagnosis of uveitis and glaucoma in paediatric patients.

Keywords: Glaucomatocyclitic crisis, child, elevated intraocular pressure, uveitis

Introduction

In 1948, Posner and Schlossman[1] described glaucomatocyclitic crisis as a condition characterised by recurrent, acute attacks of mild, unilateral, anterior uveitis associated with markedly elevated intraocular pressure. Since this first elaborate description, glaucomatocyclitic crisis has been reported almost exclusively in middle-aged and young adults.[2]

Case Report

We report a 15-year-old Nigerian boy with pain, haloes around light, photophobia, and decreased vision in the left eye for a day's duration. The pain was mild, with no nausea or vomiting. He had similar symptoms 5 years ago for which he was treated with topical corticosteroids, and symptoms and resolved within a few days. There was no known family history of glaucoma. There has been no history of trauma or surgery in the left eye.

Visual acuity unaided was 6/5 and 6/9 in the right and left eyes, respectively, and 6/6 with a pinhole in the left eye. Extraocular muscle movement was normal in both eyes. Biomicroscopy showed a normal examination in the right eye. In the left eye, biomicroscopy revealed a mild ciliary flush, a clear cornea, grade 0.5+ anterior chamber cells (1–5 cells per field), and a clear lens. Heterochromia was not present. Goldmann applanation tonometry showed 12 mm Hg in the right eye and 42 mm Hg in the left eye. Gonioscopy revealed open angles in both eyes with no evidence of angle recession, blotchy pigmentation, or synechiae. The pupil diameter was 4 mm in both eyes, they were reactive to light, and there was no relative afferent pupillary defect. On fundoscopy [Figure 1], the right eye revealed a slightly large-sized optic nerve head with a healthy neuroretinal rim and a cup-to-disc ratio (CDR) of 0.35×0.4, normal vessels, and normal macula. The left eye revealed a moderate optic nerve head with a CDR of 0.5×0.4 and a normal macula. Optical coherence tomography (OCT) right eye revealed a

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large disc with moderate cup and normal neuroretina rims and retinal nerve fibre layers, while the left eye revealed a moderate-sized disc with evidence of cupping and neuroretina rim loss as shown by the nasal rim being thicker than other rims [Figure 2]. The central visual field was essentially normal in the right eye, but the left eye showed a generalised reduction in sensitivity [Figure 3].

The clinical picture in this child was definitely in keeping with a diagnosis of Posner-Schlossman syndrome, with the appearance of some early optic nerve damage in the left eye. The patient was given oral acetazolamide 250 mg bd, topical dorzolamide 2%/timolol maleate 0.5% combination bd, and topical dexamethasone 0.1% qds to his left eye immediately. His intraocular pressure was reduced to 14 mmHg on re-check 14 hours later in the left eye, and his unaided visual acuity had improved to 6/6. His medication was reviewed, and the oral acetazolamide was discontinued.

On the fourth day after the presentation, a follow-up examination of the left eye showed unaided visual acuity of 6/5, intraocular pressure of 12 mmHg with no flares nor cells in the anterior chamber in both eyes. The corticosteroid was tapered over 2-week periods: tds for a week, bd for 3 days, and daily for 3 days. The topical dorzolamide 2% and timolol maleate 0.5% combination was also discontinued after 2 weeks. He has been reviewed 8-weeks after discontinuation of medications with stable findings and an IOP of 12 mmHg in the left eye.

Discussion

The glaucomatocyclitic crisis is characterized by recurrent, acute attacks of mild, unilateral, non-granulomatous, anterior uveitis accompanied by markedly elevated intraocular pressure.[1] Patients typically present with a mild to moderate loss of vision (range, 6/6 to 6/60)[1,3] with pain lasting hours to days.[3] The pupil is often slightly dilated or normal.[3,4] The conjunctiva in most cases is white and quiet, however, a mild ciliary flush may be present. Corneal oedema and a few discrete endothelial keratic precipitates (KPs), usually in an inferior distribution, may be observed.[3,4] The KPs usually resolve spontaneously or with anti-inflammatory treatment. The anterior chamber is deep, with mild iritis and no significant cell or flare. Iris atrophy and heterochromia have been reported in some cases, but they are not considered characteristic findings for PSS.[4] Notably, peripheral anterior synechiae are absent, and the angles are invariably open.[1-3] Intraocular pressure typically increases above 40 mmHg[5] and some have reported glaucomatous cupping[1,6] as well as transient or permanent visual field changes.[2]

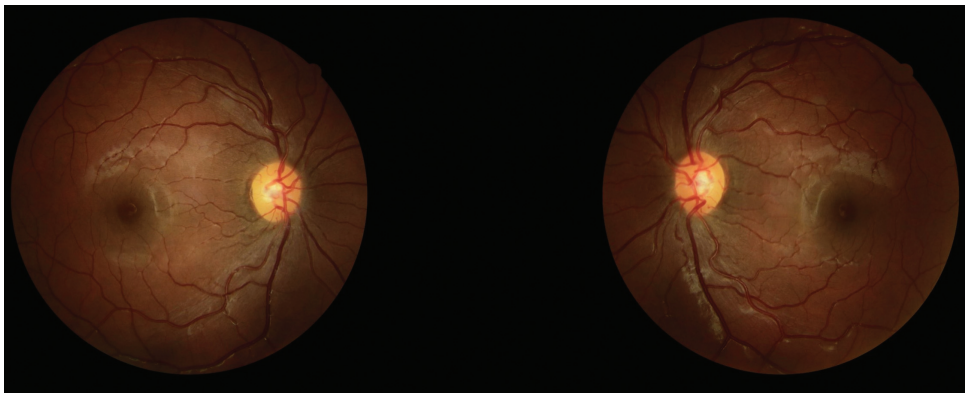

Figure 1: Optic Disc of the right and left eyes.

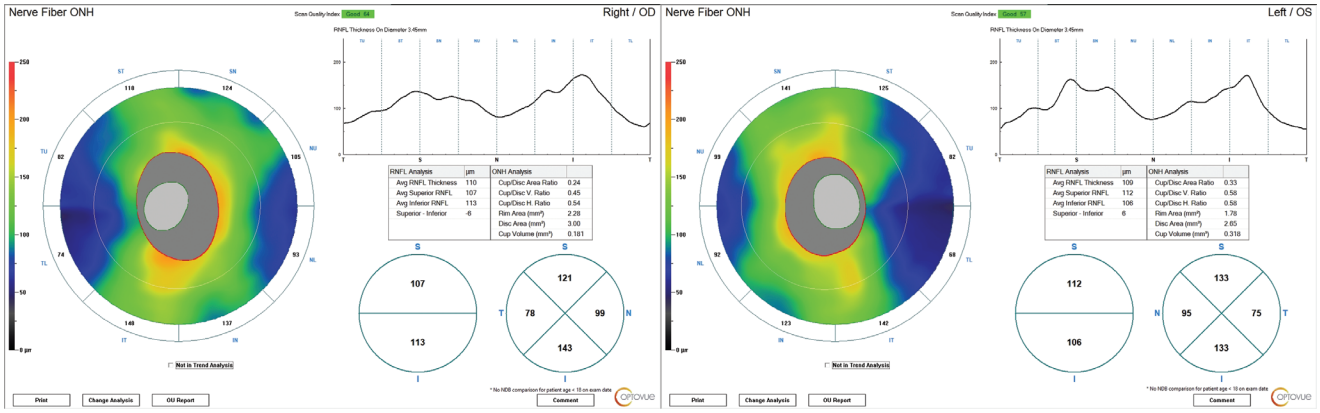

Figure 2: Optical coherence tomography of the right and left eyes.

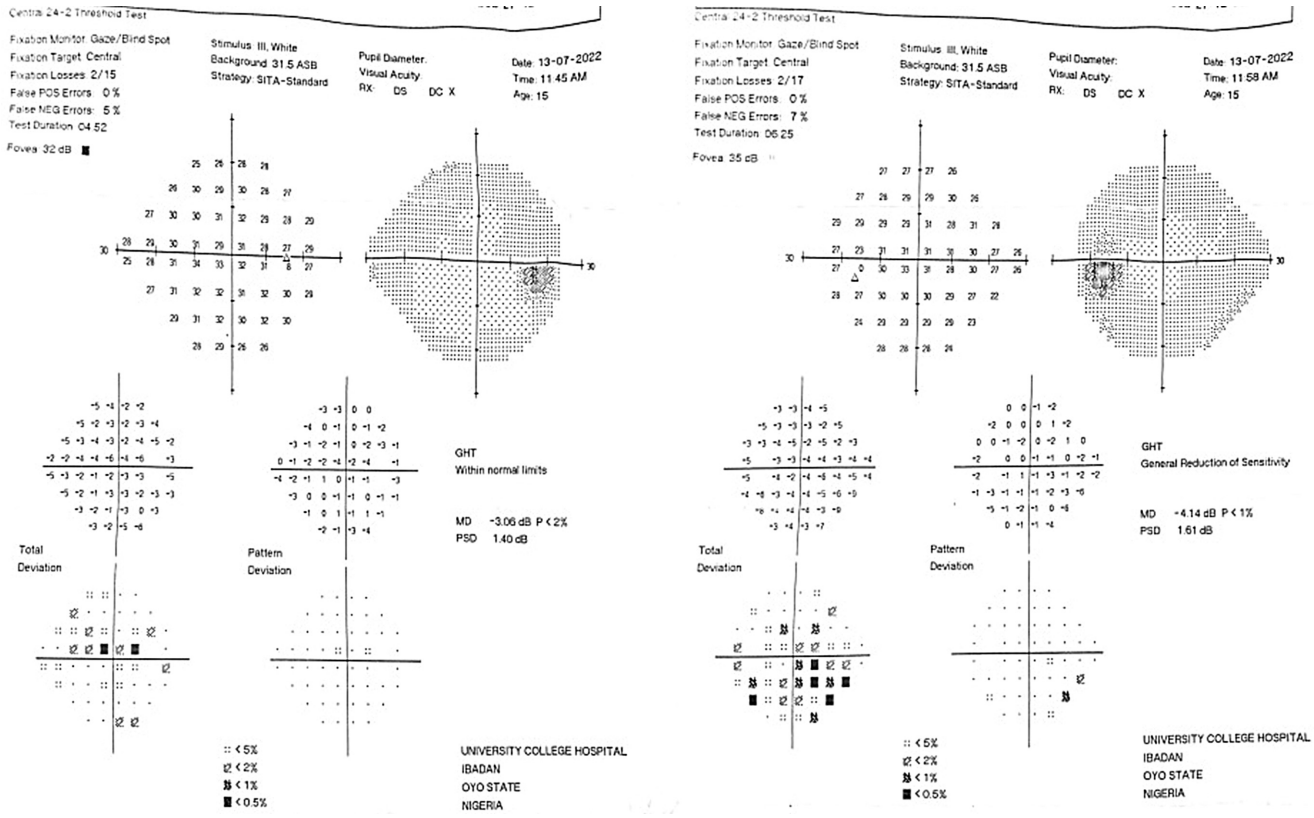

Figure 3: Central visual fields of the the right and left eyes.

The pathogenesis of glaucomatocyclitic crisis is not fully understood but is thought to involve increased aqueous outflow resistance from an acute trabeculitis[2,5] and there is growing evidence that infectious agents – particularly cytomegalovirus (with distinct genotypes of CMV being associated with glaucomatocyclitic crisis and anterior uveitis as opposed to CMV retinitis) and possibly *Helicobacter pylori*, may play a role.[2,5,7] A prostaglandin-mediated response is suggested as elevated prostaglandin levels in the aqueous have been positively correlated with IOP.[4] The presence of HLA-Bw54 haplotype has been suspected to increase susceptibility in some cases.[4,5]

The glaucomatocyclitic crisis is a rare condition, and the first reported case in Nigeria was only reported in 2004.[8] Omoti and Enock[9] have suggested that missed diagnosis may partly contribute to its relative rarity, as glaucomatocyclitic crisis could easily be misclassified as glaucoma secondary to acute anterior uveitis. An acute IOP rise in glaucomatocyclitic crisis and demonstrable CMV or other viral acute anterior uveitis can present in nearly identical manner. Some distinguishing features of glaucomatocyclitic crisis from uveitic glaucoma include mild iridocyclitis and pain, the absence of posterior synechiae or peripheral anterior synechiae, and an increase in IOP that is usually out of proportion to the inflammatory features.[1] However, serological and aqueous testing are justified if a clinical distinction cannot be made.[2]

In young patients suspected to have a glaucomatocyclitic crisis, such as our index patient, it is important to exclude juvenile open-angle glaucoma, especially with the volatility of IOP in the latter. In juvenile open-angle glaucoma, the raised intraocular pressure is more often bilateral, and without anterior chamber activity.

Treatment of glaucomatocyclitic crisis includes topical corticosteroids (or nonsteroidal anti-inflammatory) and aqueous suppressants.[2,10] Surgery remains controversial[3,5] because attacks are self-limited and the disease becomes quiescent in later years. However, those who develop glaucoma that is uncontrolled by medications will require surgery.

There is a dearth of information of glaucomatocyclitic crisis occurring in children, and reports have only been in adults except for one case reported in a 13-year-old male.[3] To the best of our knowledge, this is the first case reported case of an African adolescent. The case report by Omoti and Enock[9] was that of two adult patients, a female age 30 years and a male age 32 years. Because of the mild nature of the uveitis, it is possible that initial attacks in childhood go undetected or unreported. Retrospective evaluation of symptoms suggestive of attacks in the past has not been studied in patients with glaucomatocyclitic crisis. Furthermore, if the increased IOP in glaucomatocyclitic crisis is the result of acutely increased aqueous outflow resistance, as has been postulated, then it is reasonable to assume that the acute rise in IOP is more

marked in the older patient with comparatively diminished capacity of the trabecular meshwork.

The diagnosis of glaucomatocyclitic crisis in a child may signal an underlying predisposition to open-angle glaucoma. In fact, a positive association between glaucomatocyclitic crisis and primary open-angle glaucoma has been demonstrated.[6,11] Because of the risk of progression to chronic open-angle glaucoma, patients with glaucomatocyclitic crisis must be monitored closely, with the other eye also at risk.[5,10,12] In summary, glaucomatocyclitic crisis do occur in the paediatric age group and should be considered in the differential diagnosis of uveitis and elevated IOP in this class of patients, especially when the elevated IOP is out of proportion to the uveitis.

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Conflicts of interest

The authors report no conflicts of interest.

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