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Clinical case

Pilocytic astrocytoma mimicking cavernous angioma: Imaging features and histological characteristics

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ABSTRACT

Pilocytic astrocytoma (PA) commonly occurs during the first two decades of life. Typical locations include cerebellum, optic nerve, optic chiasm/hypothalamus and brainstem. PA should be considered in the differential diagnosis of patients with brain tumors manifesting with hemorrhagic onset. We report a case of a hemorrhagic onset of cerebellar PA in a young adult with imaging findings mimicking cavernous angioma. We also discuss imaging features and histological characteristics with a focus on the etiology of the hemorrhagic onset.

1. Introduction

Pilocytic astrocytoma (PA) commonly occurs during the first two decades of life and is one of the most common pediatric primary brain tumors. Typical locations include cerebellum, optic nerve, optic chiasm/hypothalamus and brain stem. PA is a relatively circumscribed, slowly growing astrocytoma and corresponds to World Health Organization (WHO) grade I. This tumor therefore usually presents with slowly evolving focal deficits or signs of increased intracranial pressure. The incidence of intracranial hemorrhages from gliomas is reported to be from 3.7 to 7.2%, with high incidences seen in high-grade astrocytomas, oligodendrogliomas and mixed oligoastrocytomas. Low-grade glial tumors very rarely present with intracranial bleeding, but when they do, these presentations are often catastrophic [1].

We report the case of a hemorrhagic onset of cerebellar PA in a young adult with imaging findings mimicking cavernous angioma. We discuss imaging features and histological characteristics with a focus on the differential diagnosis of this type of tumor with cavernous angioma and the etiology of hemorrhagic onset.

2. Description

A 23-year-old man presented with a 3-month history of progressively intense headache. There was no noticeable trauma history. On examination, only a static cerebellar syndrome was observed. Bilateral fundi were normal.

Brain computed tomography (CT) revealed a heterogeneous hyperdense vermis mass protruding within the fourth ventricle (V4), causing obstruction of V4 foramen and subsequent mild hydrocephalus. After administration of a contrast agent, a thick peripheral heterogeneous enhancement mainly at the inferior and anterior margin associated with prominent posterior peritumoral vessels were observed (Fig. 1). On the CT angiogram (CTA), there was no evidence of any other vascular abnormalities, such as an arteriovenous malformation or aneurysms.

Magnetic resonance imaging (MRI) demonstrated a heterogeneous mass in the inferior cerebellar vermis (uvula and nodulus), protruding in the fourth ventricle with central high-signal intensity on T1-weighted image (T1WI) and peripheral stratified low-signal intensities on T2* consistent with acute, subacute and chronic hemorrhages. The T2* hyposignal delineating the brainstem and cerebellum consisted of superficial siderosis secondary to chronic subarachnoid bleeding. Discrete edema was noted surrounding the mass. After gadolinium administration, the lesion displayed

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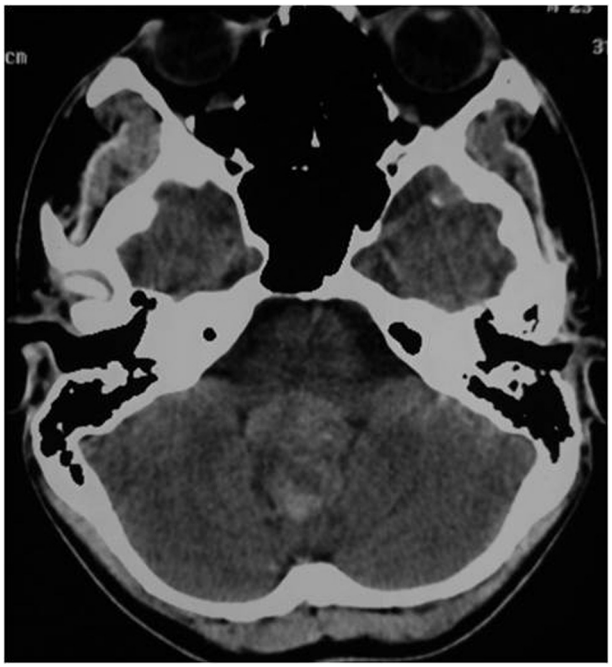

Fig. 1. Brain CT scan without contrast at admission showing a heterogeneous hyperdense mass occupying the inferior vermis.

a more obvious thick peripheral heterogeneous enhancement (Fig. 2).

A cerebellar cavernous angioma was the first diagnosis considered regarding the different stages of the hemorrhagic components but other possibility could not be excluded.

The patient underwent urgent total resection of the lesion.

The histological examination revealed a glial tumor with low MIB-1 labeling index, large zone of necrosis, hemosiderin deposition and numerous Rosenthal fibers. The histological diagnosis was pilocytic astrocytoma (Fig. 3).

There were no intraoperative or postoperative complications. Postoperative CT confirmed the total resection of the tumor and no adjuvant therapy was performed.

3. Discussion

PA, classified as a grade I astrocytic tumor according to the World Health Organization (WHO), is the most common type of glioma in children and young adults, accounting for approximately 5–6% of all gliomas.

Its typical histological characteristics include a biphasic pattern with varying proportions of compacted bipolar cells associated with Rosenthal fibers and loose-textured multipolar cells associated with microcysts and eosinophilic granular body droplets [2].

Until recently, almost nothing was known about the molecular mechanisms involved in the development of PAs. The use of high-throughput sequencing techniques interrogating the whole genome has shown that single abnormalities of the mitogen-activating protein kinase (MAPK) pathway are exclusively found in almost all cases, indicating that PA represents a one-pathway disease. The most common mechanism is a tandem duplication of a ≈2 Mb fragment of #7q, giving rise to a fusion between two genes, resulting in a transforming fusion protein, consisting of the N-terminus of KIAA1549 and the kinase domain of BRAF. Additional infrequent fusion partners have been identified, along with other abnormalities of the MAP-K pathway, affecting tyrosine kinase growth factor receptors at the cell surface (e.g. FGFR1) as well as BRAF V600E, KRAS, and NF1 mutations among others [3].

PAs are rarely associated with gross intratumoral hemorrhage despite rich vascularization and blood vessel changes, it is observed in only 1.1–3% of these tumors often accompanied by perivascular depots of hemosiderin [1]. Hemorrhagic onset of PAs is uncommon, and the etiology of hemorrhage remains unclear.

Cavernous angioma is one of the main differential diagnoses of hemorrhagic PA. Similar signal characteristics may be noted due to blood products degradation. The large spontaneous intratumoral hemorrhage with acute, subacute and discrete edema surrounding the mass chronic pattern noted in our case initially suggested the diagnosis of cavernous angioma although the demonstration of heterogeneous peripheral enhancement was against this hypothesis. In fact, even if contrast enhancement has sometimes been observed in cavernous angioma, it can be present in delayed images or in dural cavernoma.

Apart from giant cavernous malformation, in most cases, there is no surrounding edema or signals on T1 and T2 sequences vary widely, depending on the age of the blood breakdown products.

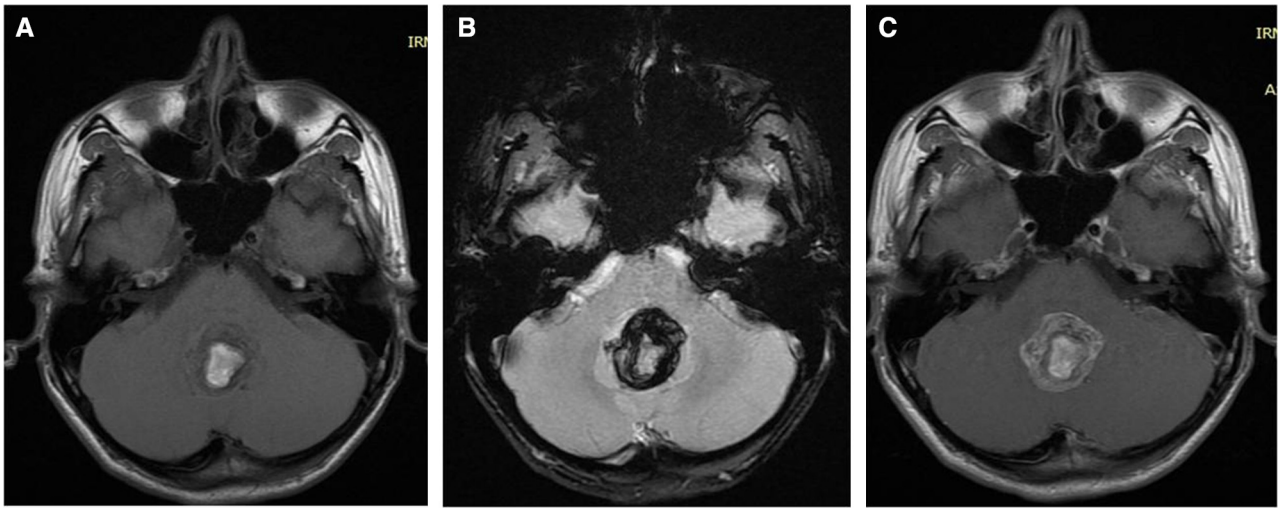

Fig. 2. Brain MRI including axial T1 (A), T2* (B) and T1 post-contrast (C) sequences showing a cerebellar mass arising from the inferior vermis and protruding within the fourth ventricle showing mixed signal intensity consisting of a central high-signal mainly on T1WI and peripheral stratified low-signal intensities on T2* with hyposignal delineating the brainstem and the ponto-cerebellar angles. Different stages of hemorrhage with superficial siderosis. Demonstration of thick and heterogeneous peripheral enhancement.

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