

Acute Neurological Deterioration in an Adult with a Cerebellar Pilocytic Astrocytoma Accompanied by Intratumoral Hemorrhage, Subdural Hematoma, and Obstructive Hydrocephalus

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Disclosures can be found in Additional Information at the end of the article

Abstract

Acute neurological deterioration from intratumoral hemorrhage within a cerebellar pilocytic astrocytoma in an adult is an extremely rare presentation of this otherwise benign, predominantly pediatric tumor. Cerebellar pilocytic astrocytomas may present with cranial nerve deficits, headaches, dysmetria, and gait disturbances, as well as symptoms of hydrocephalus, including headaches, nausea, and emesis [1]. Symptoms typically progress in a more insidious fashion over time, as it is quite uncommon for pilocytic astrocytomas to present in such an acute manner, as did the subject of our case. Our patient presented with a Glasgow coma scale of 4T, and was found to have a large right-sided cerebellar hemorrhagic mass with an associated posterior fossa subdural hematoma, effacement of the fourth ventricle, and hydrocephalus. An emergent external ventricular drain was placed as a temporizing measure prior to urgently performing a suboccipital craniotomy and tumor resection. The patient had an excellent outcome after a long hospital course, and gave birth to her first child via Cesarean section three years after the inciting event.

Categories: Pathology, Neurosurgery, Oncology

Keywords: pilocytic astrocytoma, subdural hematoma, intratumoral hemorrhage, hydrocephalus

Introduction

Pilocytic astrocytomas are histologically benign tumors generally encountered in the pediatric population. They typically have a rather non-dramatic clinical presentation. Patients with pilocytic astrocytomas can have cranial nerve deficits, headaches, dysmetria, and gait disturbances, as well as symptoms of hydrocephalus, including headaches, nausea, and emesis. We aim to present the case of an adult harboring a posterior fossa pilocytic astrocytoma presenting in an acute fashion with a rapidly deteriorating neurological exam after spontaneous intratumoral hemorrhage. Cases of hemorrhagic presentation of these tumors have been reported in the literature [1-11], although they are quite rare. The vast majority of pilocytic astrocytomas are not associated with hemorrhage, leading one to infer that a small sub-population of these tumors may have inherent features making them more susceptible to hemorrhagic complications. Intratumoral encased aneurysms or dysplastic capillary beds may be contributing factors [5, 12]. In addition to this, some specific patient factors, such as older age, coagulation defects, and hypertension, may play a role [5, 12]. The case we are presenting highlights the importance of keeping pilocytic astrocytoma within the differential diagnosis in

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otherwise healthy adult patients presenting with an unexplained intracranial hemorrhage. Informed patient consent was waived and no identifying patient data was used in this paper.

Case Presentation

A 30-year-old otherwise healthy female presented with sudden onset of headache and became unconscious. She was intubated upon arrival and her Glasgow coma scale score upon arrival was 4T with extensor posturing and bradycardia. Her eyes were deviated downward, with non-reactive pinpoint pupils. CT scan of the head showed a calcified lesion in the right cerebellar cortex, with acute intralesional hemorrhage and adjacent subdural hematoma. The fourth ventricle was effaced with associated acute obstructive hydrocephalus (Figure 1).

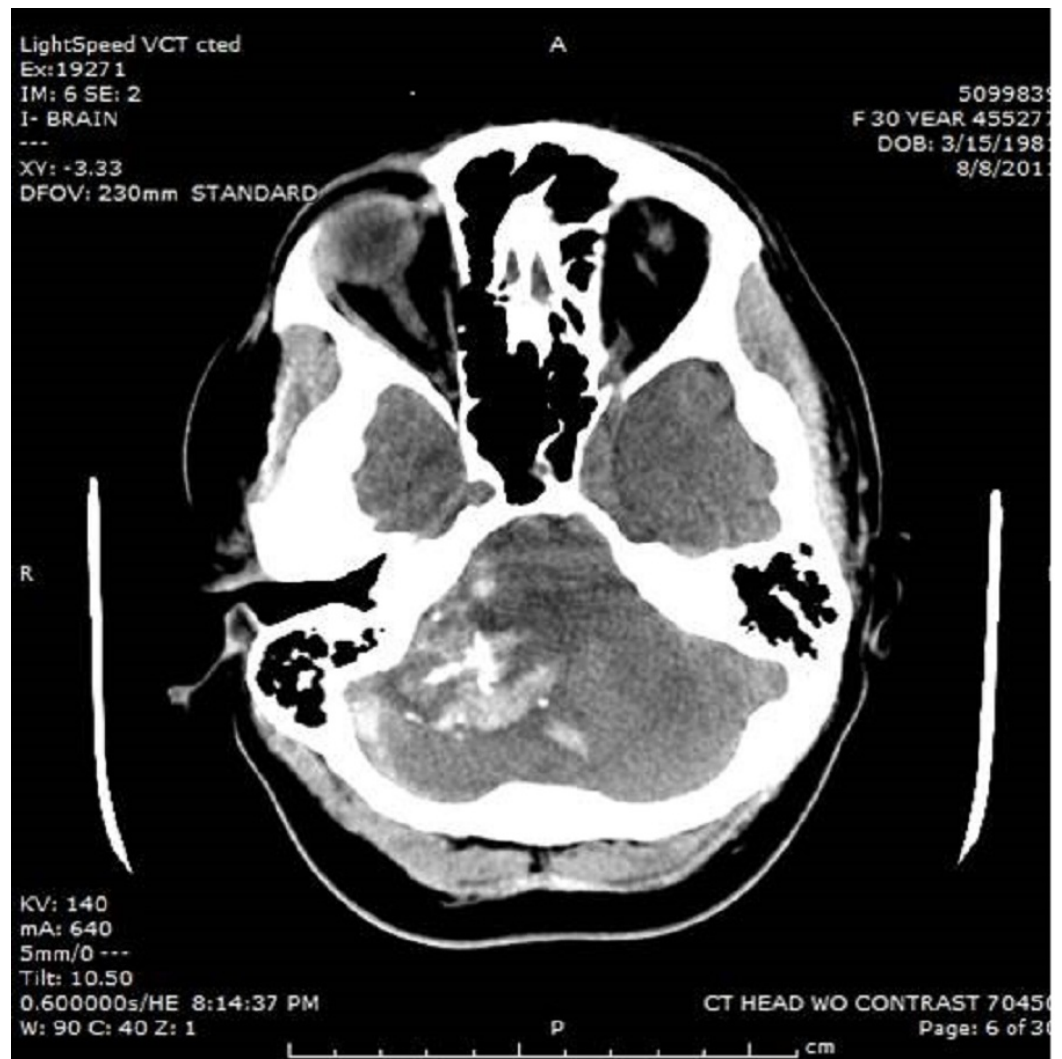


FIGURE 1: CT head revealing a right-sided hemorrhagic mass

We placed an emergent right frontal external ventricular drain in the emergency department (Figure 2).

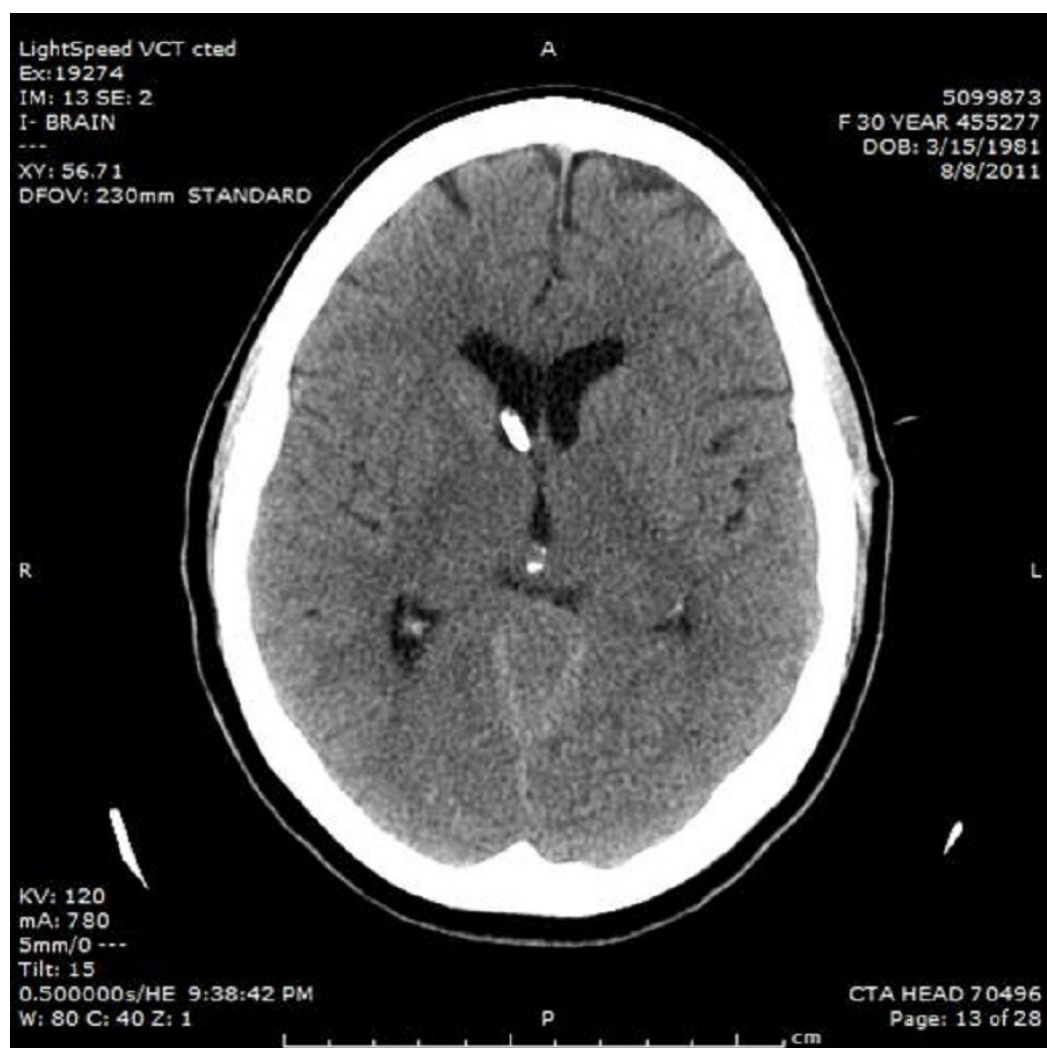


FIGURE 2: CT head revealing a right frontal external ventricular drain placed for hydrocephalus

Opening pressure was over 30 cm H₂O. CT angiogram of the head and neck to rule out underlying vascular lesions was unremarkable.

The patient was then taken to the operating room for surgical decompression. A paramedian suboccipital craniotomy was performed. The dura was opened and a subdural hematoma was encountered and subsequently evacuated. Intra-axial cerebellar tumor rapidly presented and dissection proceeded. A significant amount of intratumoral hemorrhage was noted. The cerebellar tumor was then resected in a gross-total fashion.

A very small amount of tumor was purposefully left on the brainstem to avoid significant morbidity from injury to the lower cranial nerves and the vertebral artery branches that were involved in the tumor (Figure 3).

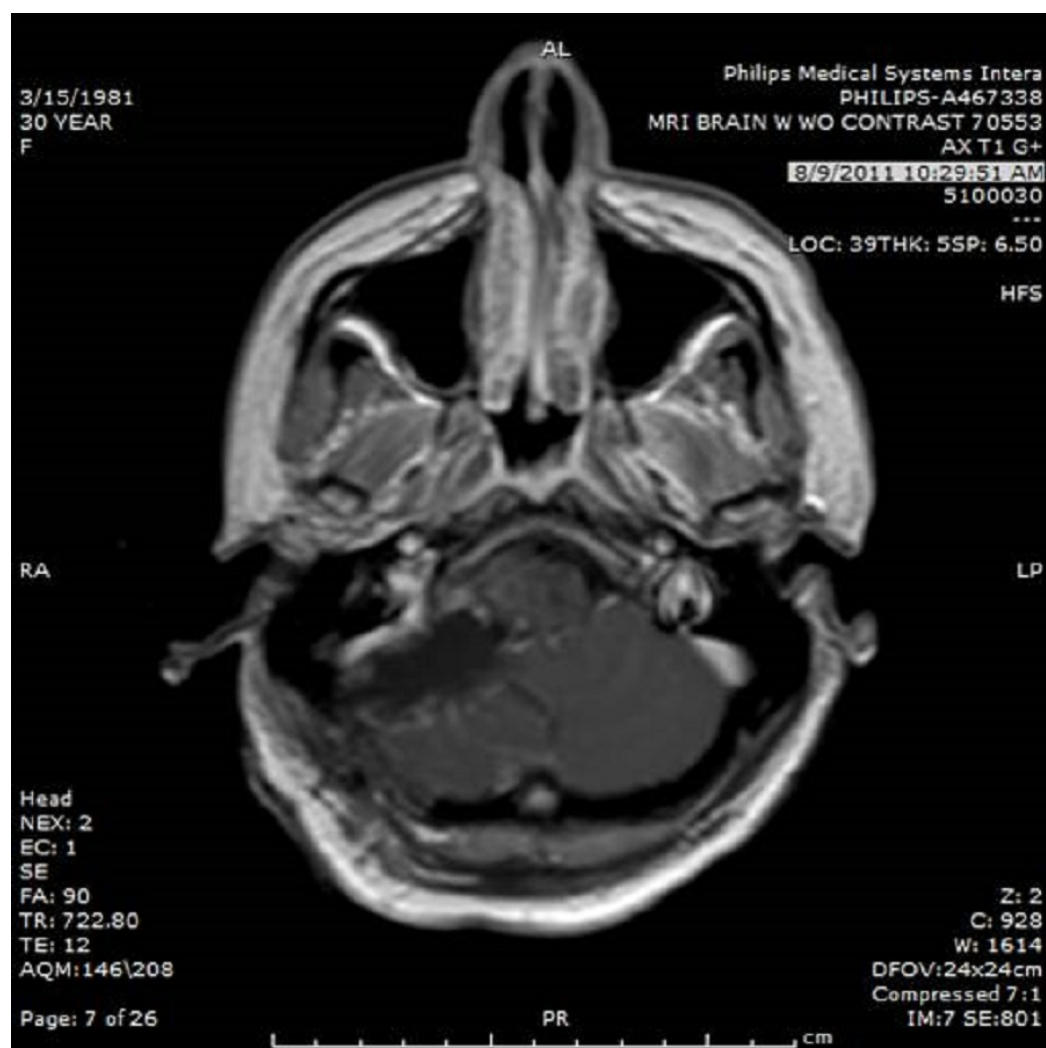


FIGURE 3: Postoperative MRI of the brain with contrast showing the tumor resection cavity

Final pathology of the tumor was pilocytic astrocytoma (Figures 4-7).

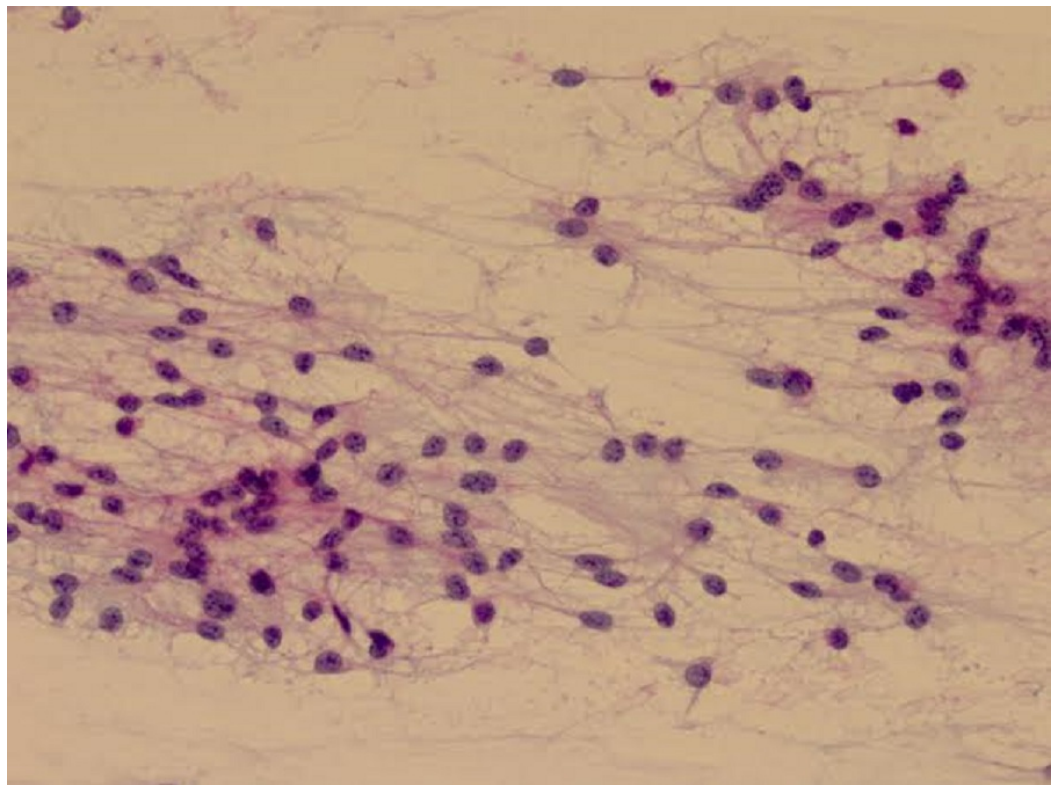


FIGURE 4: Smear at medium power. Relatively monomorphic cells with oval nuclei and bland cytology. Pilocytic processes are seen between the nuclei

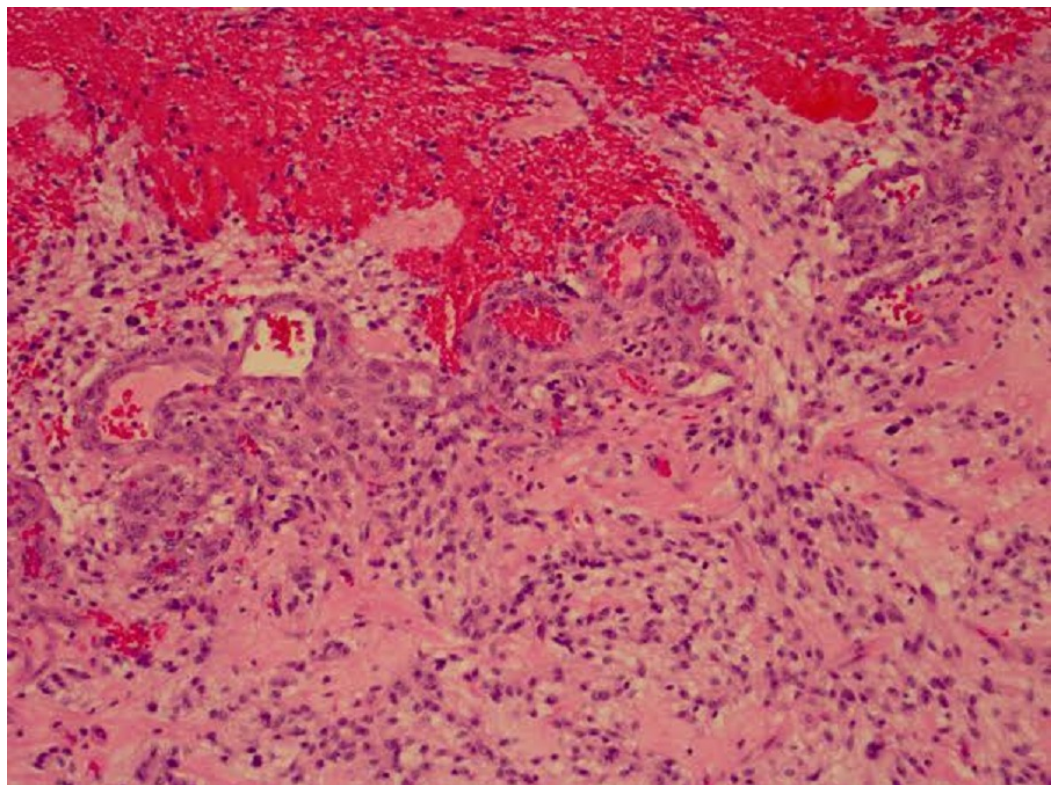


FIGURE 5: Pilocytic astrocytoma with endothelial proliferation with adjacent hemorrhage

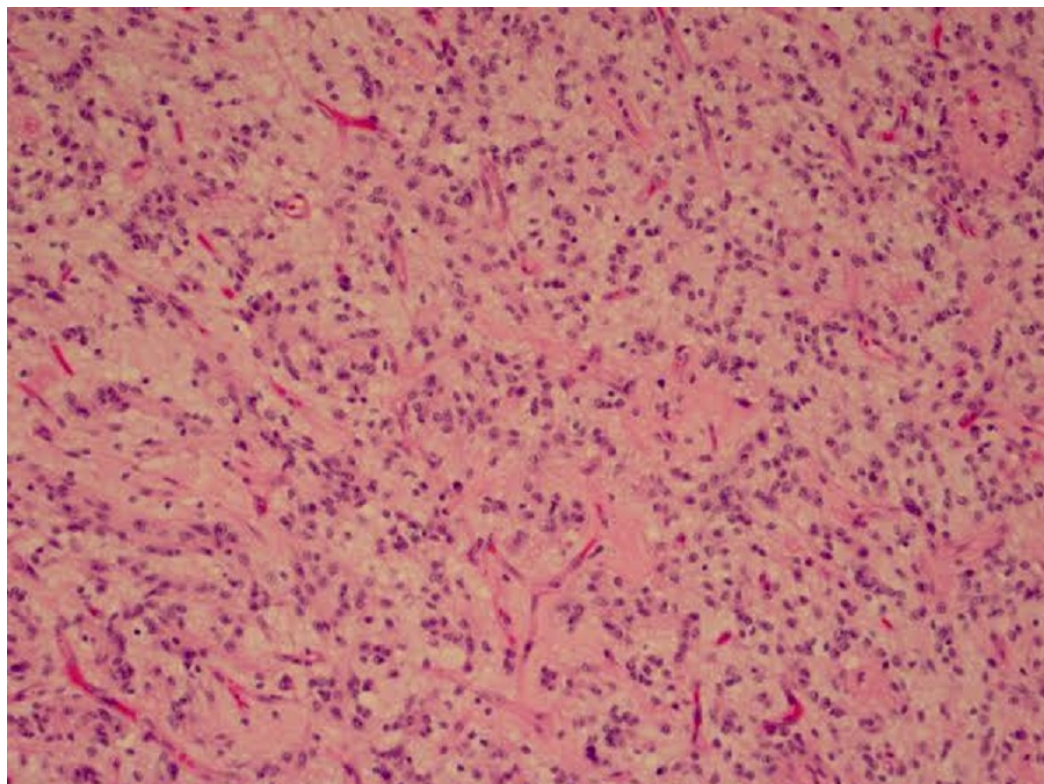


FIGURE 6: Pilocytic astrocytoma with some prominent endothelial cells but neoplastic cells without mitotic figures and with bland, oval nuclei similar to that seen in the smear

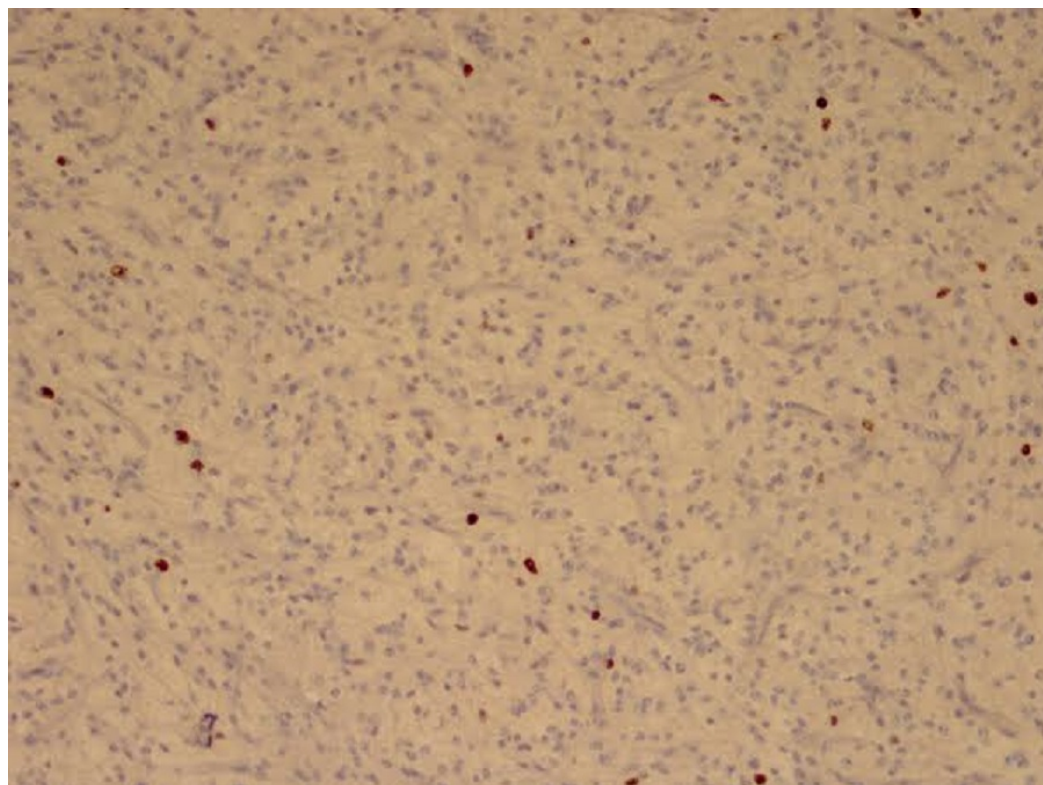


FIGURE 7: Ki-67 is low in the neoplastic cells and with some positive nuclei in the proliferating endothelial cells

The patient was following commands on the first postoperative day and was extubated after a few days. On examination, she had bilateral cranial nerve six palsies, right sensorineural hearing loss, and gait ataxia. The external ventricular drain was weaned and removed without any need for permanent CSF diversion. She was transferred to acute rehab and was eventually discharged home with significant clinical improvement. At one-year follow-up, she had minimal residual brainstem and cerebellar deficits. Adjuvant radiation therapy was considered but the radiation oncologist felt that the risks of radiation therapy were not justified. She had minimal residual brainstem and cerebellar deficits at her one-year follow-up. She was able to walk independently and go back to work. She got married and had a baby via Cesarean section. During the postoperative period and during her pregnancy, her residual tumor has remained stable and no further episodes of hemorrhage or growth has been noted over the two years of follow-up. Some of her symptoms, such as ataxia, got worse when she was pregnant but improved after delivery of the child.

Discussion

There have been very few reports of adult pilocytic astrocytomas presenting in such an acute fashion with a posterior fossa intratumoral hemorrhage, subdural hematoma, and acute obstructive hydrocephalus. Cases of hemorrhagic pilocytic astrocytomas have been reported in the literature, albeit on rare occasion. Golash, et al. reported on a case of a spontaneous intracerebral hemorrhage in a 13-year-old girl later found to be a pilocytic astrocytoma [2]. Lones and Verity described a fatal intracerebral hemorrhage in a 69-year-old woman caused by a pre-existing pilocytic astrocytoma [7]. Hwang, et al. described a case of a 34-year-old man presenting with a hemorrhagic hypothalamic pilocytic astrocytoma [3]. Oka, et al. reported on a 21-year-old man with a tectal pilocytic astrocytoma presenting with hemorrhage [9]. Lee, et al.

described a case of a hemorrhagic cerebellar pilocytic astrocytoma in a 15-month-old boy [6]. Lyons reported on a case of a spontaneous intracerebral hemorrhage in association with a pilocytic astrocytoma in a 75-year-old man [8]. Recurrent pilocytic astrocytomas are rare; however, recurrence associated with an intracerebral hemorrhage has been noted in the literature [10]. Another rare presentation of pilocytic astrocytomas has been reported in association with subarachnoid hemorrhage [1, 4]. Perhaps the most comprehensive review of hemorrhage in association with pilocytic astrocytomas was completed by White and colleagues. One hundred and thirty-eight patients with histologically-proven pilocytic astrocytomas were evaluated. The mean age at the time of diagnosis was 23 years. Approximately 8% of these individuals were found to have some degree of hemorrhage during their presentation. Although this number may seem higher than one would expect, their analysis found none of the hemorrhages to be in the cerebellum [11].

The pathophysiology underlying intra-tumor hemorrhage of a pilocytic astrocytoma remains elusive. Higher-grade astrocytomas present with hemorrhage more commonly as a result of vessel necrosis, rapidity of tumor cell proliferation, or due to neovascularization. Some contributing factors that are speculated in the literature to the etiology of hemorrhagic pilocytic astrocytomas are pre-existing hypertension, abundant neovascularization of the tumor, structural abnormalities from tumor cell invasion of the vasculature, coagulation defects, endothelial proliferation, rupture of encased aneurysms, and dysplastic capillary beds [5, 12]. Another theory is increased fibrinolytic activity secondary to the thromboplastin activity of brain tissue. When a pilocytic astrocytoma does hemorrhage, the vast majority of it is intra-tumoral. When subdural hematoma and subarachnoid hemorrhage are present, this is typically accounted for by direct extension of the intratumoral hemorrhage into other spaces [5, 12].

Another etiological factor for associated subdural hematoma is that the tumor itself can grow into the subdural space and cause traction on communicating veins. This traction can cause the vessels to become more susceptible to rupture, even from an otherwise minor trauma [3]. Shibao, et al. reported a case of a hemorrhagic pilocytic astrocytoma in an adult. They found that there was complex vascular proliferation within the tumor. This could potentially be accounted for by previously ruptured intratumoral vessels, with subsequent re-canalized thrombi. They also noted that some of the vasculature included thin-walled ectatic vessels, while other areas displayed sclerotic thick-walled vessels. Rupture within areas of this abnormal vasculature may be the etiology behind intratumoral hemorrhage of pilocytic astrocytomas. It has also been implied that some degree of degenerative vascular changes may be the underlying factor ultimately leading to hemorrhage, given the older age distribution of patients with hemorrhagic pilocytic astrocytomas [9].

Conclusions

Although hemorrhagic pilocytic astrocytomas may occur more frequently than are reported, our patient presented with several features making this a rather unique case. The patient in this report has made remarkable improvements during her rehabilitation. Two years after her surgery, she delivered a child via Cesarean section, and had no increase in the size of the tumor during the duration of her pregnancy. She has not had any further hemorrhage into the tumor. Underlying pilocytic astrocytoma should be considered in the differential diagnosis of adults presenting with unexplained cerebellar hemorrhage.

Additional Information

Disclosures

Human subjects: Consent was obtained by all participants in this study. **Conflicts of interest:**

In compliance with the ICMJE uniform disclosure form, all authors declare the following:

Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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