Cerebellar Pilocytic Astrocytomas with Spontaneous Intratumoral Hemorrhage in Adult

Min-Su Kim, M.D., Sang Woo Kim, M.D., Chul-Hoon Chang, M.D., Oh-Lyong Kim, M.D.

Department of Neurosurgery, Yewungnam University College of Medicine, Daegu, Korea

INTRODUCTION

A pilocytic astrocytoma (PA), previously known as spongioblastomas, is a slow-growing benign tumor that occurs most commonly in children and young adults. The annual incidence for all ages has been reported to be 2.9-4.8 cases per million people, and no gender predilection has been shown. Among child patients, up to 67% of them present with cerebellar lesions, whereas among adult patients, about 55% present with supratentorial lesions. The rate of intracranial hemorrhages from gliomas is reported to be 3.7-7.2%. Most glial tumors that have a propensity to present with hemorrhages include high-grade astrocytomas, oligodendrogliomas, and mixed oligoastrocytomas. However, in low-grade glial tumors such as PAs, intratumoral hemorrhages are very rare. Seven patients with cerebellar PAs associated with hemorrhage were reported and all these cases were under 15 years of age. We could not find any reports in the medical literature on hemorrhagic cerebellar PAs. We report two adult cases of cerebellar pilocytic astrocytomas with spontaneous intratumoral hemorrhage.

CASE REPORT

Case 1
A 37-year-old woman presented with sudden onset of severe headache, nausea, and vomiting. Upon neurological examination, there were no signs of cerebellar dysfunction, including ataxia, dysmetria, and dysdiadochokinesis. Computed tomography demonstrated an acute hemorrhage adjacent to the right cerebellar hemisphere and hydrocephalus. Magnetic resonance imaging (MRI) revealed a cerebellar vermian tumor with the hemorrhage as a mixed iso-intense area in the T2-weighted image, and as a mixed hyperintense area in the contrast-enhanced T1-weighted image. The second case is a 53-year-old man presenting with headache for 3 weeks. MRI revealed a cerebellar hemispheric tumor with the hemorrhage as a mixed hyperintense area. It had a cystic mass with a heterogeneous enhanced mural nodule in the gadolinium-enhanced T1-weighted image and a fluid-fluid level within the cyst in the T2-weighted image. Both of them underwent radical resections of their respective lesions. Histological examination of the specimens revealed typical astrocytoma, including a hemorrhagic portion. Both patients recovered postoperatively and continue to do well at present. The medical literature on hemorrhagic cerebellar PAs is also reviewed.

Key Words: Pilocytic astrocytoma · Hemorrhage · Cerebellum · Adult.
Cerebellar pilocytic astrocytomas with symptomatic hemorrhage

Table 1. Summary of reported cases of cerebellar pilocytic astrocytomas with symptomatic hemorrhage

| Author (year)         | Age (years)/Sex | Location | Type of hemorrhage | Histology                  | Outcome |
|-----------------------|-----------------|----------|--------------------|-----------------------------|---------|
| Mauersberger et al. (1977) | 10/M            | Cerebellum | ITB                | Spongioblastoma             | Alive   |
|                       | 10/F            | Cerebellum | ITB                | Spongioblastoma             | Dead    |
| Vincent et al. (1980) | 14/F            | Cerebellum | ITB                | Pilocytic                  | Alive   |
| Fogelson et al. (1980) | 9/M             | Cerebellum | ITB                | Pilocytic                  | Alive   |
| Specht et al. (1986)  | 8/M             | Cerebellum | ITB                | Mixed pilocytic/Oligodendroglioma | Dead    |
| Mesiwala et al. (2001) | 13/M            | Cerebellum | ITB                | Pilocytic                  | Alive   |
| Lee et al. (2009)     | 15 months/M     | Cerebellum | ITB, SAH, SDH      | Pilocytic                  | Alive   |
| Present cases         | 37/F            | Cerebellum | ITB                | Pilocytic                  | Alive   |
|                       | 53/M            | Cerebellum | ITB                | Pilocytic                  | Alive   |

ITB: intratumoral hemorrhage, SAH: subarachnoid hemorrhage, SDH: subdural hemorrhage

We could see the tumor beneath the right cerebellar tonsil. The tumor was grayish-colored, relatively firm and well-demarcated from the surrounding normal tissue with intratumoral hematomas. We were able to perform the en-bloc removal easily and completely.

The surgery was successful without neurological deficits. Histopathological examination of the specimen revealed a typical PA featuring astrocytic tumor cells with numerous Rosenthal fibers and hyalinized blood vessels (Fig. 2A). Bipolar cells with long hair-like glial fibrillary acidic protein-positive processes were observed after using an immunohistochemical staining technique (Fig. 2B). Occult coexisting vascular malformations were not observed. Three months after the operation, MRI demonstrated no residual or recurrent lesions (Fig. 1E). The patient recovered postoperatively and continues to do well.

Case 2

A 53-year-old man presented with a three-week headache. Upon neurological examination, there were no signs of cerebellar dysfunction. Brain CT demonstrated a barely high-density mass within the cystic portion of the left cerebellar hemisphere. Brain MRI revealed a left cerebellar hemispheric cystic mass with fluid-blood level in the axial T2-weighted image (Fig. 3A) and a heterogeneously enhanced mural nodule in the axial gadolinium-enhanced T1-weighted image (Fig. 3B). There was no evidence of vascular lesions from MRI. We performed the operation using a retrosigmoid suboccipital craniotomy under general anesthesia. We could see the tumor beneath the right cerebellar tonsil. The tumor was grayish-colored, relatively firm and well-demarcated from the surrounding normal tissue with intratumoral hemorrhoma. We were able to perform radical resection of the enhanced tumor with a hematoma. Histopathological examination of the specimens revealed hemorrhages and compact portions consisting of astro-
cystic tumor cells (Fig. 4A). Neoplastic astrocytes with eosinophilic granular bodies and Rosenthal fibers in the glial fibrillary background were visible (Fig. 4B). Three months after the operation, MRI demonstrated no residual or recurrent lesions (Fig. 3C). The patient recovered postoperatively and continues to do well.

**DISCUSSION**

A cerebellar PA is well demarcated, often having a cyst with a mural nodule, and is usually located in the cerebellar hemisphere. The presenting symptoms of these tumors are commonly headache, vomiting, mental change, gait ataxia, tremors, and dysmetria. Usually, these symptoms develop insidiously. White et al.\(^2\) recently reported that the rate of spontaneous hemorrhages in histologically proven cases of PAs was 8%, but no hemorrhages occurred within the cerebellum.

The association between age and tumor location is strong. Most cerebral hemorrhagic PAs occur in adults, whereas cerebellar hemorrhagic PAs occur mostly in children\(^\)\(^1\). The symptomatic hemorrhages associated with cerebellar PAs are extremely rare (Table 1\(^6,10,15-17,19\)). The mechanism of high-grade gliomas is likely related to necrosis of blood vessels, as well as to abundant neovascular proliferation. However, the mechanism of hemorrhage in low-grade gliomas is not well known and has been a subject of debate. Lyons\(^14\) reported that in an elderly adults, the potential microscopic infiltration of PAs into the adjacent parenchyma might influence the likelihood of spontaneous hemorrhage in the setting of adjacent focal congophilic angiopathy and anticoagulation. Van Querkerk and Dirven\(^18\) reported that in a child, a medullary PA including oligodendrogliotic parts manifested as acute onset of symptoms caused by the intratumoral hemorrhage. They suggested that the hemorrhage might have originated from abnormal fragile vasculatures in oligodendrogliotic parts of the tumor or in occult coexisting vascular malformations.

Endothelial proliferation, rupture of an encased aneurysm, and dysplastic capillary beds have been factors hypothesized as potential causes for bleeding\(^1,13\). In our cases, the patients had normal platelet-cell counts, prothrombin time, partial thromboplastin time, and no history of hypertension or coagulation defects related to systemic disease, chemotherapy, or radiation.

The cause of hemorrhage in our cases is unclear, but we assume that hemorrhage may be related to the abnormal vasculature within the tumor.

In general, PAs shows little or no peritumoral edema in MRI findings. However, in our cases, there were evident peritumoral edemas, which may have been due to a hemorrhagic effect. The enhancing patterns of PAs have been described as 1) a mass with an enhancing cyst wall and an intensely enhancing mural nodule, 2) a mass with a non-enhancing cyst and an intensely enhancing mural nodule, 3) a predominantly solid mass with minimal to no cystic component, and 4) a necrotic mass with a central non-enhancing zone\(^8\).

Our two cases showed a cystic mass with an enhancing mural nodule. In these cases, we performed radical resections of the enhanced lesions with hemorrhage. The surgical cures were successful without neurological deficits.

**CONCLUSION**

We report the first adult cases of cerebellar pilocytic astrocytomas presenting with intratumoral hemorrhages. It is extremely unusual for cerebellar pilocytic astrocytomas to be associated with hemorrhages, but this should be considered during differential diagnoses of intracerebellar hemorrhages with acute presentations. Such exceptional cerebellar tumors should be resected completely and should undergo histological examination to decide on further appropriate treatment.

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