HEMORRHAGIC CEREBELLAR PILOCYTIC ASTROCYTOMA IN A FILIPINO ADOLESCENT WITH SUDDEN ONSET "WORST HEADACHE OF HER LIFE"

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BACKGROUND

Non-traumatic, spontaneous cerebellar hemorrhages in pediatric patients are rare. Most are from vascular malformations or hematologic abnormalities. Brain tumors are also uncommon, but is the second most common malignancy in this age group. Brain tumors in children and adolescents do not usually present with acute symptoms and hemorrhagic onset and rarely associated with low grade tumors.

CASE REPORT

The patient is a 16-year-old Filipino healthy female who presented with **sudden onset severe headache**, which she has described as "**the worst headache" of her life**, associated with **dizziness and vomiting**. Her mother died of intracranial hemorrhage from an aneurysm. There was no family history of neurofibromatosis. On examination, she had **horizontal nystagmus (right>left).** No dysconjugate gaze, papilledema, dysmetria, dysdiadokokinesia, and ataxia. No neurocutaneous lesions were seen. CT scan revealed **intraventricular hemorrhage** within the fourth ventricle, resulting in obstructive hydrocephalus. ***No aneurysm or vascular malformations*** *was seen* on CT angiogram. Cranial MRI showed hemorrhage at the medial aspect of the right cerebellar peduncle, and a cavernous malformation was suspected. She underwent suboccipital craniotomy and gross total excision of the mass. Intraoperative findings showed a **yellowish-brown 4th ventricular tumor with old hemorrhages**. Histopathology and immunohistochemistry results supported the diagnosis of a **pilocytic astrocytoma** (GFAP+, S100+, P16 strong nuclear staining in 10-20%, Ki67 low, <3%, IDH1 and Inhibin negative). She underwent emergency VPS insertion due to progressing hydrocephalus post-excision and recovered well without any residuals. Monitoring at 3, 6 and 12 months post-operatively revealed <0.5cm residual and no recurrence.

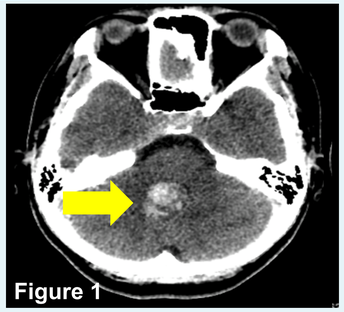

Figure 1

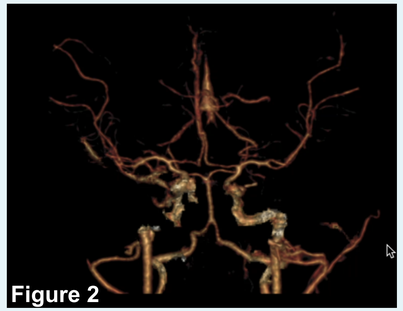

Figure 2

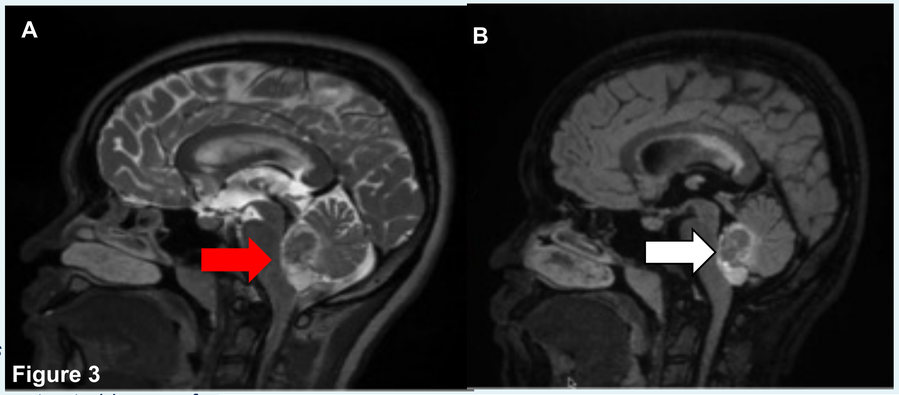

Figure 3

Figure 1. *Plain axial CT scan image of the posterior fossa at the level of the pons taken 4 hours after the onset of severe headache showing acute intraventricular hemorrhage (yellow arrow) with an estimated volume of 10.5cc).* ***Figure 2.*** *Reconstructed image of the cerebral vasculature via CT angiogram taken 4 days post ictus showing normal vasculature.* ***Figure 3.*** *Sagittal views showing T2-* ***(3A)*** *and T1-weighted MRI images* ***(3B)*** *showing subacute hemorrhage at the fourth ventricle and medial aspect of the right cerebellar hemisphere of approximately 7.9cc. On both T1- (white arrow) and T2W imaging (red arrow), the lesion shows heterogenous hyperintensity. The lesion is peripherally enhancing, with diffuse magnetic susceptibility artifacts and areas of restricted diffusion (not shown).*

DISCUSSION

Incidence rate of spontaneous intracranial hemorrhages is 8% in adult and pediatric patients with pilocytic astrocytoma [1] There are only **27 documented cases** of pediatric pilocytic astrocytomas presenting with acute hemorrhage from 1977 to present, including this case: **56% (n=15)** are located at the **cerebellum**; 1.3:1 ratio of female:male, 40% between 10-14 years old; 53% presented acutely with headache and vomiting; **80% survived with good outcomes**; 3 are mortalities; only 1 other case (Frassinato 2009) presented with sudden onset severe headache similar to our patient.

Hemorrhagic lesions **not related to aggressive pathology** (no difference between hemorrhagic and non-hemorrhagic pilocytic astrocytomas in Ki67 labeling index or microvascular proliferation)[2] **FGFR1 mutations are associated** with hemorrhagic potential of low grade gliomas.[3] One study showed spontaneous bleeding in pilocytic astrocytoma is significantly associated with tumor VEGF expression.[4] Initial neuroimaging studies may only show hemorrhage and not immediately reveal a tumor; additional radiologic techniques such as perfusion-weighted imaging may be warranted if available.

CONCLUSION

This is an atypical presentation of pilocytic astrocytoma in the pediatric age group, which usually presents with a more protracted course. Although vascular anomalies, trauma, and hematologic abnormalities are the most common etiologies of spontaneous cerebellar hemorrhage, an underlying benign intracranial tumor such as pilocytic astrocytoma should be entertained as one of the differentials. Appropriate neuroradiological investigations, close inspection of the lesion intraoperatively, and complete histopathologic examination are important to establish the diagnosis in unusual clinical presentations of brain tumors in children.

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