CASE REPORT

Angle closure as a cause for intermittent headache in a child

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SUMMARY

Angle closure is usually associated with older patients, as it typically manifests in middle to later life, being associated with an age-related increased lens volume. However, angle closure can occur in any age group if there is an anatomical predisposition that promotes pupillary block or an anterior pulling of the iris. During an acute angle closure, patients generally experience ocular pain, headache, nausea, vomiting and conjunctival hyperaemia. These attacks can be misinterpreted as migraine, particularly if subacute or chronic and the demographic characteristics of the patient do not suggest a primary angle closure event. Diagnosing a headache as ocular related is of paramount importance, since there is an effective treatment. We report a case of a child with intermittent headache which revealed a subacute angle closure in both eyes.

BACKGROUND

This case is remarkable in the sense that symptomatic angle closure can mimic migraine-like symptoms. While rare in children, this condition should be thought of in the algorithm for headache assessment as it can potentially be resolved. Furthermore, early diagnosis and treatment can prevent long-term ocular damage due to intermittent angle occlusion.

CASE PRESENTATION

A 13-year-old female was referred to the ophthalmology department as part of a multidisciplinary approach to exclude precipitating factors that could relate to her headaches. The child referred episodes of blurred vision when reading for a long period of time sometimes associated intermittent headaches. She experienced headaches for at least 6 years. She described her headache as intermittent, with asymmetric intensity between episodes, predominantly felt over the fronto-parietal areas bilaterally, which was mainly triggered with activities under dark light, and it showed mild response to paracetamol and non-steroid anti-inflammatories. The headache had a frequency of two to three episodes per week with a duration of 1 to 2 hours with the child grading it as a 5 on a scale of 1–10. She denied any symptoms of fever, nausea, vomiting, eye redness, tearing, swelling of the eyelids or pupillary changes. Her behaviour had not changed according to her mother, although she had an attention-deficit/hyperactivity disorder, for which she was not taking medication.

Her medical history was unremarkable with no previous ophthalmology-related consultations related to the headaches.

The patient was afebrile. Cardiovascular examination was unremarkable with a regular heart rhythm at a rate of 70 bpm and a normal blood pressure of 125/65 mm Hg. Examination revealed normal pupillary reflexes. Visual acuity (VA) was normal in both eyes (20/20 on Snellen Scale). The intraocular pressures were asymmetric with 24 mm Hg in the right eye (OD) and 18 mm Hg in the left eye (OS) on applanation tonometry (normal intraocular pressure: 10–21 mm Hg). Slit-lamp examination suggested a shallow anterior chamber in both eyes. Funduscopy revealed asymmetric optic discs with the right eye having an excavation of 0.5 and the left eye having an excavation of 0.3 (figure 1). Both optic discs had normal limits and colour.

INVESTIGATIONS

An optic coherence tomography of the optic disc confirmed the asymmetry found on fundus examination with a mild loss of the retinal fibre layer on the temporal quadrants of the right eye, which was normal in the left eye.

The clinical data prompted an evaluation in the glaucoma clinic, with dynamic gonioscopy demonstrating iris-trabeculum contact over 360°.

Visual field tests were unreliable due to poor cooperation.

An ultrasound biomicroscopy (UBM) was performed which revealed a narrow anterior chamber angle with an anterior insertion of the iris plate and ciliary body (figure 2).

DIFFERENTIAL DIAGNOSIS

The clinical findings at presentation were consistent with angle closure glaucoma (ACG) in OD and angle closure in OS, with both cases having a probable relation to an underlying plateau iris syndrome.

TREATMENT

After explaining the situation, an informed consent was obtained for a laser peripheral iridotomy which was performed on both eyes. Iridotomy patency and mild angle opening were observed with an UBM 1 month postoperatively, confirming a plateau iris syndrome (figure 3).

OUTCOME AND FOLLOW-UP

A follow-up appointment 6 months later revealed a stable VA, normal intraocular pressures under

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Figure 1 Retinography of the right (A) and left (B) optic discs at presentation.

no topical therapy (OD 16 mm Hg and OS 15 mm Hg) and an unchanged fundal appearance. Remarkably, the patient's headaches improved significantly, from two to three episodes a week to one per month after the laser treatment. However, due to plateau iris configuration, the patient keeps follow-up on glaucoma clinic.

DISCUSSION

Primary ACG is a leading cause of irreversible blindness worldwide.¹ Quigley and Broman² estimated that, by the year 2020, there will be 79.6 million people worldwide with glaucoma, with nearly 21 million people having ACG.² An acute ACG occurs when intraocular pressure rapidly rises as a result of blockage of the trabecular meshwork by the iris. These patients generally report ocular pain, headache, nausea, vomiting and conjunctival hyperaemia.³

Angle closure glaucoma is usually associated with older patients, since it typically manifests in middle to later life.⁴ Only a few reports of ACG have been reported in younger age groups.^{4–8} In 2003, Ritch *et al*⁹ did a retrospective study on the clinical findings of patients with angle closure aged 40 years or younger.⁹ The study population represented only 2.34% of their angle closure patients. The majority was female, and mean age at the time of consultation was 34 ± 9.4 (SD) years (range: 3–68 years). The most common diagnosis was plateau iris syndrome (52.2%) and other structural/developmental anomalies differing from the relative pupillary block seen in older age groups.⁹

Plateau iris configuration is defined as an angle appearance in which the iris root is pushed forward and centrally.¹⁰ Plateau iris syndrome is diagnosed on the basis of persistent narrow angle capable of closure after peripheral laser iridotomy accompanied by a double hump sign on indentation gonioscopy.^{9 11 12} In plateau iris syndrome, anterior inserted ciliary processes provide the apposition of the iris root to the trabecular meshwork resulting in acute or intermittent angle closure.¹³

Patients with plateau iris syndrome tend to be female, present in a younger age and often have a family history of ACG.⁹ According to Stieger *et al*,¹³ the reason for increased occurrence

Figure 2 Ultrasound biomicroscopy of the right (A) and left (B) eyes at presentation revealed a narrow anterior chamber angle with an anterior insertion of the iris plate and ciliary body.

Figure 3 Ultrasound biomicroscopy of the right (A) and left (B) eyes 1 month after laser peripheral iridotomy. Iridotomy patency (Ai, Bi) and mild angle opening (Aii, Bii) was observed confirming a plateau iris syndrome.

of angle closure due to plateau iris syndrome in young individuals is related to the anterior chamber morphology, already described above.¹³

Because angle closure in younger patients is unusual, the clinician must maintain a high index of suspicion.⁹ Indeed, angle closure is an important cause of secondary headache which is well defined in the last review made by the Headache Classification Committee of the International Headache Society (IHS). This international review describes the headache associated with acute glaucoma as being usually unilateral, caused by acute narrow-angle closure with other symptoms and clinical signs of this disorder.¹⁴ Diagnostic criteria for headache attributed to acute glaucoma by the IHS include a narrow angle with any headache with evidence of causation demonstrated by at least two of the following: (1) headache has developed in temporal relation to the onset of glaucoma; (2) headache has significantly worsened in parallel with progression of glaucoma; (3) headache has significantly improved or resolved in parallel with improvement in or resolution of the acute crisis; and (4) pain location includes the affected eye.¹⁴ However, this definition just concerns to an acute glaucoma crisis and does not include the subacute or the chronic presentation of angle closure in which eye symptoms and signs are milder and the pain might simulate a migraine. In our experience when the angle closure is intermittent, the headache may be the only symptom that drives the patient to clinic or to the emergency department delaying the diagnosis by an ophthalmologist. We think that an intermittent headache, asymmetric between episodes, that is triggered under dark light should raise the hypothesis of narrow angle induced.

Prompt treatment with laser peripheral iridotomy is indicated in patients with angle closure. However, laser peripheral iridotomy usually will not be completely effective in iris plateau syndrome, eventually requiring alternate treatments.^{9 13} This is due to the fixed anterior insertion of the iris and ciliary body. The iris procedure in these cases is meant to remove a pupillary block and to partially open the angle, hopefully to reduce both symptoms and to preclude or delay the need to further intervention. Accordingly, in our case, we did not expect a full remission of the symptoms. However, the procedures to fully resolve such block would involve a painful temporary procedure (iridoplasty) and/or a surgical intervention. Both options were considered—by consensus with patient and parents—to be overtreating. A close follow-up with periodic gonioscopy was decided, since the child

had a sustained improvement of the symptoms after the partial angle opening and good intraocular pressure control.

In examining this child, we ruled out known secondary causes of angle closure in children, such as tumours, trauma, retinopathy of prematurity and uveitis.

This clinical case sustains that we must not forget angle closure as a cause for headaches in children and shows the clinical challenge of these patients, particularly if seen first on a neurology or paediatric clinic.

Learning points

- ▶ Subacute or chronic angle closure can mimic migraine-like symptoms.
- ▶ While rare in children, angle closure should be thought of in the algorithm for headache assessment as it can potentially be resolved.
- ▶ Early diagnosis and treatment can prevent long-term ocular damage due to the intermittent angle occlusion.
- ▶ Headache triggered or worsened by dark light should raise the possibility of ACG.
- ▶ The present case suggests that angle closure must be considered in the evaluation of young patients with headache.