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Short clinical case

Hemorrhagic presentation of frontal partially calcified pilocytic astrocytoma in an 18-year-old woman: A case report and literature review as "clinical case"

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ABSTRACT

We report an unusual case of a frontal partially calcified pilocytic astrocytoma (PA) (WHO grade 1) in an 18-year-old woman who presented with acute, spontaneous intracerebral hemorrhage. Histopathology revealed the PA was mixed with psammoma bodies and areas of vascular proliferation responsible for a hypervascular pattern. The patient underwent a total gross resection. MRI showed no residual tumor at the 18-month follow-up and her neurological deficits improved after rehabilitation. Only 20 cases, including ours, of hemorrhagic presentation of PA in adults have been reported to date with enough radiological data. Furthermore, hemorrhagic presentation of a calcified PA is extremely rare. To date only two other cases of calcified PA with hemorrhagic presentation have been reported, one in an adult and one in an infant as described by Shibao et al. (2012) and Kapoor et al. (2015) respectively. Endothelial proliferation may be the main cause of bleeding in these lesions. In our case, a hypervascular pattern was exhibited by histopathological findings. A diagnosis of PA should be considered, especially when calcifications are present within a hemorrhagic tumor lesion.

1. Introduction

Pilocytic astrocytomas (PAs) occur predominantly in the pediatric population and are very rare in adults, affecting predominantly adults before the fourth decade. Radiological data of PAs with intratumoral hemorrhage have been sporadically reported in adults and children.

A recent review by Prasad et al. [1] documented a total of 53 cases of hemorrhagic PA (HPA), 27 pediatric and 26 adult cases. Only 20 cases of HPA, including ours, have been reported to date in adults with supportive radiological data [1–16]. On the other hand, calcified PAs are exceedingly rare and only two cases with associated hemorrhage have been reported, one in an adult and the other in a child [12,17]. We present an unusual case of spontaneous hemorrhage in a frontal partially calcified pilocytic astrocytoma in an 18-year-old woman.

2. Case report

An 18-year-old woman, with a past medical history of seizure, presented with sudden onset of paresthesia and right hemiparesis, headache and vomiting. She was referred to the emergency department for potential thrombolysis. Neurological examination found right hemiparesis and drowsiness.

The pattern of the lesion on plain CT and on MRI (T1, FLAIR, T2*, diffusion-weighted and 3D-T1 post-contrast images) was that of acute parenchymal hemorrhage with peripheral calcifications without enhancement on post-contrast T1 images (Fig. 1a–b). The patient was operated on. Intraoperatively, a thin-walled and grayish-colored cyst was easily distinguishable from the normal parenchyma. Hematomas were present within the cyst as well as hemorrhagic fluid (30 mm^3). Gross total resection of the tumor was performed. Histopathology confirmed PA (WHO grade I) mixed with psammoma bodies and areas of vascular proliferation

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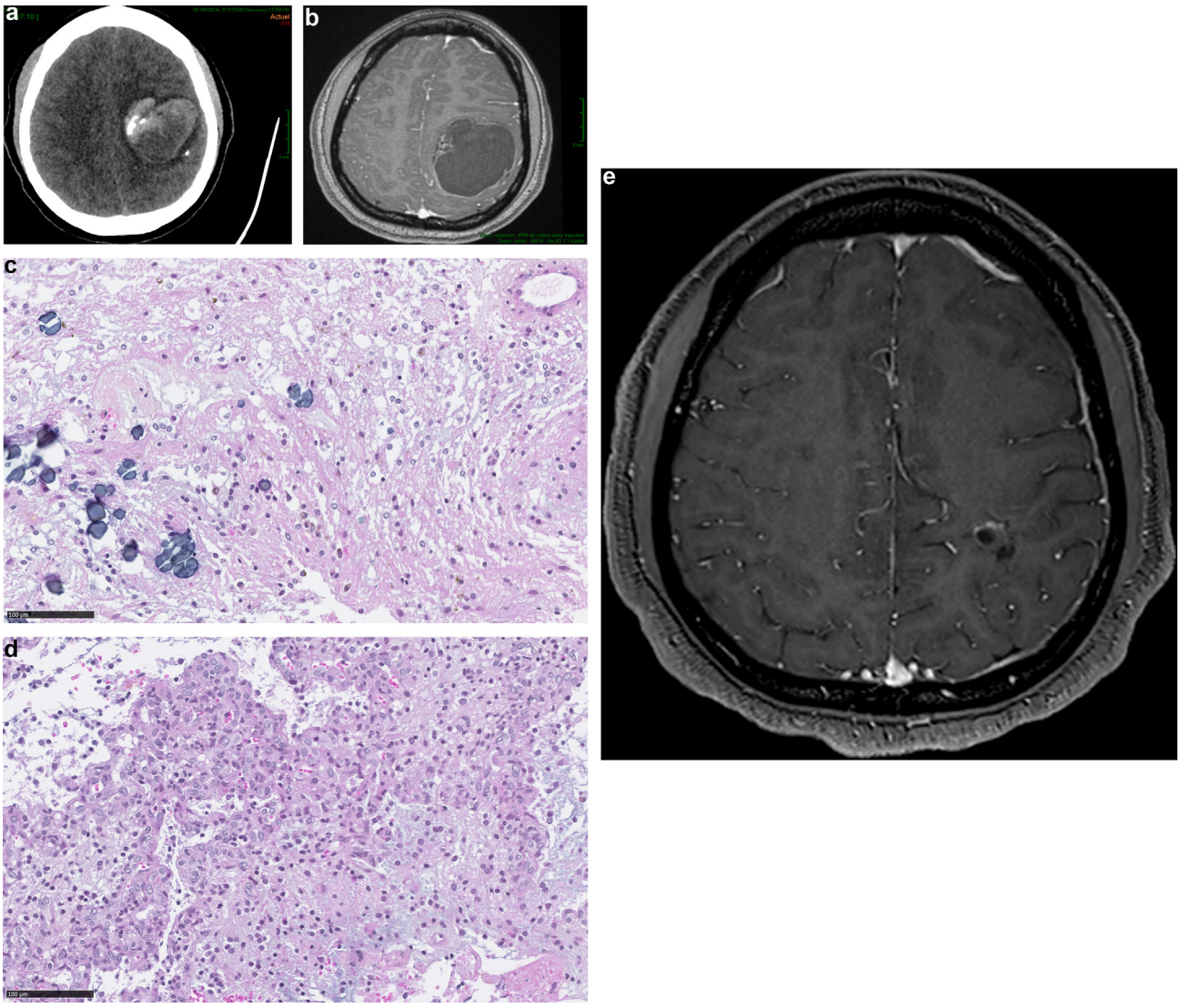

Fig. 1. a–b: non-enhanced CT and MRI at admission. Non-enhanced CT shows a slightly hypodense mass in the left frontal lobe (5.5 × 5.6 × 5.8 cm) with a hyperdense halo, surrounded by mild edema, with partial obliteration of the left lateral ventricle. A large calcification is present at the anterior corner and a punctate calcification at the posterior margin of the hematoma (a). Magnetic resonance imaging: After contrast agent injection, there is no enhancement (b); c–d: pathologic findings. Hematoxylin and eosin stain. Bars 100 μm; c: pilocytic astrocytoma showing a biphasic architectural pattern made of astrocyte cells with a typical piloid band pattern (piloid areas and cyst-like areas) mixed with psammoma bodies; d: pilocytic astrocytoma with area of vascular proliferation consisting of glomeruloïd vessels responsible for a hypervascular pattern; e: 18-month follow-up MRI-post-contrast 3-dimensional (3D) T1 acquisition: there is no residual tumor.

responsible for a hypervascular pattern (Fig. 1c–d). After tumor removal, the patient had mild residual right hemiparesis.

At the 18-month follow-up, the deficit improved with rehabilitation and MRI showed no residual tumor on T1, FLAIR, and post-contrast images (Fig. 1e).

3. Discussion

Pilocytic astrocytoma (PA) is classified as WHO Grade 1 with a rather benign, slow-growing behavior [18]. The most common location is the posterior fossa in children and the supra-tentorial area in adults, most commonly in the temporal lobes. The usual pattern is a cystic mass with an enhancing mural nodule.

The degree of tumor malignancy is highly correlated with a hemorrhagic event. The occurrence of hemorrhage in PA is said to be less than 1% [19]. However, recent pathological reports indicate that spontaneous hemorrhage in PA is more common than previously assumed: 8% in a series of 138 cases [10,20]. Nonetheless, radiological evidence of these hemorrhages in PAs without malignant transformation has been only sporadically reported in adult and pediatric populations. The etiology of these intratumoral hemorrhages remains obscure and continues to be debated [12]. Endothelial proliferation has been proposed to be responsible for the bleeding in these lesions. In the current case, we found areas of vascular proliferation made of glomeruloïd vessels resulting in a hypervascular pattern. Our findings are somewhat similar to those of Shibao who described a case of hemorrhagic and calcified PA in the cerebellar lobe of an adult [12].

A systematic search using combinations of keywords was performed in PubMed, from inception to March 10, 2018 for publications on adult cases of HPA, using the terms pilocytic astrocytoma, hemorrhagic, cranial. To be included in our review, the reported cases had to be diagnosed by CT scan and/or by MRI with a radiological description.

All studies identified were written in English and were case reports or case series, describing 1 to 2 cases per publication. In total, we found 20 cases, including ours, of pure HPA in adults with sufficient radiological data [1–16] (Table 1).

Table 1
Published cases of hemorrhagic pilocytic astrocytoma in adults with supportive radiological data.

	Author/Year/Ref No.	Case #	Age/sex	CT/MRI	Location	Surgery	Follow-up
1	Glew (1977) [2]		30/M	CT	Hypothalamic/chiasmatic	N/A	
2	Lones (1991) [3]		69/F	CT	Thalamo-caudate	Died	
3	Sorenson (1995) [4]		58/F	MRI	Hypothalamic/chiasmatic	Y	1 Yr.
4	Matsumoto (1997) [5][a]		45/M	MRI	Hypothalamic/chiasmatic	Y	6 m
5	Hwang (1998) [6]		34/M	CT/MRI	Hypothalamic/chiasmatic	N/A	N/A
6	Lyons (2007) [7]		75/M	CT	Cerebral lobe	Y	N/A
7	Oka (2007) [8]		21/M	CT/MRI	Tectum	Y	2 Yrs.
8	Li (2008) [9]	Case 1	32/M	CT/MRI	Cerebral lobe	Y	6 m
9		Case 2	34/M	MRI	Cerebral lobe	Y	6 m
10	Shibahara (2009) [10]	Case 1	22/M	CT	Tonsil – medulla	Y	N/A
11		Case 2	21/M	CT	Tectum	Y	N/A
12	Kim (2011) [11]	Case 1	37/F	CT/MRI	Cerebral lobe	Y	N/A
13		Case 2	53/M	MRI	Cerebral lobe	Y	N/A
14	Shibao (2012) [12][b]		29/M	CT/MRI	Cerebellar lobe	Y	N/A
15	Nakano (2015) [13]	Case 5	37/M	MRI	Cerebellar lobe	Y	40 m
16	Galgano (2016) [14]		30/F	CT	Cerebellar lobe	Y	3 Yrs.
17	Soliman (2016) [15]		35/M	CT/MRI	Hypothalamic/chiasmatic	Y	7 Yrs.
18	Prasad (2017) [16]		22/M	CT/MRI	Supra-sellar	Y	11 m/residual tumor
19	Gaha (2017) [1]		23/M	CT/MRI	Vermis	Y	N/A
20	Present case (2018)		18/F	CT/MRI	Cerebral lobe	Y	18 m

M: male; F: female; CT: computed tomography; MRI: magnetic resonance imaging; Yr(s): year(s); m: month; N/A: not available.
[a] Associated subarachnoid hemorrhage.
[b] Associated calcifications.

No elective location susceptible to hemorrhage has been reported even if there are preferential locations for PA in adults, especially the cerebral lobes (Table 1); these findings are consistent with previous pathological results [20]. However, it has to be emphasized that there are more case reports in adults than children with hemorrhagic presentation, whereas this tumor is more frequent in the pediatric population. In White's series of 132 PA cases, there were no pediatric cases among the hemorrhagic subgroup [20].

The typical nodular and cystic aspect is very rarely observed in cases of hemorrhagic lesions (2/30: 6% in Li's series) [9]. Usually the most common non-calcified PA has parenchymal hemorrhage on imaging, which makes the preoperative diagnosis difficult. Rare presentations include HPA associated with subarachnoid hemorrhage [5], imaging features mimicking cavernous angioma [16] or subtle enhancement at the lesions' margin [12].

Pilomyxoid astrocytoma (PMA), a recently recognized WHO grade II tumor considered as a rare variant of PA, less frequently presents with hemorrhage than PA. But PMA has a more aggressive behavior, based on the largest radiological study of 10 PMA cases vs. 38 PA cases: 20% hemorrhagic PMA versus 31% HPA [21]. These findings agree with those of Shibahara [10] who reported that 16 of 445 neuro-epithelial tumors (3.6%) had a hemorrhagic onset. Of these, there were 3 PA and 1 PMA from a total of 35 PA/PMA cases. Other than this large study, only 5 isolated cases of hemorrhage at onset have been reported in PMA [10,22–25]. In fact, few cases have been reported because PMA has only recently been described relative to PA.

Calcified PAs are even exceedingly rare. Interestingly, calcifications within PMAs have not been reported. Only 2 cases of calcified PAs presenting with intratumoral hemorrhage have been reported [12,17] (Table 1). While the radiological diagnosis of hemorrhagic PA is unlikely, the presence of calcification foci is suggestive of a benign lesion that may have bled.

Lastly, most reported cases underwent gross total resection with limited probability of recurrence.

4. Conclusion

In the current case, the very uncommon occurrence of a frontal PA with two unusual features – hypervascular pattern with hemorrhage and calcifications – is reported. Endothelial proliferation may be responsible for bleeding in these lesions. A diagnosis of PA should be considered, especially when calcifications are present within a hemorrhagic tumor lesion.

Authors contribution

MB, JPL have co-written the manuscript.

SG has operated the patient and gave the neurosurgical findings and follow-up.

D-CH: performed the pathological examination and gave the corresponding data.

Disclosure of interest

The authors declare that they have no conflict of interest.

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