Juvenile pilocytic astrocytoma presenting with subarachnoid hemorrhage

Case report and review of the literature

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The authors report a case of a 13-year-old boy with juvenile pilocytic astrocytoma (JPA) presenting with subarachnoid hemorrhage (SAH). The patient experienced sudden onset of headache, vomiting, and loss of consciousness. Cranial computerized tomography scanning revealed blood within basal cisterns and the third ventricle. Angiography demonstrated normal cerebral vasculature and upward displacement of the bilateral A1 segments of the anterior cerebral artery. Magnetic resonance (MR) imaging revealed a chiasmatic/hypothalamic mass with evidence of hemorrhage. The mass was surgically decompressed. Histopathological examination showed evidence of JPA. In all cases of SAH in which there is blood around the third ventricle and a raised A1 segment on angiography, MR imaging should be performed. The presence of a normal sella turcica, as well as indistinct margins between the tumor and the opticocarotid apparatus should raise suspicion about the lesion.

KEY WORDS • pilocytic astrocytoma • tumor • subarachnoid hemorrhage

PILOCYTIC astrocytoma is a very rare cause of tumor-related SAH and has been previously reported in only six cases.1,3,4,6,10,16 Brain tumors have been reported to be responsible for 3 to 7% of SAH or intracerebral hematoma in the pediatric population.12,13,15 Medulloblastomas and other primitive neuroectodermal tumors are the most common source. In cases of supratentorial pilocytic astrocytoma arising from the opticocarotid/hypothalamic axis, patients usually present with visual impairment, endocrine abnormalities, and diencephalic syndrome.2 Hemorrhage in cases of JPA is very rare.1,3,4,6,10,16 We describe the case of a 13-year-old boy who presented with SAH due to a chiasmatic/hypothalamic pilocytic astrocytoma.

Case Report

History. This 13-year-old boy developed sudden onset of headache, vomiting, and loss of consciousness. He was immediately taken to a local hospital where cranial CT scanning demonstrated blood in the basal cisterns and the third ventricle (Fig. 1). He regained consciousness after 6 hours and was referred to our institution for further treatment.

Presentation and Examination. We learned that the patient had a 2-month history of episodic headaches. On examination, he was alert and his higher mental function status was normal. He had neck rigidity and a positive Kernig sign was evident. Visual acuity was 6/36 on the left and 6/6 on the right side. Bilateral fundi were normal. He had right temporal and left nasal field cuts. The cranial nerve’s functions were intact. There was no history of polyuria or endocrinopathy. Routine hematological workup demonstrated normal levels, and the coagulation profile was normal as well.

Angiography revealed upward displacement of the A1 segments of the ACA bilaterally (Fig. 2). No associated aneurysm or vascular malformation was seen. Cranial MR imaging revealed a large suprasellar mass in the chiasmatic/hypothalamic region with evidence of hemorrhage (Fig. 3A and B). After contrast administration the mass showed peripheral enhancement (Fig. 3C and D). The pituitary gland was seen to be separate from the mass.

Operation. A left pterional craniotomy was performed. The tumor was approached via intraoptic and caroticooptic spaces. The subarachnoid space was filled with blood. We observed a grayish mass that was soft and easily suctioned. Large blood clots were seen within the substance...
of tumor. A near-total excision of the tumor was achieved. The portion of tumor close to the hypothalamus was left in place because it was imperceptibly merged with these structures.

Postoperative Course. Histopathological examination showed evidence of JPA (Fig. 4A). The postoperative period was uneventful. Follow-up evaluation at 7 months demonstrated minimal residual tumor in the right half of the chiasm (Fig. 4B). The patient’s vision improved, and there were no visual field defects. The patient is being followed without radiotherapy.

Discussion

Between 3.6 and 10% of patients with brain tumors in the pediatric population initially present with SAH. Yokota, et al., reported an equal incidence of aneurysm rupture and tumor hemorrhage in pediatric patients.

Subarachnoid hemorrhage associated with brain tumors usually develops secondary to intratumoral hemorrhage. The tumors, which are located along the cerebrospinal fluid pathway near the ventricles or the subarachnoid space, are more likely to cause SAH than intracerebral hematoma when they bleed. The most common lesions accounting for SAH are pituitary tumors, glioblastomas, metastatic tumors, medulloblastomas, and primitive neuroectodermal tumors; however, in pediatric patients, brain tumor–induced hemorrhages occur predominantly in the posterior fossa, with the medulloblastoma being the most common source. The incidence is 8.3% in medul-
loblastomas, 5.9% in ependymomas, and 1.9% in astrocytomas. Pediatric brain tumors have been found to bleed more frequently than those in adults (5.3 and 2.3%, respectively), which has been attributed to the higher frequency of malignant and large tumors in children.

Intracranial tumor–related bleeding may result from the following: endothelial proliferation and obstruction of the tumor vessels causing tumor necrosis and hemorrhage, disruption of tumor vessels by expanding tumor, direct tumor infiltration into vessels, or abnormal tumor vascularity.9 The pathogenesis of spontaneous hemorrhage is related to several factors. Tumor-associated aneurysm is also a possible cause.5

Hemorrhage in JPA is, however, very rarely described and its occurrence has been observed in the optic nerve and hypothalamus (Table 1). Glew9 has reported the case of a 30-year-old man who presented with acute bilateral loss of vision simulating pituitary apoplexy. The authors described a hypothalamic tumor with a hemorrhagic cyst found intraoperatively, which was a well-differentiated Grade I fibrillary astrocytoma. Hwang, et al.,6 reported on a 34-year-old man presenting with features of SAH, and a hemorrhagic juvenile astrocytoma was resected. Similarly, Golash, et al.,4 removed a hypothalamic astrocytoma in a 13-year-old girl who presented with SAH. Hemorrhage in the JPAs can be attributed to vascular proliferation, which is an occasional feature of these tumors.8,10 Retiform capillaries have been found to be associated with hemorrhage in astrocytomas and glioblastoma multiforme.8 These capillaries take a convoluted, tortuous route while lacking external support.

Although a favorable recovery from the initial catastrophic condition can be achieved, the ultimate prognosis in the majority of cases remains poor because such hemorrhages usually develop secondary to malignant tumors.

Finally, pilocytic astrocytoma, although a rare cause of tumor-associated SAH around the opticocochiasmatic axis (the most common being pituitary adenoma), should be considered in the differential diagnosis. In all cases of SAH in which blood is observed in the third ventricle, the A1 segment should be carefully examined for any upward displacement. Small tumors are often missed on routine CT scans; thus, MR images should be obtained when any suspicion exists as to diagnosis. The presence of a normal

**FIG. 3.** Magnetic resonance images. A and B: Sagittal T1 (A) and T2-weighted (B) images revealing a mass in the chiasmatic/hypothalamic region with evidence of hemorrhage, separate from pituitary gland. C and D: After contrast administration, the mass showed peripheral enhancement on sagittal (C) and coronal (D) images.
sella turcica, indistinct margin between the tumor and opticochiasmatic apparatus, and presence of cystic changes within the tumors should raise suspicion of the lesion. Surgery should always be performed via the transcranial route. Evacuation of the hematoma in conjunction with conservative decompression of the tumor is advised. When the tumor is located along the optic nerve and chiasm, it should be left in place to avoid causing any damage.

In conclusion, tumor-related hemorrhage is an important cause of SAH in the pediatric population. Although rare, JPA may be a cause of acute SAH.

References
4. Golash A, Thorne J, West CG: Low grade pilocytic astrocyto-

Table 1

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Tumor Location</th>
<th>Symptoms</th>
<th>Neurodiagnostic Findings</th>
<th>Operative Details</th>
<th>Follow-Up Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glew, 1977</td>
<td>30, M</td>
<td>hypothalamus</td>
<td>rapid loss of vision, headache, seizures</td>
<td>intraorbital mass in right orbit displacing 3rd ventricle</td>
<td>biopsy</td>
<td>more obtunded &amp; died after 2 wk</td>
</tr>
<tr>
<td>Yanoff, et al., 1978</td>
<td>26, F</td>
<td>optic nerve</td>
<td>rapid proptosis &amp; loss of vision</td>
<td>SAH &amp; intraventricular hemorrhage; enhanced suprasellar mass on CT, elevation of proximal ACAs, &amp; fat displacement; CTA on angiography</td>
<td>total excision of cystic mass with evacuation ofSAH</td>
<td>NS</td>
</tr>
<tr>
<td>Charles, et al., 1981</td>
<td>45, M</td>
<td>hypothalamus</td>
<td>sudden-onset severe headache, nausea, vomiting &amp; loss of consciousness</td>
<td>hypervascular mass on MR; &amp; ventriculography</td>
<td>partial excision of tumor via right pterional approach; residual tumor removed via orbitozygomatic approach</td>
<td>no residual mass on 2 mos follow-up</td>
</tr>
<tr>
<td>Matsumoto, et al., 1997</td>
<td>13, F</td>
<td>hypothalamus</td>
<td>sudden-onset headache, nausea, vomiting &amp; loss of consciousness</td>
<td>CTA on angiography</td>
<td>near-total excision of hypothalamic astrocytoma; no further tumor growth</td>
<td>partial improvement in vision, no field defect</td>
</tr>
<tr>
<td>Golash, et al., 1998</td>
<td>13, M</td>
<td>hypothalamus</td>
<td>sudden-onset headache, nausea, vomiting, &amp; loss of consciousness</td>
<td>MR of hypothalamic astrocytoma; confirmation of bleed, frank SAH</td>
<td>MRA; residual mass in right half of chiasm</td>
<td>no residual mass at 11 mos</td>
</tr>
<tr>
<td>present case</td>
<td>13, M</td>
<td>hypothalamus, optic chiasm</td>
<td>sudden-onset headache, nausea, vomiting &amp; loss of consciousness</td>
<td>MRA; residual mass in right half of chiasm</td>
<td>near-total excision of hypothalamic astrocytoma; confirmation of bleed, frank SAH</td>
<td>no residual mass at 11 mos</td>
</tr>
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* ACA = anterior cerebral artery; CA = carotid artery; NS = not stated.
† Details not available.

Fig. 4. A: Photomicrograph showing diffuse proliferation of astrocytic cells with microcystic spaces in the background suggestive of pilocytic astrocytoma. H & E, original magnification × 200. B and C: Postoperative MR images revealing minimal residual tumor.
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