Case report of concomitant intermittent exotropia as a rare presentation in a child with temporal arachnoid cyst

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Abstract

Aim: To report a case of arachnoid cyst incidentally detected in a child who initially presented with concomitant intermittent right exotropia that progressed to incomitancy.

Methods: This is a case report of a 3-year-old boy with concomitant intermittent right exotropia who was initially listed for strabismus surgery. However, 2 months later, prior to the operation, he was found to have developed constant V-pattern right exotropia and bilateral inferior oblique overactions.

Results: Urgent CT and MRI revealed the incidental finding of a temporal arachnoid cyst measuring 2.8 cm × 3.8 cm × 3.2 cm displacing the temporal lobe posteriorly. Urgent marsupialisation of the cyst was done with subsequent improvement to the strabismus.

Conclusion: Arachnoid cysts are developmental anomalies that occasionally cause sudden neurological deficit due to cyst expansion or bleeding. Although concomitant exotropia is generally benign in a child, intracranial lesions need to be investigated if the child shows progressive worsening of strabismus, especially with the recent onset of incomitance.

Key words: Arachnoid cyst, Concomitant exotropia, Incomitant strabismus, Marsupialisation

Introduction

Although acute concomitant strabismus in children is generally benign, numerous past reports have shown that acute comitant esotropia can be associated with intracranial pathology such as brainstem tumours or hydrocephalus.¹ However, no such association was found in the literature with regard to acute concomitant exotropia in children. Nevertheless, the clinician should be extra careful when evaluating children with acute concomitant strabismus, especially those who present with progressive change in the angle of deviation, and incomitancy as well as nystagmus.² These children should have immediate radiographic investigation such as computed tomography (CT) or magnetic resonance imaging (MRI) performed, even in the absence of signs suggesting raised intracranial pressure and neurological deficits.²

We report a child with temporal arachnoid cyst who initially presented with intermittent exotropia of the right eye.

Case report

A 3-year-old Indian boy presented to the University of Malaya Medical Center with concomitant right intermittent exotropia for the past year with increasing frequency, albeit normal ocular movement. It occurred mostly when he was tired or fixating on a distant object. Perinatal history was uneventful and he had normal developmental milestones. However, family members on the mother’s side had a history of divergent strabismus.

Since the previously intermittent right exotropia was becoming constant, we decided to proceed with recess and resect surgery to the horizontal recti muscles of the right eye in order to improve ocular alignment. However, the child developed a left head tilt and constant V-pattern right exotropia with bilateral superior oblique underactions with associated inferior oblique overactions (Right > Left) during pre-operative assessment 2 months later (Fig. 1). There were neither symptoms nor signs of raised intracranial pressure, neurological deficits or recent trauma. Ocular examination revealed healthy globes with no fundus pathology. Although binocular visual acuity tested with both eyes open was 6/19, no stereopsis was demonstrable. As the patient’s strabismus had progressed acutely with the additional element of incomitance (V-pattern) as well as features suspicious of bilateral superior oblique palsies, further investigations were warranted to rule out an intracranial lesion.

Urgent CT and MRI scans of the brain were performed which showed the presence of a large left temporal lobe subarachnoid cyst in the middle cranial fossa measuring 2.8 cm × 3.8 cm × 3.2 cm (Fig. 2). It arose from the left sylvian fissure and displaced the temporal lobe posteriorly without midline shift. Pediatric neurosurgical expertise was sought and open craniotomy with marsupialisation of the cyst wall was performed. During the operation, it was noted that the cyst wall was tense (Fig. 3). Despite successful initial evacuation of the cyst, the procedure was complicated by subdural hygroma, prompting a second craniotomy to drain the sequestered

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cerebrospinal fluid. Following that, the child recovered uneventfully.

Strabismus in our patient improved after the first craniotomy. Right exotropia became intermittent again while the inferior oblique overactions had resolved. However, the frequency of right exotropia worsened a year later. Orthoptic assessment then showed right constant exotropia with inferior oblique overaction. Neither recurrence of the arachnoid cyst nor other intracranial lesions were detected on a repeat CT scan of the brain. Thus, medial rectus resection and lateral rectus recession (6 mm each) of the right eye were subsequently performed. Post-operatively, good ocular alignment was achieved with glasses but stereopsis was not restored.

Discussion

In concomitant strabismus, the angle of ocular deviation remains unchanged regardless of the direction of gaze. Children commonly present with concomitant strabismus due to familial inheritance or associated refractive error. Yet there have also been instances where acute comitant esotropia was a known indicator of potential intracranial tumour. Although rare, tumours of the cerebellum, brainstem, pituitary region and corpus callosum have been found.

Ciancia and Lennestrand had postulated a possible theory for this esodeviation and suggested that the vergence mechanism in the mesencephalon was affected either directly by the expanding tumour or indirectly via hydrocephalus. As the supranuclear vergence pathway in humans was poorly understood, this postulation was based on primate studies that showed the vergence neurons located in the dorsal and dorsolateral portion of the oculomotor nucleus in the midbrain. These studies had also noted that both elements of divergence and convergence arise from the same vergence neurons. Therefore, similar intracranial pathology could theoretically give rise to concomitant exotropia if these vergence neurons were compromised.

However, very few cases pertaining to acute second-
ary concomitant exotropia have been reported in the literature compared with acute comitant esotropia with an underlying intracranial lesion. Comoglu et al., who reported the case of a 60-year-old man with acute exotropia and bilateral blepharospasm secondary to bilateral paramedian thalamic infarct, believed such an acute concomitant exodeviations could be due to convergence paralysis or excessive stimulation of the divergence neurons. The other reported cause of acute secondary concomitant exotropia is heroin intoxication, where the opiate was suspected to have had an adverse effect on the vergence center in the midbrain, although no opiate receptors were shown in that area.

In the case of our patient, we believe that the initial intermittent concomitant exotropia could have been the result of either direct compression of the vergence center in the midbrain by the temporal arachnoid cyst which had displaced the temporal lobe posteriorly, or indirect compression via raised pressure transmitted through the cerebrospinal fluid, despite the lack of evidence suggesting raised intracranial pressure such as papilledema and dilated ventricles. Over time, as the cyst expanded, the pressure on the vergence center increased. This caused the exotropia to become constant with the added element of incomitance. Apparently, this damage was reversible, as the exotropia reverted to its previous intermittent concomitant nature after evacuation of the cyst. Yet this reversal was temporary as the patient’s exotropia decompensated a year later despite the absence of tumour recurrence. Therefore, the decompensation is not readily explainable.

As opposed to concomitant strabismus, incomitant strabismus in general is indicative of infranuclear cranial nerve lesion or mechanical limitation of movements due to extraocular muscle myopathy or orbital lesion where the primary pathology needs to be thoroughly investigated. Primary treatment aims to eradicate potentially life-threatening disorders while at the same time the ophthalmologist strives to relieve diplopia, improve binocular vision and deter amblyopia. Hence the acute onset of incomitant strabismus in a child with pre-existing concomitant strabismus warrants immediate radiological investigation to rule out a potential intracranial lesion, as illustrated in the case above. Apart from incomitance, acute onset of other ocular motility disorders such as nystagmus, with increasing frequency and angle of deviation, as well as poor oculomotor fusion after strabismus treatment, could also herald the presence of an underlying intracranial insult.

Ocular misalignment resulting from brain tumour interrupts binocular sensory fusion and causes strabismus in children. This misalignment can either be permanent or correctable after treating the underlying intracranial tumor by radiotherapy, chemotherapy or surgical excision. Some may require further strabismus surgery after the primary treatment to improve ocular alignment. Shalev and Repka found that these children may regain the ability to fuse if their misalignment can be corrected. They observed that 10 of 14 children with incomitant strabismus regained good stereopsis once their brain tumour was treated and their eyes realigned. This was in contrast to those children with acute comitant esotropia secondary to brain tumour where most literature reported absence of recovery despite optimal treatment. These reports concurred with our case finding: our patient who also presented initially with concomitant strabismus did not regain stereopsis despite optimal ocular alignment. Shalev and Repka also reported that patients with a shorter duration of ocular misalignment and older age at diagnosis regained better stereopsis. These factors were also related to other forms of strabismus and were thought to be associated with reduced likelihood of suppression.

Arachnoid cyst is a benign cystic collection of fluid within the arachnoid membrane and subarachnoid space of the cisterns and major cerebral fissures. Although most of these cysts are asymptomatic and remain stable throughout life, some cause sudden and rapid expansion, compressing adjacent brain parenchyma and resulting in neurological deficit. Overall, arachnoid cysts account for 1% of all intracranial mass lesions. They commonly involve the middle cranial fossa such as the temporal lobe (50–60%), predominantly in the left hemisphere. The male to female ratio is 4 to 1.

Arachnoid cysts are either congenital or acquired. Congenital cysts occur as a result of splitting or duplication of the primitive arachnoid membrane in early embryonic life. Acquired cysts result from cerebrospinal fluid sequestration in the arachnoid membrane due to inflammation, trauma, tumour, intracranial haemorrhage and surgery. Yet most of these cysts are incidental finding at intracranial CT or MRI and do not require treatment if they remain static in size. However, they may become symptomatic early in life due to rapid expansion following minor closed-head injury. Frequently, the absence of head trauma in the history cannot rule out such intracranial injury, because the parents or guardians might not be aware of it. Cysts can also rupture, resulting in subdural hygromas, haematomas and intracranial hypertension.

Clinical features of arachnoid cysts vary according to their anatomical relations. Ninety per cent of them are found at supratentorial locations, where 60% are located in the middle cranial fossa as in our patient. This is a common site for arachnoid cyst formation due to the arachnoid covering of the temporal and frontal lobes failing to merge at the sylvian fissure early in life. Thirty per cent of these middle cranial fossa cysts are large; they can occupy the whole temporal lobe and occasionally extend to the frontal region and convexity. Cysts in this location have been reported to cause acute psychosis, seizures, headaches, hemiparesis, raised intracranial pressure, craniomegaly and developmental delay.

The literature has also shown on numerous occasions that ophthalmologists are the first-line doctors in the multidisciplinary team approach to treating patients with arachnoid cysts when they present primarily with ophthalmic symptoms. The progression of strabismus in our patient from intermittent concomitant exotropia to incomitant V-pattern alternating exotropia with bilateral superior oblique palsy is similar to a case reported by Ohtsuka et al. Their patient presented with acute diplopia and bilateral trochlear nerve palsy due to an arachnoid cyst in the quadrigeminal cistern which had spontaneously enlarged. However, the suspected bilat-
eral superior oblique palsy in our patient was not readily explainable from the scan that showed the cyst located in the left anterior temporal fossa extending into the interpeduncular cistern and away from the quadrigeminal cistern. Another report, by Pagni et al., also showed left trochlear nerve involvement secondary to an arachnoid cyst of the quadrigeminal plate.19

Patients with arachnoid cyst may also present to ophthalmologists with problems other than strabismus. Ishii et al. and Stefanii et al. recently reported an adult with a middle cranial fossa arachnoid cyst with rapidly deteriorating visual field defect and visual acuity impairment due to optic nerve compression.20,21 Kaisho et al. also reported optic nerve compression in a 16-year-old with a suprasellar arachnoid cyst that resulted in optic atrophy.22 Not all compression leads to severe morbidity, as shown by a case study by Keller and Flammer, who reported a 36-year-old woman with a large fronto-temporo-parietal arachnoid cyst who presented with headache and unilateral papilloedema of the left eye without visual impairment.23 In addition to nerve compression, proptosis is also a feature of middle cranial fossa arachnoid cyst. Tsitouridis et al. described a case series of 23 patients with proptosis due to anterior temporal arachnoid cyst which caused posterior and lateral orbital wall remodelling and subsequent anterior displacement of the orbital content.24

Not all arachnoid cysts require surgical intervention. Yamauchi et al. and Weber et al. reported spontaneous regression of temporal arachnoid cyst after 7 years and 10 years of observation, respectively.25,26 However, surgical treatment is indicated in cases where the cyst causes neural compression, hydrocephalus and raised intracranial pressure. Post-operatively, the cyst can recur in about 25% patients.27 Surgical techniques remain controversial and these include craniotomy, open fenestration or marsupialisation of the cyst, stereotactic cyst aspiration, endoscopic fenestration and, last but not least, shunt placement.13 Subdural hygroma is a well-known complication of the standard technique of marsupialisation whereby the cyst wall opens into the subdural fluid space, brain cistern or ventricles; its incidence is 6%,13 As with our patient, such a complication necessitates a second craniotomy to drain the fluid. Although endoscopic fenestration was once discouraged due to the increased risk of bleeding, it has become the future direction of treatment strategy as endoscopic technology improves.28 This technique avoids the need for craniotomy as well as complications associated with the shunting tube.

Conclusion

Although concomitant strabismus can be associated with underlying intracranial tumour, most cases need no further investigation as they are generally benign. However, sudden acute changes such as nystagmus, worsening of the angle of deviation and incomitancy in a child with concomitant strabismus are indicative of an underlying intracranial pathology that needs to be investigated urgently. Early and prompt treatment not only saves lives, it can help to restore binocularity and stereopsis.

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