CASE REPORT

Unusual Presentation of Spasm of Near Reflex Mimicking Large-Angle Acute Acquired Comitant Esotropia

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ABSTRACT

We report the case of an 11-year-old boy who presented with sudden esotropia, binocular diplopia, and blurred vision. The patient was neurologically normal. He had a large, constant, comitant, alternating esotropia associated with minimal accommodative spasm. Ocular motility and pupillary reactions were normal. He was diagnosed to have spasm of the near reflex presenting as acute onset of esotropia. The esotropia was persistent despite treatment and eventually resolved with prolonged cycloplegic therapy. This unusual case illustrates that spasm of the near reflex can have unique and variable presentations. Spasm of the near reflex needs to be considered in the differential diagnosis of every case of acute, acquired, comitant esotropia. This is the first case of spasm of the near reflex where persistent esotropia is reported in the absence of any neurological disorder.

Keywords: Acute onset comitant esotropia, atropine, spasm of near reflex

The near reflex consists of accommodation, convergence, and miosis. Spasm of the near reflex (SNR) is a rare and usually self-limiting disorder that involves intermittent and variable episodes of esotropia, pseudomyopia, and pupillary miosis.^{1,2} Limited abduction, unilateral or bilateral, may be associated with SNR.

Due to the rarity of this condition, affected children may be misdiagnosed as having childhood esotropia, sixth nerve palsy, divergence insufficiency, horizontal gaze palsy, ocular motor apraxia, or myasthenia gravis.³

Acute acquired comitant esotropia (AACE) is an unusual presentation of strabismus in older children and adults characterised by acute onset with diplopia, concomitance of strabismus, a large angle of deviation, absence of paretic element, and potentially normal binocular vision.⁴

SNR is usually not considered in the differential diagnosis of AACE because a typical presentation of SNR with intermittent, usually small-angle esotropia, pseudomyopia, limitation of abduction, and miosis is easily differentiated. We present an unusual case of SNR presenting with a constant large-angle esotropia, diplopia, and variable accommodative spasm that may be misdiagnosed as an AACE.

CASE REPORT

A 11-year-old boy presented with complaints of inward deviation of the eye, diplopia, and blurred vision for the past 1 month. The parents reported that the onset of squint and diplopia was sudden and had been constant since then. There was no history of recent illness, head trauma, or taking any medications.

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FIGURE 1 The patient at presentation with a large-angle alternating esotropia and normal abduction.

The parents had previously consulted multiple eye care practitioners, and the child had been prescribed glasses with -1.00 dioptre sphere (DS) in the right eye and -0.50 DS in the left eye after non-cycloplegic refraction. He had been diagnosed to have an acute, acquired esotropia, and neuro-imaging was advised.

The examination revealed unaided visual acuities of 20/40 in each eye for distance. Ocular motility was full. Pupils were equal in size, and reflexes were brisk and equally reacting to light in both eyes. Cover test showed a large-angle, comitant, alternate esotropia (Figure 1). The esotropia measured 60 prism dioptres (PD) at distance and near. Static retinoscopy showed a variable myopia of approximately -4.00 DS in the right eye and -1.00 DS in the left eye. Dynamic retinoscopy was unstable and revealed a lead of accommodation of -1.00 DS in both eyes. The fundus was normal.

However, the examination was limited because the patient was in obvious distress and his fixation was unstable. The neurological evaluation was normal.

The patient was advised cycloplegic refraction after application of homatropine (2%) twice a day for 3 days. On review, it was noted that the cycloplegia was incomplete.

The patient was then advised to use 1% atropine eye drops twice a day for 1 week, with punctal occlusion to reduce systemic absorption. On review, the cycloplegic refraction of right eye (RE) was $+0.25$ DS / -0.50 DC $\times 160$ and left eye (LE) was $+0.50$ DS. The visual acuity and strabismus were evaluated under cycloplegia to assess the effect of accommodative relaxation. The vision had improved to 20/30 in the right eye and 20/20 in the left eye. The near vision was N24 in both eyes. However, the alternating esotropia and diplopia persisted without any change in magnitude. The patient was comfortable with the medication but distressed about the persistent squint, diplopia, and blurred vision for near. He was prescribed bifocal glasses with a near addition of $+3.00$ DS and advised to continue 1% atropine eye drops once a day.

The patient was diagnosed to have an acute SNR in view of the convergence spasm with esotropia and induced pseudomyopia that disappeared under cycloplegia. A contrast-enhanced magnetic resonance imaging (MRI) scan of the brain and orbit, which was normal, had already been obtained.

Further neurological evaluation was not performed in view of this.

We presumed that the aetiology was functional. The child came from a patriarchal background where any mental illness is stigmatised, and as such the parents refused a suggestion to obtain psychological counselling. However, the child appeared emotionally stable, with no obvious domestic or academic pressures that might have precipitated the SNR.

The patient was reviewed weekly after starting therapy. He tolerated the atropine eye drops well with no adverse effects. The accommodative spasm was relieved with clear vision for both distance and near with glasses. But the patient was anxious about the persistent diplopia and objectively, the angle of esotropia remained constant at 60 PD.

After 1 month of continuous use of atropine eye drops, the patient reported that the squint and diplopia had suddenly improved over 2 days. On evaluation, the vision was 20/20 in each eye for distance and N6 for near with the near addition. The ocular motility was full. There was a minimal esophoria of 2–3 PD for distance and near with a stereoacuity of 60 arc seconds. He was advised to continue treatment with 1% atropine eye drops once a day and bifocal glasses. The patient was maintained on atropine eye drops once a day for 2 months. The duration of atropine was prolonged in view of the time taken for initial resolution of the esotropia. The patient was then gradually weaned off medication over a period of 2 months after being counselled about the possibility of recurrence and need for long-term follow-up.

On evaluation after complete cessation of atropine, his unaided visual acuity was 20/20 in both eyes and ocular alignment was orthophoric for distance and near (Figure 2). He had an accommodative facility of 4 cycles per minute with flippers of power $+2.00/-2.00$ DS, with difficulty in relaxation of accommodation. The patient has been prescribed accommodative facility exercises to improve the flexibility of the accommodative system.

DISCUSSION

Near fixation is normally accompanied by convergence, accommodation, and miosis. A SNR occurs

FIGURE 2 The spasm of near reflex was relieved with a combination of atropine eye drops and bifocals.

when one or more of those three components exceeds the demand required by the stimulus of interest.² SNR represents a constellation of clinical findings that may appear together or separately, in varying degrees. Whereas some patients may have isolated spasm of accommodation without involving convergence and pupils, others may show spasm of convergence without abnormal accommodation.^{1,2}

Hussaindeen et al. presented a similar case of a 23-year-old male with comitant esotropia (30 PD), pseudomyopia, full ocular motility, and no miosis.⁵ Their patient had a functional aetiology and developed the convergence spasm after wearing overcorrected glasses of -5.00 DS. Similar to our case, they have also noted persistence of esotropia despite cycloplegic therapy. They have speculated that their patient could have initially been in a state of ill sustained accommodative dysfunction that could have been aggravated by the increased minus prescription precipitating a state of SNR. In their case, the diagnosis was relatively simple in view of the intermittent esotropia, fluctuating myopia, and functional aetiology.

SNR is typically considered to be a self-limiting functional disorder. In our patient, the aetiology was assumed to be functional in the absence of any neurological signs and a normal neuro-imaging. It is to be noted that the child did not fit into the usual profile of patients with accommodative dysfunction. He seemed to be a well-adjusted child with no history of any complaints of a functional nature.

Patients with SNR usually present with intermittent episodes of diplopia, eyestrain, headache, and blurred vision.^{1,2} Limitation of abduction may be seen during episodes of convergence spasm, but frequently the esotropia component may appear with full abduction.² The occurrence of miosis is also not a reliable aid for diagnosis of SNR because it is difficult to document, as the fixation pattern at the onset of spasm may be unstable and variable. This task may be further hampered by the dark pigmentation of the iris.^{2,5}

AACE is an uncommon presentation of strabismus in older children and adults.⁴ Three main types have been described. The Swan type (type 1) refers to an esotropia due to the interruption of fusion. Burian-Franceschetti type, or AACE type 2, lacks clear

associations; however, physical or psychological stress has been suggested as precipitating factors. Type 3, or Bielschowsky type, has been described in myopic subjects.

AACE type 2 is characterised by an acute onset with diplopia, a large angle of deviation, absence of abducens nerve palsy, and potentially normal binocular vision.⁶ A low amount of hypermetropia and no accommodative component are regarded to be typical of this entity.⁷ The preferred treatment option is surgery in order to restore ocular alignment and binocular vision. Previous series on AACE type 2 have consisted of heterogeneous groups, with Legmann Simon and Borchert including patients with late-onset refractive-accommodative esotropia in their study.^{6,7} It is possible that some of the children with suboptimal alignment after squint surgery had an associated dysfunction of the near reflex that was overlooked.⁴

A number of neurological associations for accommodative spasm have been reported in literature, including trauma, Wernicke-Korsakoff syndrome, Arnold-Chiari malformation, pituitary disease, pineal gland tumour, and sixth nerve palsy.³ Persistent accommodative spasm, resistant to conventional treatment, has been reported in association with organic causes, including closed head trauma.^{3,8} It is therefore important to rule out a central nervous system aetiology in cases of SNR by detailed ophthalmic and neurological evaluation in combination with neuro-imaging.

Cycloplegic agents, in combination with reading glasses for near work and orthoptic exercises, are recommended in the management of SNR.² Atropine, a non-selective muscarinic antagonist, breaks the SNR by paralysing the ciliary muscle. Conversely, atropine may stimulate accommodative effort and induce convergence spasm as the patient attempts to overcome the blurred near vision induced by cycloplegia. Hence, it is essential to combine cycloplegic therapy with near addition glasses to minimise the accommodative effort and induced convergence. Long-term use of atropine eye drops has the risk of side effects, including skin rashes and tachycardia, due to systemic absorption. The Atropine for the Treatment of Childhood Myopia trial studied the long-term effect of 1% atropine eye drops on myopia progression

in children and reported no serious adverse effects.⁹ The patients on long-term treatment need to be counselled regarding the potential side effects and instructed to perform punctual occlusion after application of the medication to reduce systemic absorption.

This case of SNR had a unique manifestation as a large-angle, constant esotropia with minimal pseudo-myopia. Ocular motility was full despite the large angle of esotropia, and pupillary miosis was absent. The esotropia was eventually relieved only with a combination of prolonged atropinisation and bifocals. Isolated persistent accommodative spasm has been previously reported in cases with neurological damage, but these cases did not have an associated spasm of convergence.^{3,7}

This case emphasises the need to consider SNR in the differential diagnosis of every case of AACE. A careful assessment of vision, ocular alignment, dynamic retinoscopy, and cycloplegic refraction to detect induced myopia are key to making the correct diagnosis. Recognizing SNR is important to prevent unnecessary, expensive, and potentially harmful treatment, including strabismus surgery.¹⁰

As SNR appears more tenacious in eyes with dark pigmented irides, therapy should be initiated with a combination of atropine eye drops and near addition glasses. Weaker cycloplegics such as homatropine seem to be ineffective. Even with the use of topical atropine, prolonged therapy may be needed to completely relax the accommodative effort.

In conclusion, this is the first reported case of SNR with persistent esotropia, despite cycloplegia, in a child with no history of head trauma or associated neurological disorder.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Note: Figures 1 and 2 of this article are available in colour online at <http://informahealthcare.com/oph>.

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