

## Adult Pilomyxoid Astrocytoma with Hemorrhage in an Atypical Location

### Abstract

Pilomyxoid astrocytomas (PMAs) are generally seen in young children and tend to occur in the hypothalamic–chiasmatic region. Their presence in other parts of the brain in the nonpediatric age group is uncommon. In addition, hemorrhage in such low-grade tumors is an occasional event. We describe a case of PMA that presented with spontaneous bleed in an atypical location (temporoparietal region) in an adult. A concise literature of the cases of PMA which were associated with bleed is also presented. Occasionally, low-grade tumors such as PMA may present with bleed. These benign looking neoplasms behave differently from the commonly occurring pilocytic astrocytomas (PAs) and should be considered as a differential. It is important to differentiate these from PA, as the management and prognosis differs.

**Keywords:** *Astrocytoma, hemorrhage, intratumoral, pilocytic, pilomyxoid, tumor*

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### Introduction

Pilomyxoid astrocytoma (PMA), once considered as a subtype of pilocytic astrocytoma (PA), has been recently characterized as a distinct entity in the World Health Organization (WHO) classification 2007. In contrast to the PA (WHO grade I), this tends to be locally aggressive, with higher chance of leptomeningeal dissemination, recurrence, and poor prognosis.<sup>[1,2]</sup> The usual location is in the hypothalamic/chiasmatic region and occurs in infants and young children. At times, the pilomyxoid tumors may present with bleed in these areas.<sup>[3-6]</sup> Their presence in adults, especially in nonchiasmatic location, is unusual.<sup>[4,7]</sup> Furthermore, associated spontaneous hemorrhage in such eccentric sites is extremely rare.<sup>[7]</sup> We hereby report a case of PMA in the temporal region in an adult that presented with tumoral bleed. This is the second of its kind in the literature.

### Case Report

Informed consent was obtained from the patient. A 40-year-old man presented to our emergency services with features of raised intracranial pressure of 2-week duration. Neurological examination showed no deficits. Computed tomography showed an ill-defined, hypodense lesion in the left temporoparietal region with areas of bleed

and lateral ventricular hemorrhage. The lesion caused mass effect over the surrounding brain. Magnetic resonance imaging revealed a T1 isointense, T2 hyperintense lesion with intraventricular extension, and heterogeneously enhancing on gadolinium contrast [Figure 1]. Preliminary diagnosis was that of a high-grade glioma/choroid plexus tumor. A left temporoparietal craniotomy with near-total excision of tumor was performed. Intraoperatively, the tumor was soft, suckable, and moderately vascular. A small portion adherent to choroid plexus was left behind.

On histopathology, the tumor was moderately cellular arranged in sheets in abundant myxoid matrix [Figure 2]. The tumor cells were oval to bipolar and showed moderate nuclear pleomorphism. Microcystic changes and few mitotic figures were also noted. The stroma was rich in blood vessels with few dilated staghorn-like blood vessels. The tumor cells were focally positive for glial fibrillary acid protein and negative for CD 34 and B-cell lymphoma-2 2. A diagnosis of PMA (WHO grade II) was made. Postoperatively, he received radiotherapy and was well at 3-month follow-up.

### Discussion

Tihan *et al.* in 1999 identified the term “pilomyxoid astrocytoma” for a more aggressive variant of PA in infants and

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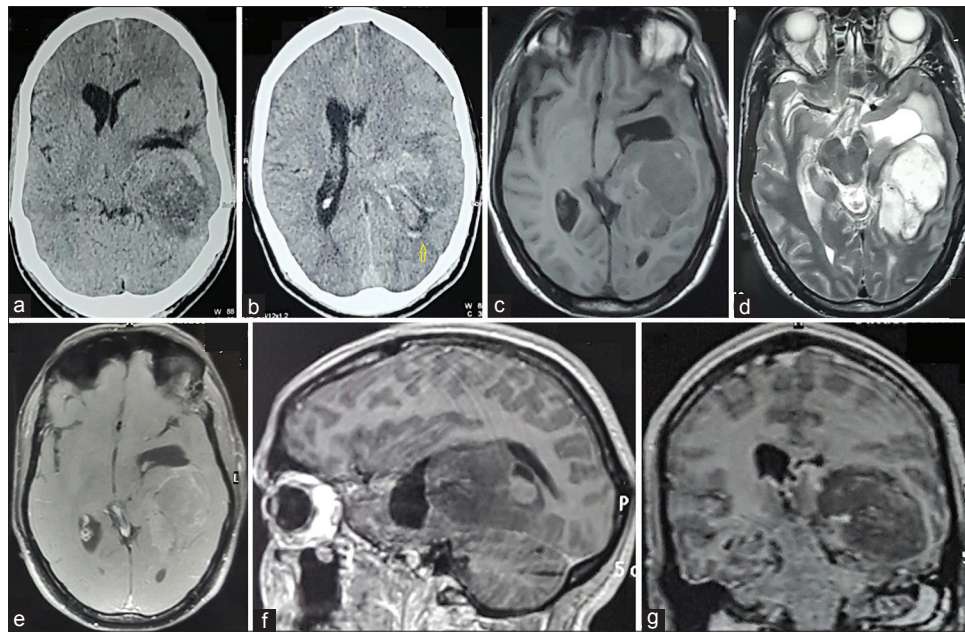
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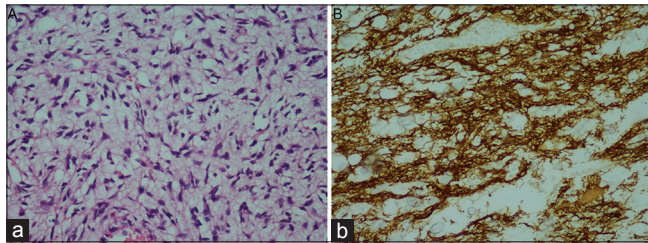
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**Figure 1:** (a and b) Computed tomography shows ill-defined lesion in the left posterior temporal and parietal region with areas of hemorrhage within the tumor. In addition, note the presence of blood in the left lateral ventricle (yellow arrow in b). (c-g) In magnetic resonance imaging, the lesion is isointense on T1 weighted (c), hyperintense on T2 weighted (d) sequence and heterogeneously enhancing on gadolinium contrast (e-g)



**Figure 2:** (a) Photomicrograph showing a moderately cellular tumor consisting of slender astrocytes in a background of myxoid tissue (H and E,  $\times 200$ ). (b) Immunostaining showing glial fibrillary acid protein-positive slender processes (H and E,  $\times 200$ )

young children.<sup>[2]</sup> In 2007, WHO classification designated PMA as Grade II tumors, different from PA owing to its distinct histological picture and relatively poor clinical outcome. A prominent myxoid background and cellular monomorphism with bipolar tumor cells that lack Rosenthal fibers and eosinophilic granular bodies are noticeable features which distinguish PMA from the classical PA.<sup>[1,4]</sup> The cells may also frequently exhibit an angiocentric pattern and nuclear pleomorphism; however, calcification is uncommon.

Mean age at diagnosis varies from 18 months to 7 years, with most cases occurring in infants and young children.<sup>[1,2,4]</sup> In adults, PMAs are only occasional with very few reports in literature.<sup>[4,7]</sup> Other than the usual hypothalamic/chiasmatic region, few cases have been reported in the posterior fossa, spinal cord, and sporadically in the supratentorial location (parietal, temporal, and basal ganglia regions).<sup>[4]</sup>

It is difficult to reliably differentiate PMA from PA due to its shared common location and a similar radiological

picture. However, certain subtle clues may favor a PMA. It is predominantly a solid tumor with less frequent cystic component and shows more homogeneous enhancement, higher chance of leptomeningeal dissemination, and probably an increased propensity to bleed than their PA counterpart.<sup>[8]</sup> No feature is unique to PMA and hence the diagnosis is confirmed only by histopathological studies.

Spontaneous intratumoral hemorrhage is predominantly a feature of high-grade tumors and metastasis, more so in children. Low-grade gliomas with such presentation are only occasionally seen and represent  $<1\%$  of hemorrhagic brain tumors.<sup>[9]</sup> The etiology underlying the bleed in low-grade neoplasms is less understood. Sporadically, PMA and PA can present in such manner.<sup>[7,10]</sup> In recent times, studies have attempted to identify the abnormal vasculature that predisposes to hemorrhage in these low-grade neoplasms.<sup>[9,10]</sup> They have noted thick-walled densely hyalinized vessels, glomeruloid endothelial hyperplasia, and thin-walled ectatic vessels in such cases. Local metabolic factors are also suggested, evidenced by microcalcifications in about 25% of PA.<sup>[9]</sup> Degenerative vascular changes in the form of sclerosed thick-walled and ectatic thin-walled vessels may also contribute to hemorrhage.<sup>[10]</sup> The other possible factors implicated are endothelial proliferation from the associated oligodendroglial component in PA which has a tendency to bleed, rupture of encased aneurysms, dysplastic capillary beds, amyloid angiopathy, and increased vascular endothelial growth factor expression.<sup>[9,10]</sup>

The index case reported is an adult with PMA bleed located in the temporal periventricular area. The presentation has

certain unique features. It differs from its usual occurrence in the chiasmatic/hypothalamic area in the pediatric age group. An additional noticeable feature was the associated hemorrhage (both intratumoral and intraventricular) that was evident on preoperative imaging. Previously, Gottfried *et al.* reported the first case of PMA in temporal lobe that presented with bleed.<sup>[7]</sup> However, the histopathology in their case was mixed, with predominant features of PMA and a small focus of PA. It was unclear whether the PA component had caused the tumoral bleed. In our patient, the histopathology was only that of PMA.

To date, eight cases of PMA with spontaneous hemorrhage have been described.<sup>[3-7]</sup> Noticeably, the majority (75%) were seen in their typical location except for two patients who had tumors outside the suprasellar region (one each in posterior fossa and temporal lobe). The relevant

literature is summarized in Table 1. PMA, as compared to PA, is more likely to display intratumoral bleed. It has been reported in up to 12%–25% of pilomyxoid tumors, whereas PA showed hemorrhage in nearly 8%.<sup>[4]</sup> Steadily, there has been increased recognition of incidence of hemorrhage in these low-grade tumors (PMA and PA).<sup>[3-7,10]</sup> Hence, it is worthwhile to consider them in the differential diagnosis of intracranial tumors presenting with bleed.

The correct identification and differentiating PMA from PA on imaging as well as histology reflect on its outcome.<sup>[1,2]</sup> A suspicion of PMA on preoperative radiology may help in appropriate prognostication of patients. In a follow-up that compared outcomes of PMA with PA, the former showed higher rate of local recurrence (76% vs. 50%).<sup>[1]</sup> The progression-free survival (mean 26 vs. 147 months) and the

**Table 1: Reported cases of pilomyxoid astrocytoma with hemorrhage**

Author (year)	Age, gender	Site	Clinical presentation	Radiology	Management	Intraoperative finding	Follow-up
Gottfried <i>et al.</i> (2003) <sup>[7]</sup>	24, male	Right posterior temporal	Confusion, vomiting, headache, gait instability	Cyst with fluid-fluid levels (hemorrhage), T1 and T2 hyperintense, heterogeneously enhancing solid component with calcifications in medial margin	Gross total resection	Cyst fluid with motor oil consistency, solid-soft, mucinous	6 months, no recurrence
Hamada <i>et al.</i> (2008) <sup>[3]</sup>	5, male	Suprasellar region and third ventricle	Headache, vomiting	Heterogeneously enhancing lesion with partial hemorrhage	Partial resection and adjuvant chemotherapy (cisplatin and vincristine)	Soft, mucinous and easily suctioned	4 months, expired
Linscott <i>et al.</i> (2008) <sup>[4]</sup>	4, female	4 <sup>th</sup> ventricle	Seizure	T1 hypo, heterogenous on contrast with rim enhancement	NA	NA	NA
	46, male	Hypo-thalamus, optic chiasma	Headache and vomiting	T1 hypo-isointense, T2 hypointense core with hyperintense rim, contrast rim enhancing	NA	NA	NA
	9, male	Hypothalamus, optic chiasma, 3rd ventricle, medial temporal lobe	Visual changes	T1 hypo-hyperintense, T2-heterogeneously hyperintense, heterogeneous on contrast with rim enhancement	NA	NA	NA
	21, male	Hypo-thalamus, optic chiasma	Headache	T1 hypo, T2 hypointense core with hyperintense rim, heterogenous on contrast with rim enhancement	NA	NA	NA
Shibahara <i>et al.</i> (2009) <sup>[5]</sup>	18, male	Chiasma and hypothalamus	Loss of consciousness	Subarachnoid (basal and ambient cistern) and intraventricular hemorrhage	Biopsy	NA	NA

*Contd...*



Table 1: Contd...

Author (year)	Age, gender	Site	Clinical presentation	Radiology	Management	Intraoperative finding	Follow-up
Wang <i>et al.</i> (2016) <sup>[6]</sup>	13, male	Suprasellar	Headache, vomiting, visual impairment, coma, endocrine dysfunction	Intratumoral hemorrhage and hydrocephalus	EVD, ventriculoperitoneal shunt, gamma knife radiosurgery, subtotal resection	NA	10 months, no recurrence in MRI, neurology stable, endocrine dysfunction present
Present case	40, male	Left posterior temporal and parietal	Headache, vomiting	T1 iso, T2 hyperintense lesion with intraventricular extension and heterogeneously enhancing	Near total excision and adjuvant radiotherapy	Soft, suckable and moderately vascular	3 months, neurology stable

NA – Not available; EVD – External ventricular drain

overall survival rate were also less compared with that of PA (mean 63 vs. 213 months).

Management of PMA remains controversial and varies with the location.<sup>[1,2]</sup> Surgical resection is the primary treatment modality; however, its occurrence often in the hypothalamic/chiasmatic region precludes complete excision. In the posterior fossa, total excision could be achieved. Adjuvant chemotherapy and radiotherapy are offered in tumor recurrence, partially resected symptomatic cases, and those that progress on follow-up imaging.<sup>[1,2]</sup>

## Conclusion

The report highlights an atypical (nonchiasmatic) site of occurrence of PMA with spontaneous intratumoral hemorrhage in an adult. Occasionally, low-grade tumors may have such unusual presentations. The possibility of PMA has to be borne in mind in such cases. Differentiating PMA from PA has management and prognostic implications.

## Consent

Informed consent was obtained from the patient.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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