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CASE REPORT

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Adult cerebellar pilocytic astrocytoma presenting as hemorrhage—A confounding entity

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ABSTRACT

Pilocytic astrocytomas are histologically benign low-grade astrocytic tumors seen predominantly during the first two decades of life. Pilocytic astrocytomas in the adult population as well as the hemorrhagic manifestation of pilocytic astrocytomas are quite uncommon. Here, we present a case of the hemorrhagic manifestation of cerebellar pilocytic astrocytoma in a 48-year-old male who presented with a history of vertigo and headache for a week.

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KEYWORDS

Pilocytic astrocytoma; hemorrhage; cerebellum; adults

Introduction

Pilocytic astrocytomas are typically seen in the pediatric population and young adults and are pathologically benign tumors. Only one-third of patients are over the age of 18, and only 17% are over the age of 30 [1]. Among the pediatric age group, 67% are present as cerebellar lesions, whereas, among the adult population, 55% are present as supratentorial lesions [2]. Intratumoral hemorrhages are very rare in low-grade glial tumors such as pilocytic astrocytoma. Since the majority of cases of pilocytic astrocytomas are not associated with hemorrhage, it helps us to infer that a small percentage of these tumors may have inherent features such as a dysplastic capillary bed or an encased aneurysm that make them more susceptible to hemorrhagic complications [3]. We report a unique case of a cerebellar hemorrhagic tumor lesion in a 48-year-old male patient, which in subsequent pathological reports was identified as pilocytic astrocytoma—WHO grade I.

Case Report

A 48-year-old male patient presented to our hospital with complaints of vertigo and headache for the past 1 week, associated with vomiting for the past 2 days. The patient had previously undergone transsphenoidal excision of a pituitary macroadenoma in 2009. The patient was referred to our department for brain imaging to look for any organic cause for the headache and giddiness and was subjected to magnetic resonance imaging-Brain in a 3 Tesla scanner (Skyra, Siemens, Healthineers, Erlangen, Germany). The standard institute tumor protocol was fluid–attenuated inversion recovery, diffusion weighted imaging, and susceptibility weighted imaging sequences. Precontrast gradient (MP RAGE) sequences followed with T1 sagittal, T2 axial, 3D SPACE, and postcontrast fat-suppressed T1 weighted images. Subtracted images were also obtained.

It revealed a well-defined T1 heterogeneously hyperintense and T2 heterointense lesion in the left cerebellopontine angle causing a mass effect with compression of the fourth ventricle and anteromedial displacement of the pons. (Figs. 1a and b and 2). The lesion showed areas of restricted diffusion (Fig. 3) with a few foci of gradient blooming (Fig. 4a and b). An magnetic resonance (MR) angiogram revealed no significant abnormality. The lesion shows no contrast enhancement in the subtracted images (Fig. 5a and b). The 3 Tesla Scanner (Skyra, Siemens, Healthineers, Erlangen, Germany) used in our institution is a left-handed system. Calcifications are predominantly hypointense in all sequences and show gradient blooming with hypointensities in-phase images (left-handed systems). However, in

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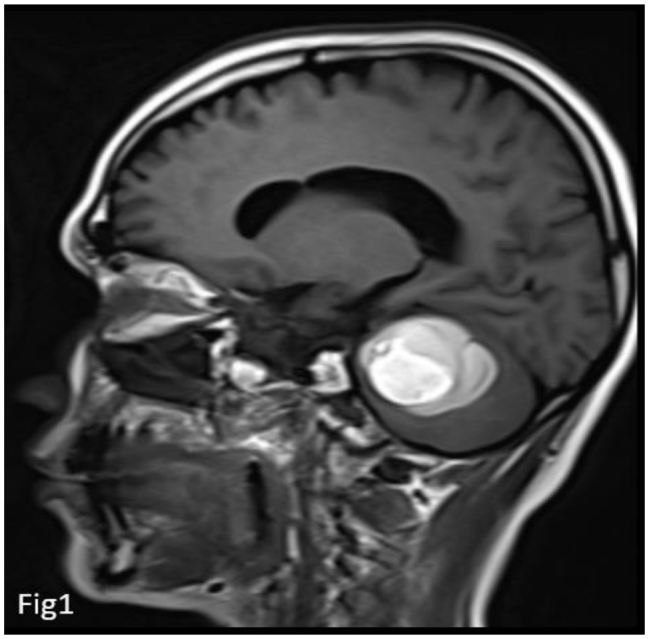


Figure 1. Preoperative sagittal T1-weighted brain magnetic resonance imaging showing a well-defined hyperintense cerebellar mass lesion with perilesional T1 hyperintense signal.

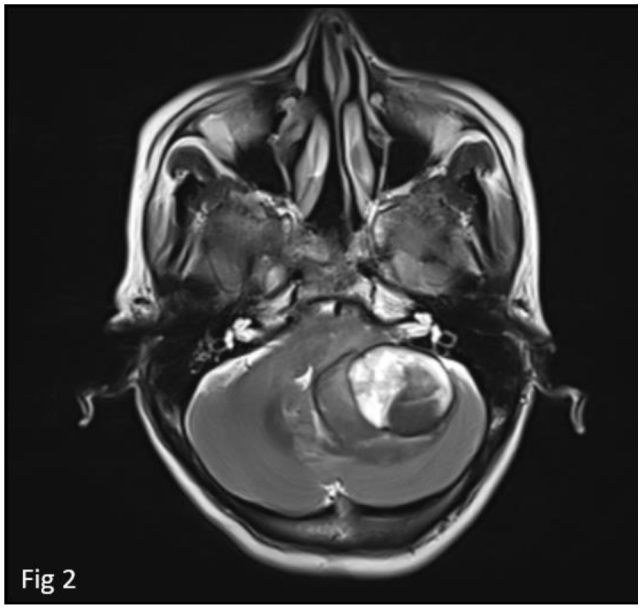


Figure 2. Preoperative axial T2-weighted brain magnetic resonance imaging shows a cerebellar mass with central mixed-signal intensity and peripheral low-signal intensity fat-suppressed T1 weighted images were followed by 3D.

our case, the lesion shows near uniform T1 hyperintensity in fat-saturated sequences and hyperintensity in phase images which are consistent with a hemorrhagic lesion and the possibility of a vascular

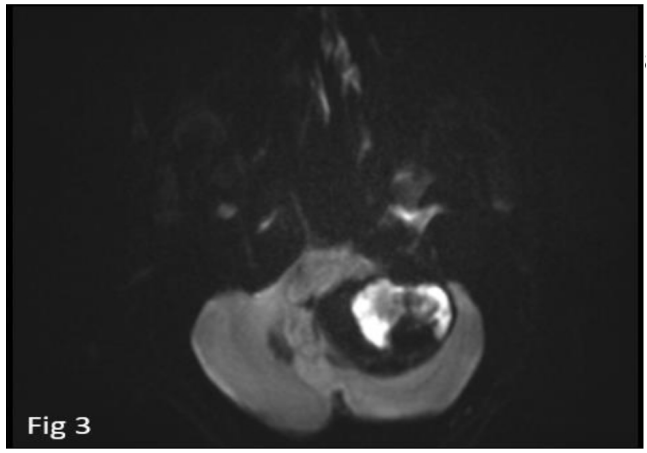


Figure 3. Axial diffusion-weighted images show areas of diffusion restriction within the lesion.

lesion like cavernous angioma, hemorrhagic tumor, and thrombosed aneurysm were considered.

The patient was later referred for diagnostic cerebral digital subtraction angiography. Through right femoral puncture, four vessel diagnostic cerebral digital subtraction angiography was done in our Biplane digital subtraction angiography, which revealed a paucity of vessels in the left cerebellar hemisphere with an upward displacement of the P1 segment of the left posterior cerebral artery and branches of the superior cerebellar artery, with no aneurysmal or tumor blush (Fig. 6). The patient then underwent near-total excision of the lesion. The histopathological report confirmed Pilocytic Astrocytoma WHO Grade-1, with neoplastic astrocytes in a glial fibrillary background, numerous Rosenthal fibers and eosinophilic granular bodies, numerous dilated, congested blood vessels, and areas of hemorrhage (Figs. 7 and 8).

Discussion

Pilocytic astrocytomas typically occur in young patients, have a relatively good prognosis, and are classified as WHO Grade-1 tumors with slow-growing benign biological behavior [4]. It has no gender predilection and has an overall incidence of about 2.9 to 4.8 cases per million people [5]. There exists a documented association with neurofibromatosis type 1. The clinical features depend on the size and location of the lesion, with the cerebellum being the most common, followed by the optic pathway (especially in NF1) [5]. The presenting complaint in our patient was headache and vertigo, which reflects the slow growth and indolent nature of the

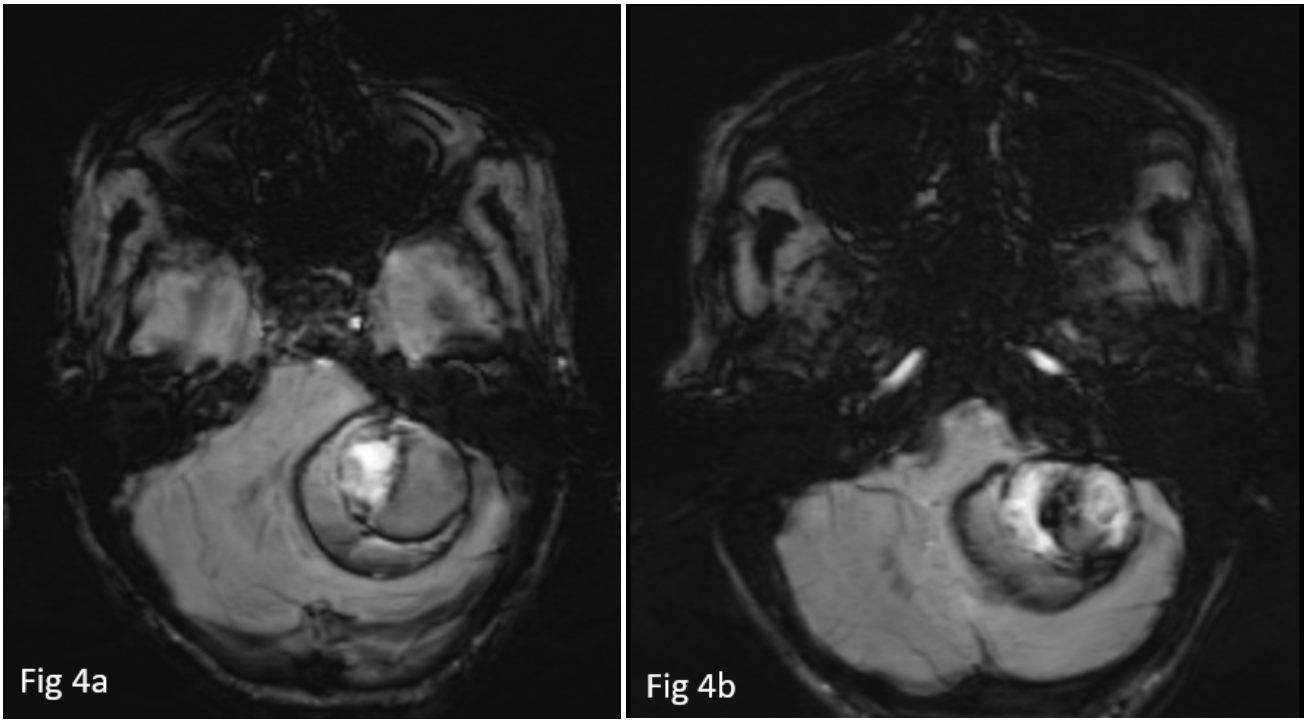


Figure 4. a, b. Axial susceptibility-weighted images show few foci of gradient blooming.

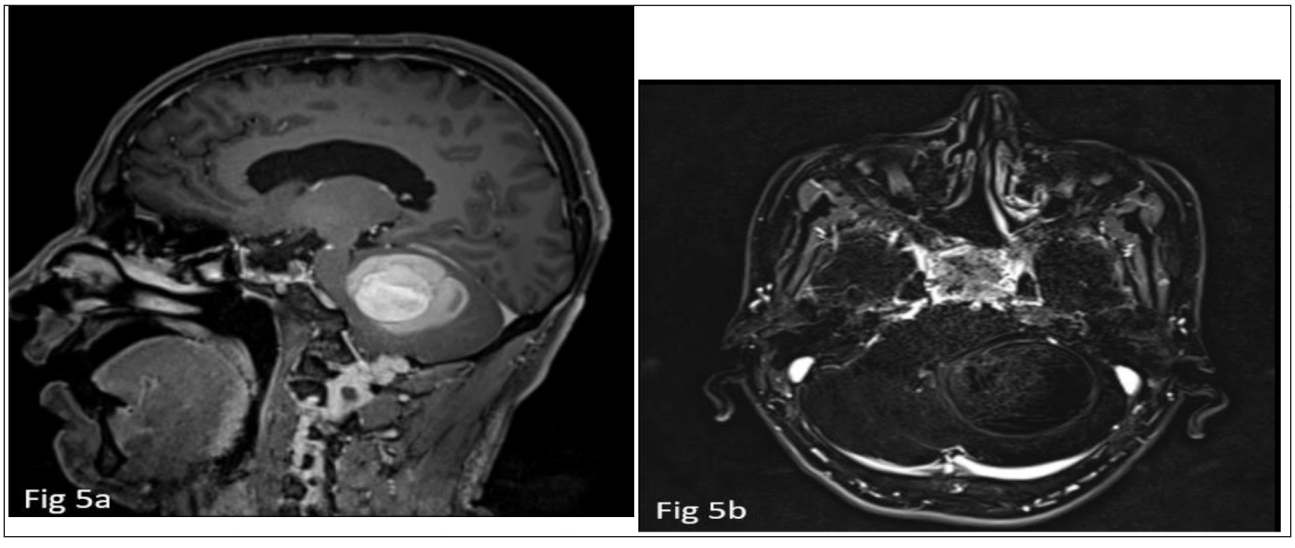


Figure 5. a and b. Sagittal T1 post-contrast and axial subtracted images show no contrast enhancement.

lesion [6]. A typical pilocytic astrocytoma macroscopically appears as a well-circumscribed cystic mass with a mural nodule, whereas microscopically it has a biphasic appearance in which compact areas containing brightly eosinophilic Rosenthal fibers alternate with spongier areas with microcysts [1]. There have been a few case reports of spontaneous hemorrhage of pilocytic astrocytoma in children. The presence of pilocytic astrocytoma in an elderly adult is extremely rare [7]. The two most common enhancement patterns in pilocytic astrocytomas are: i) a mass with an enhancing cyst wall and an intensely enhanced mural nodule (46%) and ii) a mass with a non-enhancing cyst wall and an intensely enhanced mural nodule (21%). However, the enhancement pattern of our case does not adhere to the aforementioned patterns.

Reported studies have documented the rate of spontaneous hemorrhage in a histologically proven case of pilocytic astrocytoma at 8%. Cerebellar hemorrhagic pilocytic astrocytomas are seen mostly in children and cerebral hemorrhagic

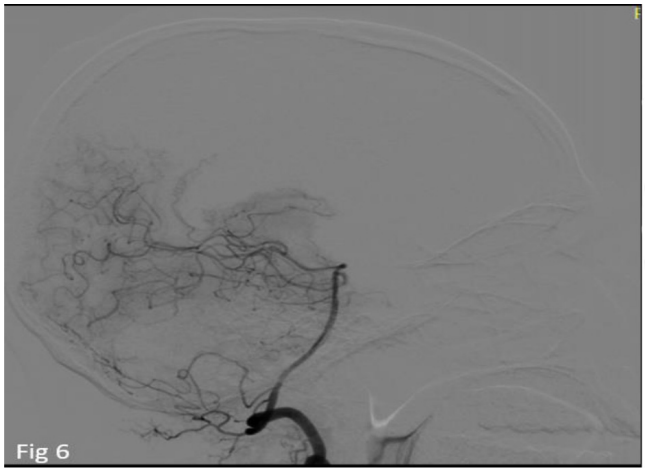


Figure 6. Selective catheterization of left vertebral artery shows a paucity of vessels in left cerebellar hemispheres with an upward displacement of P1 segment of the left posterior cerebral artery and branches of the superior cerebellar artery, with no aneurysmal blush.

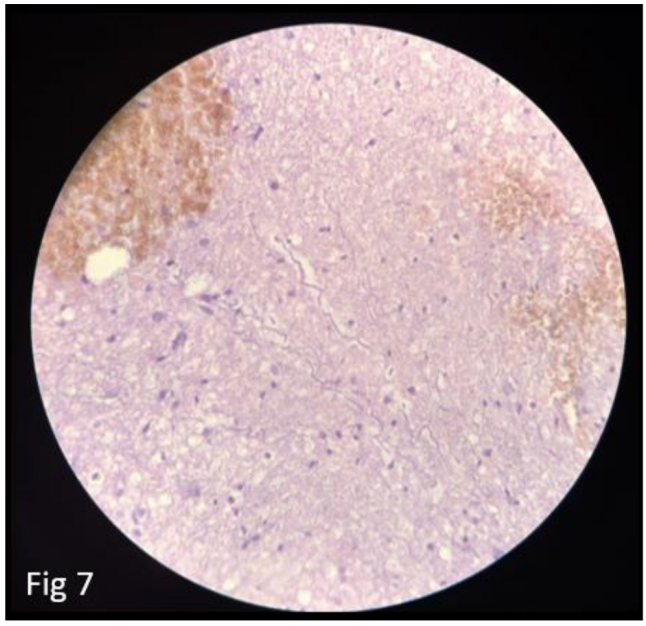


Figure 7. Histopathological image reveals irregular distribution of tumor cells with nucleus and fiber-rich regions and intratumoral hemorrhage. Rosenthal fibers present.

pilocytic astrocytomas in adults [2]. The pathophysiology behind the hemorrhage in high-grade gliomas is likely due to neovascularization and necrosis of blood vessels, whereas the mechanism of hemorrhage in pilocytic astrocytomas remains unclear [2]. The typical enhancement pattern these tumors exhibit may be attributed to their characteristic vascularization, which may have contributed to the intratumoral hemorrhage. Documented studies have reported that the hemorrhage may be

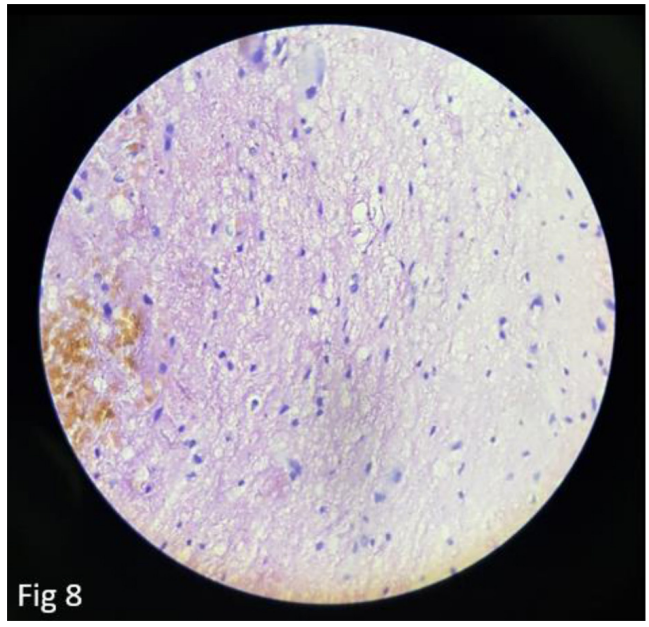


Figure 8. High magnification of eosinophilic granular bodies and Rosenthal fibers.

due to abnormal vasculatures such as thick-walled hyalinized vessels, glomeruloid endothelial proliferation, and thin-walled ectatic vessels [8]. In our case, the patient had normal platelet counts and no coagulation defects, hence the hemorrhage may be related to the abnormal vasculature within the tumor. The treatment of choice for such a lesion is usually total macroscopic resection [6]. It is uncommon for cerebellar pilocytic astrocytoma to present as a hemorrhagic lesion in an adult. A focal cerebellar hemorrhagic mass may be due to a neoplastic hemorrhage as well as a non-neoplastic hemorrhage. Hemorrhagic metastases may manifest as an acute intracerebral hemorrhage with no obvious enhancing mass as seen on MR images. Due to the perilesional T1 hyperintense signal, the diagnosis of a vascular lesion like cavernous angioma was also considered [9]. The typical appearance of the cavernous angioma with a mixed-signal intensity core and a reticulated popcorn appearance was not evident in this case. This case highlights this very unusual manifestation of an adult cerebellar pilocytic astrocytoma and one should be aware of this entity while reporting a hemorrhagic cerebellar lesion in an adult.

Conclusion

We report a case of cerebellar pilocytic astrocytoma presenting with intratumoral hemorrhage. It is extremely unusual for cerebellar pilocytic astrocytomas to be associated with hemorrhage, but this

should be considered during differential diagnosis of a vascular hemorrhagic lesion in the cerebellum. This case reveals the confounding features of adult hemorrhagic cerebellar pilocytic astrocytoma in various sequences of magnetic resonance imaging.

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