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Pilomyxoid Astrocytoma Presenting as Fetal Ventriculomegaly and Obstructive Hydrocephalus in a Newborn: A Case Report

Sreelekshmy K [1], Narayanan E [1], Muthukumaran Natarajan [1]

1. Neonatology, Madras Medical College, Chennai, IND

Corresponding author: Sreelekshmy K, sreelekshmyk94@gmail.com

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Abstract

Pilomyxoid astrocytoma (PMA) is a histologically distinct and biologically aggressive variant of pilocytic astrocytoma (PA), with neonatal-onset presentation being extremely rare. We report a term neonate who presented with refractory seizures and progressive macrocephaly, in whom antenatal ventriculomegaly had been detected at 26 weeks of gestation. Postnatal neuroimaging revealed a large suprasellar mass causing obstructive hydrocephalus, for which an emergency ventriculoperitoneal shunt was performed, followed by subtotal surgical resection. Histopathology showed monomorphic bipolar cells in an angiocentric pattern within a myxoid matrix, with diffuse S-100 positivity and focal perivascular GFAP immunoreactivity, confirming PMA. The postoperative NICU course was prolonged with multisystemic complications, and the infant was discharged on day of life 77. This case highlights that antenatal ventriculomegaly, even when mild, may herald an underlying congenital intracranial neoplasm, supporting consideration of postnatal neuroimaging follow-up. PMA should be considered in the differential diagnosis of a neonate presenting with a suprasellar mass or unexplained obstructive hydrocephalus, and requires histopathological confirmation given significant radiological overlap with classic pilocytic astrocytoma.

Categories: Pathology, Pediatrics, Neurosurgery
Keywords: antenatal ventriculomegaly, congenital hydrocephalus, neonatal brain tumor, pilomyxoid astrocytoma, suprasellar mass

Introduction

Brain tumors in the neonatal period are exceptionally rare, accounting for less than 1% of pediatric malignancies, with an estimated incidence of 1-3 per million live births [1]. Pilomyxoid astrocytoma (PMA) is a histologically distinct variant of pilocytic astrocytoma (PA), generally associated with a more aggressive clinical course, with a marked predilection for the hypothalamic-chiasmatic region in young children, first delineated as a distinct clinicopathological entity in 1999 [2]. Compared with classic PA, PMA carries a higher recurrence rate, greater propensity for cerebrospinal fluid (CSF) dissemination, and significantly shorter progression-free survival [2,3]. The most recent WHO classification of central nervous system (CNS) tumors has refrained from assigning PMA a specific histological grade, reflecting ongoing uncertainty about its biological behavior [4]. Neonatal presentation of PMA is exceedingly rare globally; to the best of our knowledge, no previous report of neonatal-onset PMA from India has been identified [5,6]. This case illustrates the diagnostic value of correlating antenatal imaging abnormalities with postnatal neurological deterioration and histopathological findings, highlighting the diagnostic and therapeutic challenges of managing this neoplasm in a neonate.

Case Presentation

The mother was a 26-year-old second gravida with a previous healthy, alive child, a history of febrile exanthem during the first trimester, and third-trimester anemia necessitating packed red blood cell transfusion. Serial antenatal ultrasonography was unremarkable until a growth scan at 26 weeks and 5 days of gestation identified fetal ventricular dilatation; fetal neurosonography confirmed mild left lateral ventriculomegaly of 11 mm without intracranial hemorrhage or calcification. A subsequent routine antenatal ultrasound at 32 weeks gestation showed mild progression of the ventriculomegaly to 12 mm and postnatal neuroimaging was recommended. Fetal MRI was not done during the antenatal period.

A female neonate was delivered at 38 weeks and 6 days of gestation by normal vaginal delivery, with a birth weight of 2.9 kg and Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. At 40 hours of life, the infant developed generalized tonic-clonic seizures and was commenced on anticonvulsant therapy at the referring hospital before transfer to our tertiary neonatal unit on day 6 of life. On admission, the infant required invasive mechanical ventilation and was hemodynamically stable. Recurrent seizures within hours of admission necessitated escalation to dual anticonvulsant therapy. Neurological examination revealed lethargy, reduced spontaneous movements, hyperreflexia, and a tense, bulging anterior fontanelle. Occipitofrontal circumference (OFC) was 37 cm, above the 97th percentile for gestational age, consistent with rapidly progressive macrocephaly.

Baseline investigations, including sepsis screen, serum electrolytes, CSF analysis (normal glucose and protein, sterile culture), viral polymerase chain reaction (PCR) panel, and CSF Toxoplasma serology, were non-contributory. Cranial ultrasonography demonstrated grossly dilated bilateral lateral ventricles with a Levine ventricular index of 21.7 mm bilaterally, exceeding the 97+4th percentile action line, confirming severe progressive hydrocephalus. Non-contrast cranial computed tomography (CT) demonstrated a mixed-density suprasellar space-occupying lesion causing obstruction at the foramina of Monro with resultant severe bilateral ventricular dilatation, periventricular lucency, and sulcal effacement (Figure *1A*). Following emergency CSF diversion, magnetic resonance imaging (MRI) of the brain characterized a large T1-hypointense, mildly T2-hyperintense, suprasellar mass without diffusion restriction, extending into the bilateral medial temporal lobes, posteriorly to the prepontine cistern abutting the ventral pons, and anteriorly into the anterior cranial fossa, encasing the bilateral middle cerebral, anterior cerebral, and posterior communicating arteries, with associated bilateral cerebral white matter vasogenic edema (Figure *1B*).

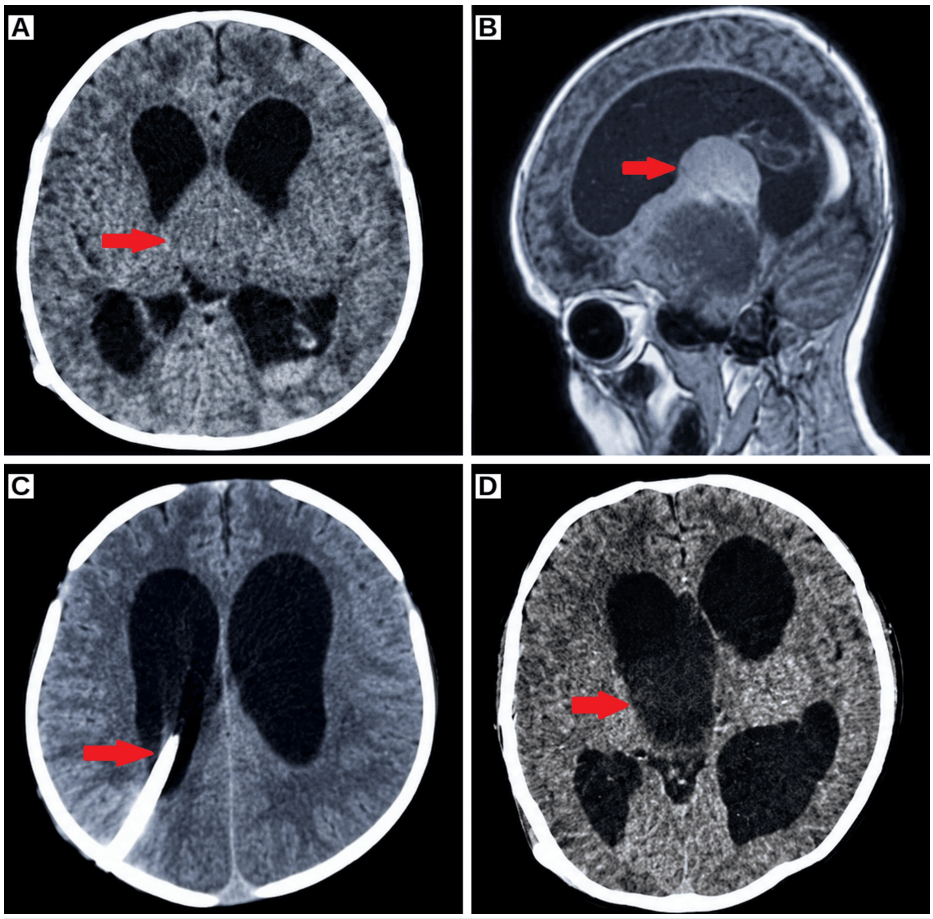


FIGURE 1: Neuroimaging of the suprasellar mass

(A) Non-contrast axial CT of the brain demonstrating a suprasellar mass (red arrow) with resultant dilated bilateral lateral ventricles, illustrating the obstructive mechanism of hydrocephalus. (B) T1-weighted sagittal MRI demonstrating a 46×50×41 mm suprasellar mass (red arrow), delineating its extent and relationship to adjacent structures. (C) Axial CT following emergency cerebrospinal fluid diversion, showing the ventriculoperitoneal shunt catheter tip (red arrow) in situ with interval decompression of the ventricular system. (D) Axial CT following subtotal excision of the suprasellar mass (red arrow), demonstrating reduction in mass volume with a small residual component.

Emergency ventriculoperitoneal (VP) shunt insertion was performed on postnatal day 10 to restore CSF circulation (Figure *1C*). Following clinical stabilization and definitive MRI characterization of the lesion, the case was reviewed at a multidisciplinary tumor board involving neonatology, neurosurgery, neuro-oncology, and pathology, and written informed parental consent was obtained after counselling regarding prognosis and complications. The infant subsequently underwent craniotomy via a pterional subfrontal approach for tumor resection on postnatal day 22. Intraoperatively, the lesion was identified as a grey-white, friable, moderately vascular suprasellar mass with significant associated cerebral edema. Subtotal resection was performed(Figure *1D*); a small residual component was deliberately left in situ where densely adherent to a major intracranial artery, to avoid catastrophic vascular injury. Adjuvant radiotherapy was not administered, as it is generally avoided in children younger than three years of age owing to severe long-term

neurocognitive, endocrine, and cerebrovascular sequelae. Chemotherapy was deferred until interval neuroimaging assessment of the residual lesion.

Histopathological examination of the resected specimen revealed a cellular glial neoplasm comprising monomorphic bipolar cells arranged in a characteristic angiocentric pattern within an abundant myxoid fibrillary matrix, with absence of Rosenthal fibers and eosinophilic granular bodies (Figures *2A*, *2B*). Immunohistochemistry demonstrated diffuse strong S-100 protein positivity (Figure *2C*), focal perivascular glial fibrillary acidic protein (GFAP) immunoreactivity, negative chromogranin staining (Figure *2D*), and a Ki-67 proliferation index of 30-34% - a combined morphological and immunophenotypic profile diagnostic of pilomyxoid astrocytoma according to the WHO Classification of Tumors of the Central Nervous System (2021) [4].

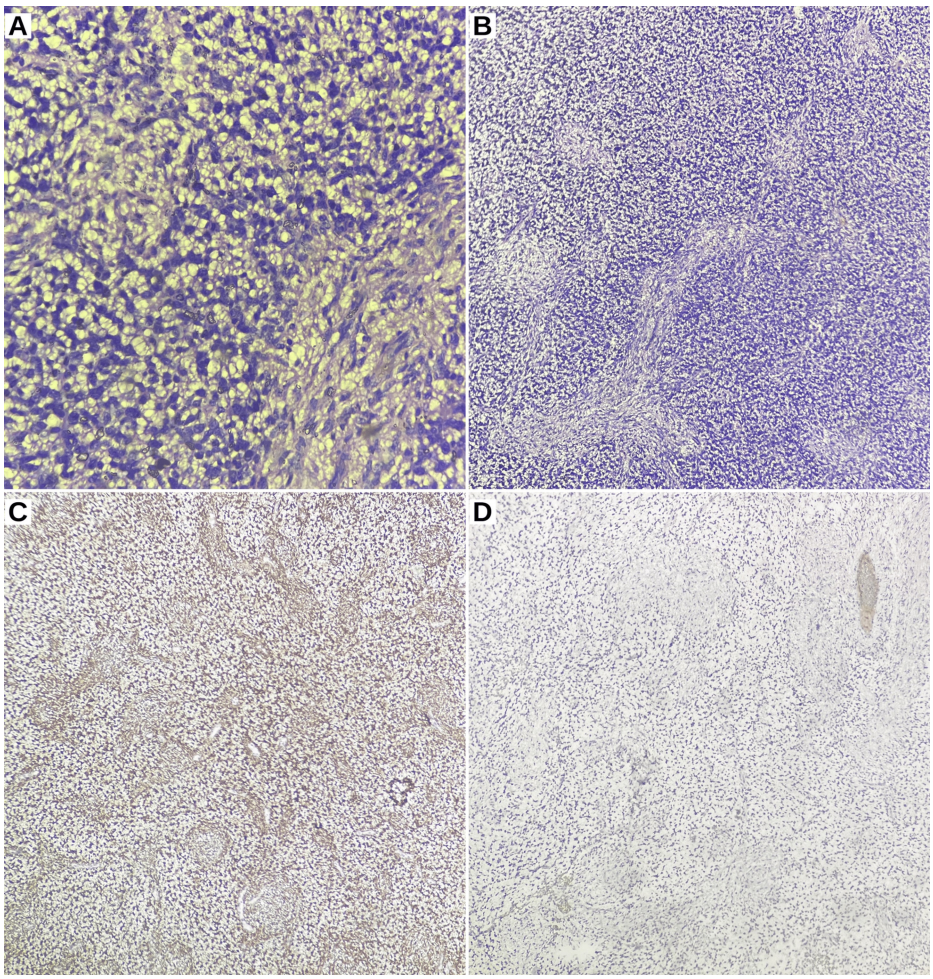


FIGURE 2: Histopathological and immunohistochemical features of pilomyxoid astrocytoma

(A) Hematoxylin and eosin stain (H&E), high-power view (400x), demonstrating monomorphic bipolar tumor cells embedded in a myxoid background with absence of Rosenthal fibers and eosinophilic granular bodies. (B) H& E stain, low-power view (100x), showing a cellular glial neoplasm with an angiocentric growth pattern in an abundant myxoid matrix. (C) Immunohistochemistry (100x) showing diffuse strong S-100 positivity in the tumor cells. (D) Immunohistochemistry (100x) showing negative chromogranin staining.

The postoperative course was prolonged with multisystemic complications requiring intensive multidisciplinary supportive care. These included cerebral edema managed with hypertonic saline, fluid-refractory shock requiring vasopressor support, recurrent seizures requiring escalation through several anticonvulsant agents, disseminated intravascular coagulation (DIC) treated with blood product support, late-onset sepsis, and a secondary surgical site infection, both treated with targeted antibiotics, symptomatic anemia requiring transfusion, and recurrent ventilator-associated pneumonia with delayed extubation. Mechanical ventilator support was required until day of life 55. The infant was discharged on day of life 77 on maintenance anticonvulsant therapy in stable clinical condition. At one month of follow-up since discharge, repeat neuroimaging demonstrated a stable, unchanged residual tumor. Neurodevelopmental assessment was performed and showed warning signs in head control and posture, for

which the infant has been referred for early intervention.

Germline genetic testing by whole exome sequencing (WES) of peripheral blood revealed no pathogenic single-nucleotide variant (SNV) or copy number variant (CNV) associated with known hereditary cancer predisposition syndromes. Somatic tumor molecular profiling was not performed. Follow-up is ongoing under a structured multidisciplinary program encompassing neurosurgery, neuro-oncology, neurology, ophthalmology, clinical genetics, and developmental pediatrics, with planned surveillance neuroimaging to monitor the residual lesion.

The parents received detailed counselling regarding the histological diagnosis of pilomyxoid astrocytoma, its biological behavior and recurrence risk, the rationale and limitations of subtotal resection, and the requirement for long-term oncological surveillance with potential adjuvant chemotherapy. The family demonstrated adequate understanding of the guarded prognosis and provided written consent for the ongoing management plan and follow-up program.

Discussion

The rarity of neonatal brain tumors, with an incidence of 1-3 per million live births, ensures most clinicians will rarely encounter one [1]. The antenatal detection of mild fetal ventriculomegaly at 26 weeks as the earliest sonographic harbinger of the underlying neoplasm is instructive: mild isolated ventriculomegaly (atrial width 10-12 mm), while most commonly benign or structural in etiology, may occasionally represent the earliest detectable manifestation of a congenital intracranial neoplasm and might require postnatal neuroimaging follow-up [7]. PMA represents a small but distinct subset of tumors previously classified as pilocytic astrocytoma, with a predilection for the hypothalamic-chiasmatic region [8]. The rapid postnatal evolution to severe obstructive hydrocephalus within weeks reflects the aggressive biological behavior of PMA, which is associated with higher recurrence rates and shorter progression-free survival than classic PA [2,3]. The prior Indian reports describe PMA in older children [5,6]; the present case is the youngest documented Indian presentation to the best of our knowledge, with antenatal ventriculomegaly as the first clinical harbinger--a pattern not previously reported in this population.

The pathognomonic histological triad of angiocentric monomorphic bipolar cells within a myxoid matrix, with conspicuous absence of Rosenthal fibers and eosinophilic granular bodies, distinguished PMA from classic PA in this case [2,3]. The Ki-67 proliferation index of 30-34% is notably higher than typically reported for classic pilocytic astrocytoma (generally less than 5%), consistent with PMA's intermediate biological aggressiveness, more aggressive than classic PA yet retaining a low-grade phenotype [3]. This index was quantified by immunohistochemistry on the resected tumor specimen, assessed as the percentage of positively staining tumor cell nuclei across multiple high-power fields in areas of highest labeling (hot spots); no additional proliferative features, such as increased mitotic activity or necrosis, were identified on histopathological examination. Neuroimaging-based differentiation of PMA from PA is unreliable, as both present as solid, T2-hyperintense, non-calcified hypothalamic masses without diffusion restriction [9]; histopathological confirmation is therefore mandatory. Negative chromogranin immunostaining argued against neuroendocrine neoplasm, although this finding alone is insufficient to definitively exclude all germ cell tumors; this exclusion was further supported by the characteristic histopathological features described above and the absence of calcification or cystic change on neuroimaging, which also argued against craniopharyngioma. The diagnostic rationale in this case therefore reflects the limitations of the available immunohistochemical panel, and more extensive marker profiling may be warranted in diagnostically ambiguous cases. The absence of pathogenic germline variants on WES is consistent with the predominantly sporadic nature of PMA [3], though this germline finding does not substitute for somatic tumor molecular characterization, which was not performed in this case.

Surgical resection of hypothalamic-chiasmatic PMA in neonates poses unique operative challenges. The tumor's intimate relationship with the optic chiasma, hypothalamus, pituitary stalk, and major cerebral vessels renders gross total resection hazardous; planned subtotal resection is a widely accepted strategy to minimize morbidity while achieving tissue diagnosis [8]. In this case, subtotal resection was the appropriate strategy given the tumor's proximity to major intracranial vessels; the optimal extent of resection in neonatal PMA more broadly should be individualized based on anatomical location and operative risk rather than generalized from a single case. The multisystemic complications encountered postoperatively reflect the profound physiological vulnerability of the neonatal host to major intracranial surgery. Long-term sequelae specifically associated with hypothalamic-chiasmatic PMA include diencephalic syndrome, panhypopituitarism, visual field defects, and neurocognitive impairment, all necessitating lifelong multidisciplinary surveillance [8].

Cranial irradiation is generally avoided below three years of age owing to the severe risks of irreversible neurocognitive, endocrine, and cerebrovascular injury to the immature brain. Chemotherapy, principally carboplatin-vincristine or the TPCV regimen, constitutes the established standard of care for residual or progressive pediatric low-grade glioma in infancy, with five-year overall survival of 86-87% in randomized trial data [10]; it should be noted that these survival figures are derived from a broader pediatric low-grade glioma population, and their direct applicability to neonatal-onset PMA specifically remains uncertain given differences in age at diagnosis and tumor subtype. Molecularly targeted therapy directed at BRAF

alterations, KIAA1549-BRAF fusion, or BRAF V600E mutation has received regulatory approval for subsets of pediatric low-grade glioma [11]; however, safety and efficacy in the neonatal age group remain unestablished. At the population level, outcomes for pediatric brain tumors in India remain constrained by delayed referral, limited access to specialized neuro-oncological services, and the absence of a national registry [12]--challenges reflected in the prolonged and resource-intensive course documented in this case.

Conclusions

This case underscores that antenatal ventriculomegaly, even when mild, can be the earliest harbinger of a congenital intracranial neoplasm. While we advocate for heightened clinical vigilance and consideration of postnatal neuroimaging follow-up in such cases, this recommendation should be interpreted within the context of established fetal ventriculomegaly surveillance guidelines and the clinical presentation of the newborn. Early multidisciplinary involvement spanning neonatology, neurosurgery, neuro-oncology, and pathology remains central to optimizing outcomes in this rare and biologically aggressive tumor. Long-term structured surveillance is essential given the risk of recurrence and the potential for delayed endocrine, visual, and neurocognitive sequelae.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Sreelekshmy K, Narayanan E, Muthukumaran Natarajan

Acquisition, analysis, or interpretation of data: Sreelekshmy K

Drafting of the manuscript: Sreelekshmy K

Critical review of the manuscript for important intellectual content: Narayanan E, Muthukumaran Natarajan

Supervision: Narayanan E, Muthukumaran Natarajan

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. Institutional Ethical Committee,Madras Medical College issued approval 27062026. **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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