Delayed Diagnosis of Enhancing Posterior Fossa Tumors Mimicking the Tela Choroidea of the Fourth Ventricle

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Key words
- Brain tumor
- Choroid plexus
- Congenital
- Nystagmus
- Tela choroidea

Abbreviations and Acronyms
MRI: Magnetic resonance imaging

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BACKGROUND: **Posterior fossa brain tumors are common in children. Symptoms typically develop when the tumors have reached sufficient size to cause compression of adjacent neural structures or cause obstructive hydrocephalus. Many tumors in this region originate from the tela choroidea and choroid plexus of the fourth ventricle. Enhancement of the fourth ventricular tela choroidea and choroid plexus is uncommon in children, and when such enhancement is present, it may represent early tumor growth.**

METHODS: **A 5-year-old girl with a history of congenital nystagmus, for whom initial work-up was reported as negative, presented again several years later with headache, nausea, and vomiting. She was found to have a large posterior fossa lesion on repeat neuroimaging that was retrospectively seen on the first neuroimaging scan as prominent enhancement in the region of the fourth ventricular choroid plexus. The second patient presented with congenital nystagmus and a lingual tremor and was found to have a slowly growing lesion situated in the fourth ventricle. Initial imaging was read as nodularly enhancing tela choroidea, but subsequent scans revealed enlargement of the lesion.**

RESULTS: **The first patient underwent gross total resection, and neuropathology was consistent with an atypical teratoid rhabdoid tumor. The patient has done well with postoperative adjuvant therapies. In the second patient, resection of the lesion revealed ependymoma; the patient has done well after adjuvant radiation therapy.**

CONCLUSIONS: **Pediatric patients who have enhancing tela choroidea or choroid plexus without an obvious mass lesion of the fourth ventricle may harbor early tumors. Surveillance imaging in these patients may be warranted given the aggressive nature of certain posterior fossa tumors in children. Failure to recognize abnormal enhancement patterns in this region may lead to delayed diagnosis.**

INTRODUCTION

The tela choroidea is a connective tissue layer that separates the pia mater from the ependymal lining of the ventricular system. During embryologic development, ependymal cells of the lateral, third, and fourth ventricles invaginate to form longitudinal folds; the tela choroidea invaginates along with these cells and acts as a richly vascularized lamina propria. These ependymal cells eventually differentiate into the choroid plexus of the ventricular system. The tela choroidea forms the roof of the inferior fourth ventricle (10). The choroid plexus that it supports can extend into the foramen of Magendie and Luschka and covers the lateral recess of the fourth ventricle (5, 8).

The genetic development of the neuronal and ependymal lining of the brain is complex. Some studies showed that the same genes that are involved in neuronal development and differentiation may also be involved in congenital nystagmus (1). Other studies showed that the cells of the ependymal lining have stem cell–like properties (9). It is plausible that genetic alterations may lead to aberrant growth and tumor formation, in addition to neurologic signs or symptoms that may or may not be related to a tumor.

We present 2 cases of patients with congenital nystagmus who developed posterior fossa brain tumors that manifested as enhancement of the tela choroidea and choroid plexus of the fourth ventricle on initial imaging without an obvious mass lesion. One patient was found to have an atypical teratoid/rhabdoid tumor 2 years after initial imaging was read as negative, and the other was found to harbor an ependymoma.

CASE 1

History

A 5-year-old girl with no family history of congenital nystagmus or intracranial mass lesions presented at 1 month of age with abnormal eye movements. An ultrasound scan of the head was done in infancy, which revealed optic nerve hypoplasia and thickening of the corpus callosum. She was lost to follow-up and presented again at 15 months of age with nystagmus and septo-optic dysplasia. This work-up was ultimately negative for intracranial mass lesions, but initial imaging showed striking enhancement of the fourth ventricular

tela choroidea. The patient presented again to an outside facility 2 years later with headache, nausea, and vomiting. Imaging revealed a large mass in the posterior fossa, and she was transferred to our institution for management.

Examination

During the initial evaluation, the patient had a normal neurologic examination other than strabismus and horizontal nystagmus. The patient was transferred emergently from an outside facility 2 years after the initial evaluation; she was noted to be stuporous but localizing to noxious stimuli. At this time, her pupils were equal and reactive to light with no obvious sunsetting. On arrival to our facility, she was intubated for airway protection and external ventricular drain placement.

Neuroimaging

Initial magnetic resonance imaging (MRI) during work-up for septo-optic dysplasia was read as optic nerve hypoplasia with no evidence of intracranial masses (Figure 1, top 2 images), but did show striking enhancement of the tela choroidea of the fourth ventricle. Additional MRI sequences (i.e., fluid attenuated inversion recovery, T2) did not show abnormalities. A head computed tomography scan obtained at an outside facility 2 years later when she presented with nausea and vomiting showed a 4 cm × 3 cm mass in the posterior fossa with obstructive hydrocephalus. Repeat MRI revealed a large, heterogeneously enhancing mass situated in the roof of the fourth ventricle (Figure 1, bottom 2 images).

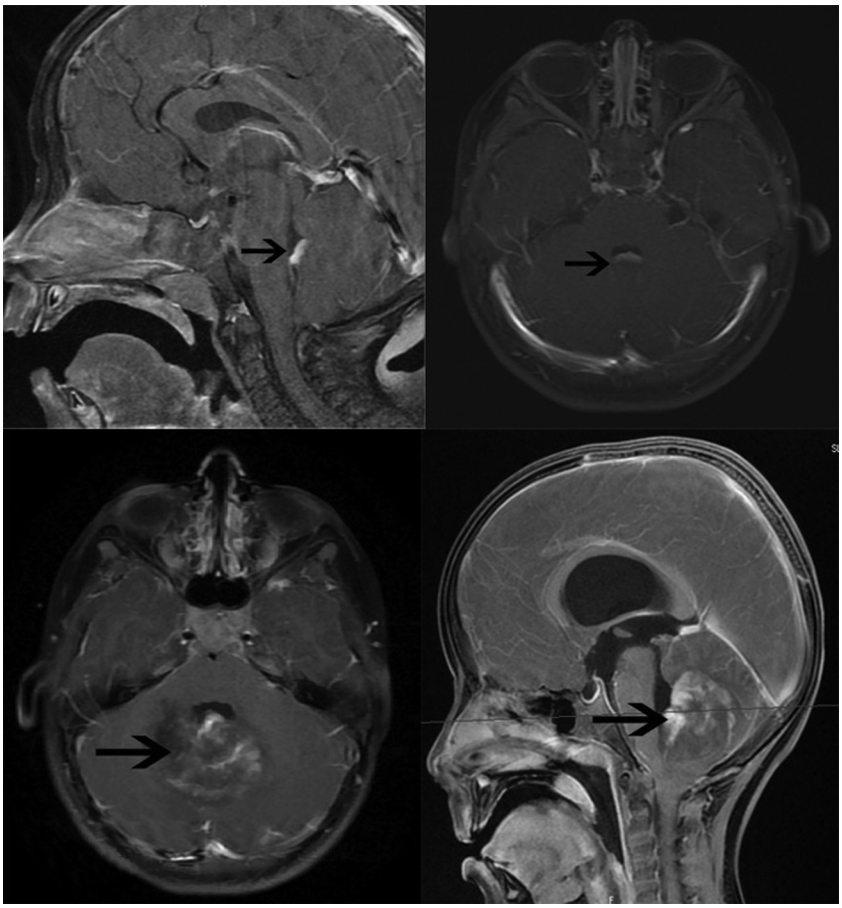

Figure 1. Case 1. Axial and sagittal T1-weighted magnetic resonance images with contrast showing strikingly enhancing choroid plexus and tela choroidea of the fourth ventricle but no obvious mass lesion (*arrows*; top 2 images). Repeat magnetic resonance imaging scan revealing a large heterogeneously enhancing lesion in the posterior fossa (*arrows*; bottom 2 images).

Treatment

The patient underwent gross total resection of an atypical teratoid/rhabdoid tumor followed by adjuvant radiation and chemotherapy. The radiation dose was 5040 cGy. Chemotherapy included vincristine, methotrexate, cyclophosphamide, etoposide, and cisplatin. Autologous stem cell rescue therapy was used after adjuvant therapy.

Postoperative Course

Postoperatively the patient developed cerebellar mutism that resolved slowly. Other than her mutism, her immediate postoperative course was uncomplicated. At follow-up evaluation 1 year after surgery, she was noted to have hypophonic, dysarthric, and ataxic speech, which was improved substantially from her postoperative mutism. She was able to ambulate independently, but showed signs of ataxic gait. She had mild dysmetria of the right upper extremity. Her extraocular movements were intact, although lateral gaze nystagmus was noted.

CASE 2

History

A 19-year-old woman with no family history of congenital nystagmus or intracranial mass lesions presented during infancy with congenital nystagmus and hypotonia. Extensive work-up at that time revealed no abnormalities other than some nodular enhancement of the fourth ventricular tela choroidea. She presented again at age 12 with symptoms of a postural extremity tremor, which was followed by intermittent episodes of urinary incontinence. Work-up at that time did not reveal any abnormalities. These symptoms were followed by the onset of a lingual tremor roughly 2 years later. Further work-up at this time revealed the interval development of a 6-mm enhancing lesion centered in the inferior fourth ventricle.

Examination

On neurologic examination, the patient was noted to have oscillatory nystagmus with primary gaze that abated with convergence, a lingual tremor with extrusion that abated with lateral movements of the tongue, and intention tremors of the upper and lower extremities. She was able