Acute V Pattern Esotropia Without Abduction Deficit, Secondary to a Posterior Fossa Pilocytic Astrocytoma

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ABSTRACT
A case of acute esotropia with bilateral inferior oblique muscle overaction in a 4-year-old boy with a posterior fossa pilocytic astrocytoma is reported. After tumor excision, the esotropia and oblique dysfunction resolved. Acute esotropia with bilateral inferior oblique muscle overaction without abducens palsy is an unusual sign of brain stem mass. [J Pediatr Ophthalmol Strabismus 2009;46:235-237.]

INTRODUCTION
Pilocytic astrocytomas predominantly occur in children and account for 10% to 20% of all childhood tumors. Common presenting signs and symptoms include nausea and vomiting, headache, ataxia, and visual complaints, such as diplopia or blurred vision. It is common for symptoms to occur for many months or years before medical attention is sought. Symptoms are usually caused by increased intracranial pressure as a direct result of the mass effect of the tumor or from direct invasion of the cranial nerve, as in the case of a 9-year-old girl with trochlear palsy.1 After prompt diagnosis and total resection, the 5-year survival rate is good, at 95%.2

This report describes a case of acquired esotropia with bilateral inferior oblique muscle overaction. This is a common presentation to an orthoptic department; however, in this case, a posterior fossa tumor was diagnosed and excised, with resolution of the strabismus 4 months later.

CASE REPORT
A 4-year-old boy had a history of recent onset of convergent strabismus noticed by his parents and teachers for the past 4 days. He had complained of increasing headaches for 5 weeks that were worse in the mornings and were relieved by paracetamol (acetaminophen). He had no vomiting or nausea. There was no other significant history. He was born at 34 weeks’ gestation, after a normal delivery. He spent 4 weeks in the special care baby unit and was discharged. There was a maternal family history of strabismus. He was otherwise fit and well.

Initial orthoptic examination showed visual acuity within normal limits (0.1 logarithm of the minimum angle of resolution in the right and left eyes) and moderate to marked right esotropia for near and distance, measuring greater than 45 prism diopters. Ocular movements showed full abduction in both eyes, with bilateral overacting inferior oblique muscles. Right overaction (+3) was greater than left overaction (+1). Measurements in side gaze positions were not possible because of lack of patient cooperation.

Funduscopy showed healthy optic discs and maculae, with no papilledema. Cycloplegic refraction showed low hypermetropia on the right and left (+0.75/+0.25), and glasses were not prescribed. Computed tomography and subsequent magnetic resonance imaging showed evidence of a large posterior fossa tumor involving the right side of the cerebellum, the vermis, and also part of the left side. Two months later, the patient underwent posterior fossa craniectomy and excision of the cerebral tu-
mor. Histopathologic examination confirmed a glial tumor with a biphasic appearance, with both compact fibrous spindle cells and loose microcystic areas. The tumor was diagnosed as a pilocytic astrocytoma, World Health Organization grade 1.

At the first postoperative visit, the convergent strabismus and ocular movement findings were unchanged. Two weeks later, there was no change in ocular status and the patient was scheduled for botulinum toxin injection in the right medial rectus muscle, but he did not attend the appointment. At the next visit, 5 months after the onset of symptoms, the esotropia and oblique dysfunction had almost resolved and visual acuity was still within normal limits. On examination, small, well-controlled esophoria was present in the primary position, measuring 8 prism diopters at near and 2 prism diopters at distance. The esophoria was still compensated at distance and on extreme right and left gaze. Fusional vergences were noted and within the normal range (40 prism diopters base out to 10 prism diopters base in). Ocular movements showed minimal underaction of the right eye (to -1) on dextroversion, but ductions were full. Otherwise, findings on ocular movement examination were unremarkable. The patient remains under observation.

**DISCUSSION**

Pilocytic astrocytomas can involve the midline, basal, and posterior fossa structures and are usually considered benign tumors of childhood. Astrocytic tumors were reported as the most common tumors in a consecutive series of 216 children. Strabismus is a common presenting sign of cerebellar and brain stem tumors, especially esotropia, and is usually associated with cranial nerve VI palsy. In a study of 63 patients with intracranial chordoma, VI cranial nerve palsy was the sole presenting sign in 29% of patients, with 62% having various combinations of muscle palsies. Cranial nerve palsies were also the most common presenting sign (33%) in a study of 45 cases of brain stem gliomas, with cerebellar signs occurring in 29.8% of cases. Trochlear palsies are more rarely reported. A case involving tumor invasion into the nerve itself has been documented; however, in many cases, increased intracranial pressure from the mass or from hydrocephalus can compromise lateral rectus muscle function. Compression by hydrocephalus may be the leading cause of symptoms, such as papilledema, visual field defects, and visual loss.

Although rare, concomitant esotropia can present as a brain stem or cerebellar tumor. Acute esotropia was the presenting sign in a series of eight patients (three children and five adults) with intracranial neoplasm. Two further case reports show striking similarities to this case, one of a 5-year-old girl with a large cerebellar astrocytoma and moderate hydrocephalus and one of a 2-year-old child with pilocytic astrocytoma, both presenting with acute esotropia. In a prospective study of the clinical characteristics and features suggestive of underlying neurologic pathology in patients with acute-onset esotropia, 1 of 10 patients was found to have a cerebellar tumor, and the authors concluded that no single clinical sign can consistently be considered an indication of tumor.

Concomitant esotropia with oblique dysfunction is a common finding in an orthoptic clinic; however, sudden onset of esotropia with bilateral inferior oblique muscle overactions, even in the presence of normal abduction, should alert the examiner to consider expanding the differential diagnosis from nonorganic etiology to include brain stem mass, for example, especially in the presence of other symptoms of increased intracranial pressure. In a retrospective case series of 92 children (younger than 8 years) with superior oblique muscle palsy, 3 children were found to have a brain tumor. Skew deviation can also cause acquired vertical deviation that can mimic oblique dysfunction, and although not frequently caused by tumor, it has been reported. The oblique dysfunction in this case may have been caused by either skew deviation or superior oblique muscle palsy, although increased intracranial pressure may explain the oblique abnormality in this situation.

Initial postoperative findings implied that the esotropia may persist and that further intervention would be required; however, in this case, spontaneous resolution occurred, suggesting that conservative management initially may be prudent in these cases. However, it is important to consider the possible effects of manifest strabismus on the developing binocular system, depending on the age of the child. In a further case of pilocytic astrocytoma presenting as late-onset esotropia in a 3-year-old child, surgery was required to restore rudimentary binocularity because the esotropia did not resolve after tumor removal. In this case, neurologic signs of brain stem mass were not apparent until 8 months after presen-
The esotropia had been present for considerably longer than in the current case, reflecting the lack of spontaneous resolution and poorer binocular outcome.16

Although pilocytic astrocytoma is rare, intracranial neoplasm should be considered a diagnosis of exclusion in any child presenting to an orthoptic department with acute nonaccommodative esotropia, with or without oblique dysfunction, even in the absence of symptoms of increased intracranial pressure. In this case, prompt diagnosis and surgical removal led to an excellent result.

REFERENCES


