

A Boy With Worsening Esotropia

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Dustin* is a 7-year-old boy with accommodative esotropia. Our office has been following him since he was 2 years old. Recently, his mother asked if his regular eye appointment could be moved forward by a few weeks. She was concerned because for the previous 2 to 3 weeks, he had been holding reading material very near and closing his right eye to read. She also noted that his right eye was now crossing constantly; previously, this right-eye esotropia had been present only when he wasn't wearing glasses. He also had a mild but persistent headache above his right eye, which was relieved by ibuprofen.

We Get a Look

Dustin had no significant medical history other than a tonsillectomy in 2012, and he was not taking medication apart from ibuprofen as needed for his headache. He had been patching the left eye because of right-eye amblyopia with vision of 20/40.

On exam, he appeared to be his usual smiling self. He was wearing his glasses, which were $+6.50 +1.50 \times 60$ for the right eye and $+6.00 +1.50 \times$

130 for the left. His visual acuity with glasses had been 20/40 in the right eye, but it was now 20/100; and the left eye had dropped from 20/20 to 20/40. There was a constant 45-D right esotropia, which was a change from his prior 6-D esotropia. We could not detect any limitation in horizontal gaze, and pupil reactions were equal.

Initial Misdiagnosis

The initial thought was that his accommodative esotropia was decompensating, and he was dilated with Cyclogyl 1% (cyclopentolate hydrochloride).

Retinoscopy was $+9.00 +1.00 \times 60$ in the right eye and $+6.75 +0.50 \times 130$ in the left. This reinforced the impression that he merely needed stronger glasses and possibly more patching for esotropia.

However, our hearts sank when indirect ophthalmoscopy revealed 2 very edematous optic nerves with severely blurred margins and splinter hemorrhages (Fig. 1). This was definitely a new finding that had not been seen during prior examinations. After noting the optic nerve edema, we looked at the eye movements again and observed a mild bilateral abduction deficit indicating a sixth cranial nerve palsy. Upon further questioning about his headache, he said it hurt just a little above his right eye. An immediate MRI was ordered.

What Tests Revealed

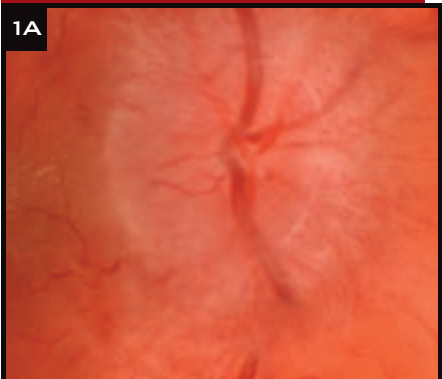
Unfortunately, the MRI showed a mass lesion, measuring $4.5 \text{ cm} \times 2.6 \text{ cm} \times 3.2 \text{ cm}$, in the fourth ventricle. It extended down to compress the cervicomedullary junction with dilation of the third and lateral ventricles (Fig. 2). The tentative radiologic diagnosis was ependymoma with obstructive hydrocephalus. Dustin was transferred that evening to the nearest pediatric specialty hospital, 3 hours away, and was scheduled for surgery.

Treatment

Dustin had a ventriculostomy placed for the elevated intracranial pressure, and the tumor was then resected via a posterior fossa suboccipital craniotomy.

What's Your Diagnosis?

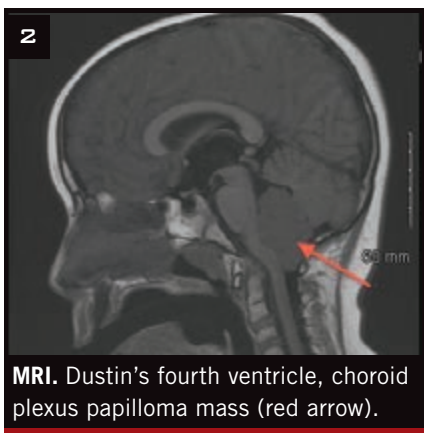
1A



1B



INDIRECT OPHTHALMOSCOPY. Dustin's edematous optic nerves.



MRI. Dustin's fourth ventricle, choroid plexus papilloma mass (red arrow).

omy. The tumor was pushing the cerebellum rostrally, allowing the neurosurgeon to work beneath the cerebellar tonsils without having to retract them. Even during the surgery, the neurosurgeon thought it was an ependymoma. However, the frozen section indicated a choroid plexus tumor, which has a significantly better prognosis. At the end of the case, the surgeon felt that the entire tumor had been resected.

About the Diagnosis

Choroid plexus papillomas develop from the ventricular choroid plexus, which produces the cerebrospinal fluid (CSF) in the brain.¹ Dustin's case was atypical because the lesion was located in the fourth ventricle. In pediatric patients, these lesions are typically found in the lateral ventricle of the brain, whereas in adults, they are more commonly found in the fourth ventricle.¹

The World Health Organization (WHO) classifies choroid plexus tumors as ranging from more benign papillomas, referred to as WHO Grade I, to carcinomas, referred to as WHO Grade III.² Excluding rare cases, choroid plexus papillomas of WHO grade I and II are almost always benign, as opposed to choroid plexus carcinomas (WHO grade III), which are malignant.² Choroid plexus papillomas cause the choroid plexus to overproduce CSF, and they can block normal CSF circulation, which leads to hydrocephalus.¹ Hydrocephalus in turn causes headache and sixth nerve palsy in addition to papilledema with blurred vision. A choroid plexus papilloma is usually treated with a total mass resec-

tion.² These resections have a high success rate even in recurrent papilloma cases, and WHO grades I and II rarely develop malignancy.²

Differential Diagnosis

Fortunately for Dustin, he did not have an ependymoma, which was the original working diagnosis. Even though ependymomas—the most common fourth-ventricle tumor in children—are not malignant, they are difficult tumors to treat, with a 5-year survival rate of around 55%, as opposed to choroid plexus papillomas (WHO grade I), which have a 5-year survival rate of 80% to 100%.^{2,3}

Another possible diagnosis was an intraventricular meningioma, which accounts for 20% of pediatric meningiomas.⁴ Approximately 80% of these are seen in the lateral ventricle, 15% are seen in the third ventricle, and only 5% are seen in the fourth ventricle.⁴ Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is another cause of increased intracranial pressure and was another possible diagnosis, although it is rare in prepubertal children.

Patient's Progress

Dustin has done amazingly well following the surgery, with complete resolution of papilledema and return of his prior visual acuity and his well-controlled accommodative esotropia. Other than the scar on his scalp, there are no residual signs of his illness.

What We Learned

Overall, a surprising aspect of the case was that Dustin had mild headaches despite the severe papilledema and significantly worsened esotropia. This case is a strong reminder to look at the optic nerve in any child with a change in vision, especially one with a significantly worsened esotropia, even in the setting of accommodative esotropia. ■

*Patient name is fictitious.

1 Jaiswal AK et al. *J Pediatr Neurosci.* 2009;

4(1):10-16.

2 Fuller C. Pathology of Choroid Plexus Neo-

Catch Up on Your Rounds

Read these recently published cases at www.eyenet.org (click "Archives" at the top of the screen).

A case of a migrating white retinal lesion. Vision in his right eye started to deteriorate 10 days after a seizure. He suffered occasional headaches and discomfort of the right brow but no eye pain. (*October*)

A slow growth. For years, she felt that her upper left eyelid was swollen. Vision in that eye became progressively blurry. Then she developed double vision on upgaze. (*September*)

A growth spurt and a growing shadow in his vision. His vision was blurry in his right eye, and he said it felt like "a shade" was covering the superior aspect of his visual field. (*August*)

A boy without color vision. First he had difficulty with color vision, and then he began losing his central vision. (*July*)

A mysterious iris mass. The 61-year-old had suffered 4 months of blurry vision, intermittent photophobia, and right conjunctival injection before seeking help. (*June*)

The nuisance protuberance that grew and grew. She first noticed the bump on her eyelid a year earlier. Initially small, it had grown and grown. Adding injury to insult, it was now starting to hurt. (*May*)

plasms. Medscape. <http://emedicine.medscape.com/article/1744050-overview#a1>. Accessed Aug. 24, 2015.

3 MacDonald TJ. Pediatric Ependymoma. Medscape. <http://emedicine.medscape.com/article/986333-overview>. Accessed Aug. 24, 2015.

4 Muzio BD, Gaillard F. Intraventricular Meningioma. Radiopaedia. <http://radiopaedia.org/articles/intraventricular-meningioma>. Accessed Aug. 24, 2015.

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