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Pilocytic Astrocytoma

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A 12-year-old girl presented to our pediatric clinic for a drop-in visit with intermittent headaches of 1 week's duration. She described the headaches as a dull, moderate, nonradiating pain localized to the right frontal region. She reported that the headaches occurred particularly in the afternoon and with no aggravating factors. The head pain had been associated with 1 episode of nonbilious nonbloody (NBNB) emesis. The pain was relieved with medications such as nonsteroidal anti-inflammatory drugs.

History. The patient denied any trauma, fever, auras, night sweats, weight loss, visual changes, anorexia, extremity weakness and numbness, and recent illness. She had no significant medical history. She reported having stress and anxiety related to her school and home life.

Physical examination. Physical examination findings were unremarkable during her clinic visit. Her pupils were equal, round, and reactive to light and accommodation, with intact extraocular movements. No lymphadenopathy was present. The patient was awake, alert, and

oriented to person, place, and time. Sensation was intact, and she had normal motor function with no focal neurological deficits. Function of cranial nerves II through XII was intact. She had normal deep-tendon reflexes and normal gait.

A DASS-21 (Depression Anxiety Stress Scales) screening tool was administered, the results of which indicated mild depression, severe anxiety, and moderate stress. Results of a SCARED (Screen for Child Anxiety-Related Emotional Disorder) screening tool were positive for social anxiety disorder, significant school avoidance, and separation anxiety.

She was advised to keep a symptom diary documenting her headaches and to follow up in the clinic in 1 week.

The patient did not follow up in 1 week as directed, since her headaches had improved, and she was busy at school. Nevertheless, 13 days after the initial encounter, she experienced a second episode of NBNB emesis. Our clinic was closed, and the patient instead visited a local emergency department (ED).

Diagnostic tests. A computed tomography (CT) scan of the head without contrast was ordered in the ED. The results showed a large cystic mass in the right cerebellum with minimal hemorrhage and calcification, resulting in obstructive hydrocephalus (**Figure 1**).

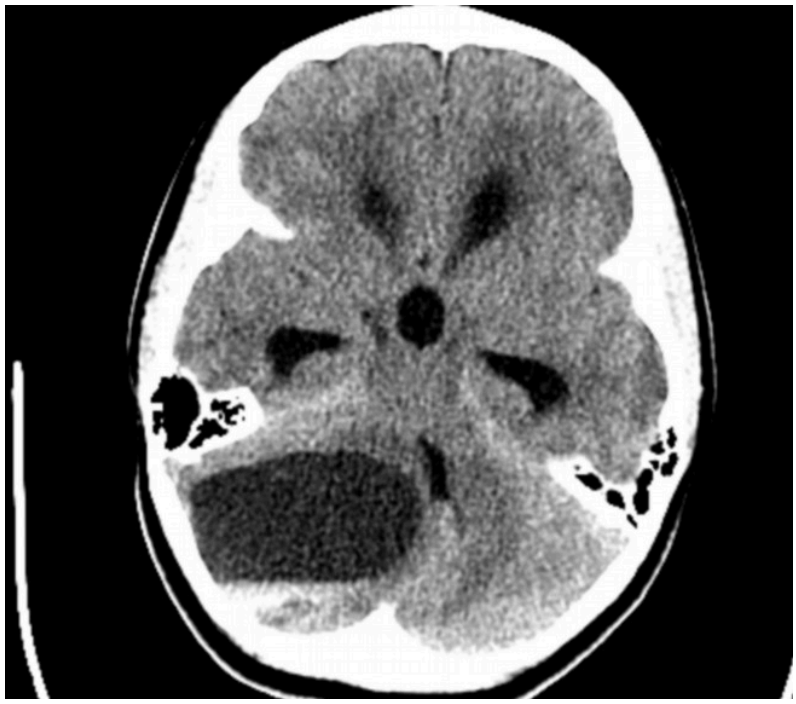


Figure 1. *Noncontrast CT scan of the head showed a large cystic mass in the right cerebellum measuring 5.5 × 3.5 cm on an axial image.*

Subsequent magnetic resonance imaging (MRI) of the brain without contrast showed a 5.9-cm cystic right cerebellar intra-axial mass with enhancing nodule, consistent with pilocytic astrocytoma, with spontaneous hemorrhage within the cyst and marked tonsillar herniation and compression of the fourth ventricle (**Figure 2)**.

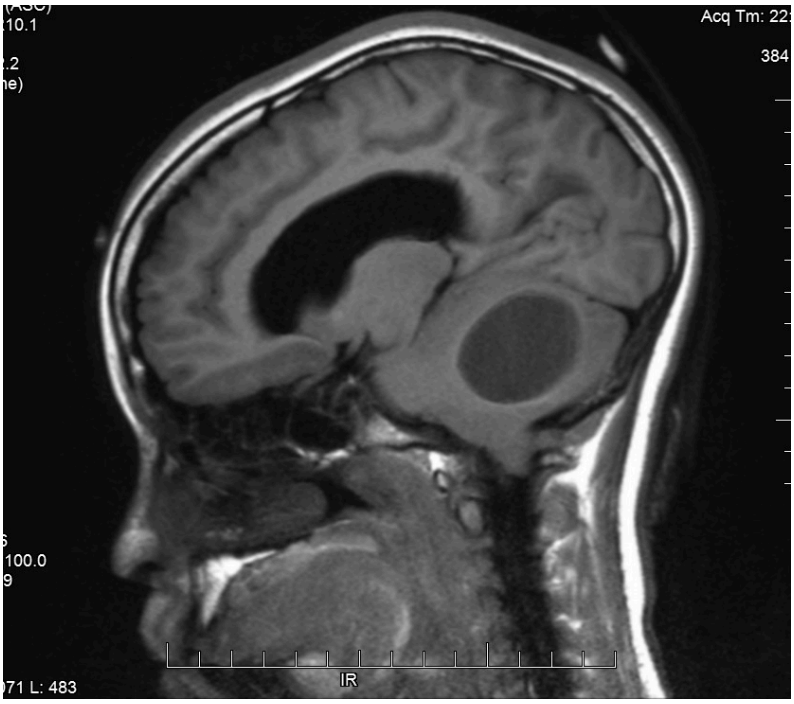
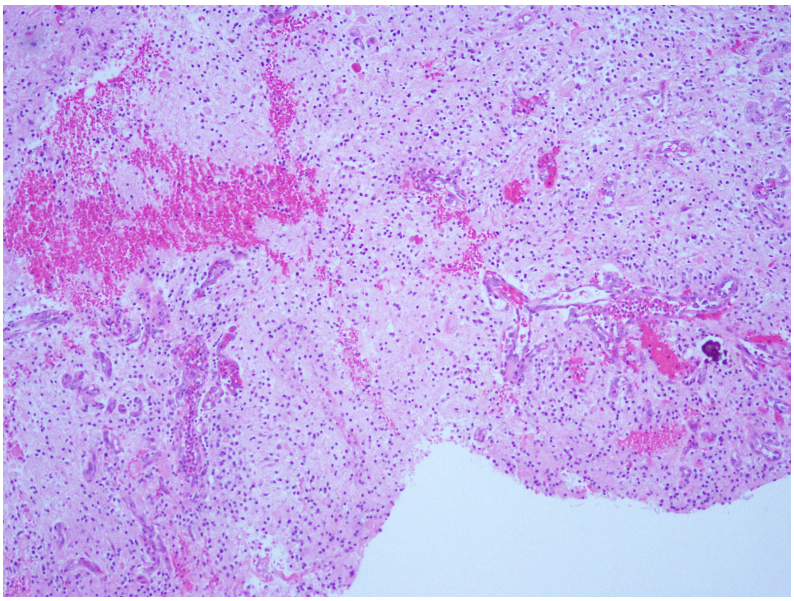
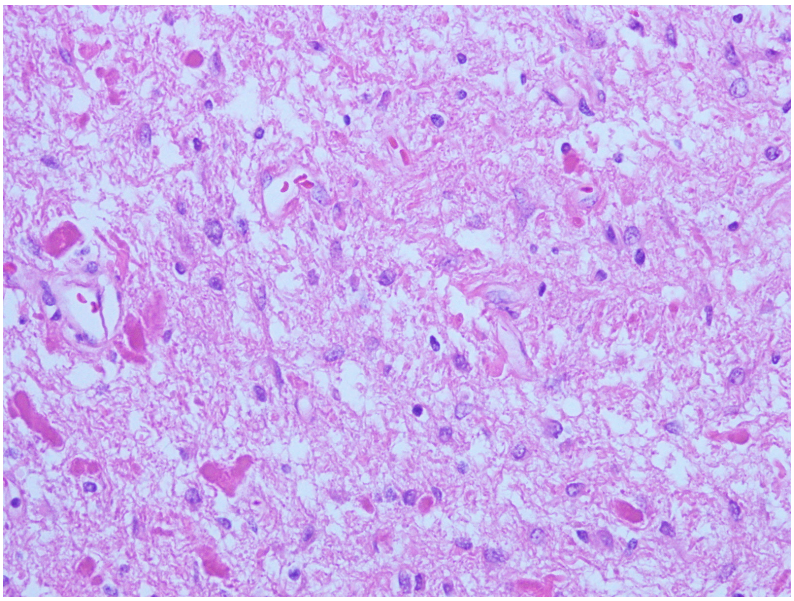


Figure 2. *MRI of the brain with contrast showed 4.7-cm (anteroposterior) × 5.9-cm (transverse) × 3.5-cm (craniocaudal) intracranial mass. Severe compression of fourth ventricle is resulting in acute hydrocephalus and tonsillar herniation of 12 mm through the foramen magnum.*

Posterior fossa craniotomy was performed to remove the right cerebellar tumor, along with stereotactic right occipital external ventricular drain placement for obstructive hydrocephalus. Pathological slides confirmed pilocytic astrocytoma, World Health Organization (WHO) grade 1, with low (1%) proliferation index (**Figures 3 and 4**).





Figures 3 and 4. *Tumor cells surrounding blood vessels with areas of necrosis and densely eosinophilic Rosenthal fibers (Frozen section, high-power field).*

Outcome of the case. This patient was managed surgically; no chemotherapy or radiation was required. Postoperatively, the patient did not display any neurological defects. At 2- and 4-week follow-up visits, the patient reported having mild neck pain and denied having any symptoms of anxiety. Her neck pain had completely resolved by the sixth week postoperatively.

Discussion. Pilocytic astrocytoma is the most frequent relatively benign (WHO grade 1) pediatric brain tumor. It is mainly caused by a mutation in *BRAF*.[1] It accounts for 15.4% of primary brain and central nervous system (CNS) tumors among persons younger than 19 years old and 18.6% of those younger than 14 years old.[2] The most common site is the cerebellum.[3] The treatment of choice is total surgical resection, and irradiation is typically not required. Patients with pilocytic astrocytoma have a long-term survival rate of 80% to 95%.[3]

The clinical presentation of a brain tumor varies due to the location of the tumor and is insidious due to the slow growth of the tumor. Cerebellar tumors greatly affect gait and motor and cranial nerves. If the tumor is present at the hypothalamus, endocrinologic symptoms may result. In addition, due to the mass effect resulting in an increase in intracranial pressure, headaches, nausea, and vomiting are the most common symptoms. In rare cases, brain tumors can present primarily with psychiatric symptoms.[4]

In 1938, Keschner and colleagues reported that 78% of 530 adult patients with primary brain tumor indeed had psychiatric symptoms, and 18% of the 530 patients solely had psychiatric symptoms.[5] A 2003 study in Finland demonstrated that anxiety resulted from mass effect rather than the psychological reaction to the illness.[6] The findings showed that patients with a tumor in the right hemisphere had a significantly higher mean anxiety score than patients with a tumor in the left hemisphere. The level of anxiety declined postoperatively in patients with tumors in the right hemisphere; a decline was not found in patients with a tumor in the left

hemisphere. In 2012, a case report by Assefa and colleagues discussed a 26-year-old man with left temporal lobe meningioma who displayed symptoms of anxiety and fear due to compression of the left side of the amygdala.[7] Although the amygdala is commonly implicated in the pathophysiology of anxiety disorders, emerging studies are supporting the emotional regulation functions of the cerebellum.

The primary role of the cerebellum traditionally has thought to be balance and motor control. The cortico-ponto-cerebellar and cerebello-thalamo-cortical pathways are responsible for emotional processes due to information processing between the cerebellum and cortical areas.[8] Although the exact neural mechanism underlying the etiology of anxiety disorder is unclear, it has been discovered that anxiety disorder resulted in a hyperactive cerebellum.[9] Therefore, a mass in the cerebellum could potentially result in an anxiety disorder.

Summary. Clinicians must be aware of the red flags associated with an intracranial mass, especially in the pediatric population. Although morning headaches accompanied by vomiting are the most common presentation, these may not always be the first clinical sign of a brain tumor. Our patient did not exhibit the motor and balance dysfunction as would be expected with such a large cerebellar mass. Rather, she displayed significant anxiety as demonstrated by the results of the DASS-21 and SCARED screening tools. Although the cerebellum deeply influences thoughts and emotions, no studies have demonstrated a direct correlation between a cerebellar tumor and anxiety. The neural mechanism of anxiety is quite complex and is still unclear; however, any new psychiatric change should alert the clinician to a potential organic cause and prompt further investigation.

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