

Pilomyxoid Astrocytomas (PMA)

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Pilomyxoid Astrocytoma (PMA)



- Previously classified within the pilocytic astrocytoma (PA) category
- Behave more aggressively than PA
- Was described first time by *Tihan T. Et al in Johns Hopkins, 1999.*
- Has been added to the 2007 WHO Classification of tumors of the central nervous system
- Is a rare WHO grade II tumor
- Has Decreased duration of disease-free survival and higher mortality rates
- infiltration to the surrounding tissues, are usually focal and minimal

PMS



- Occurs in infants and young children, 10-18 month (few cases in adult patients are published)
- Associated with worse outcome as compared to the pathologically related pilocytic astrocytoma
- It is typically located in the hypothalamic/chiasmatic region (also posterior fossa and spinal cord)
- Local recurrence and CSF spreading are frequent
- Origin: peculiar radial glia?



PMA Histological Features

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Table 1.

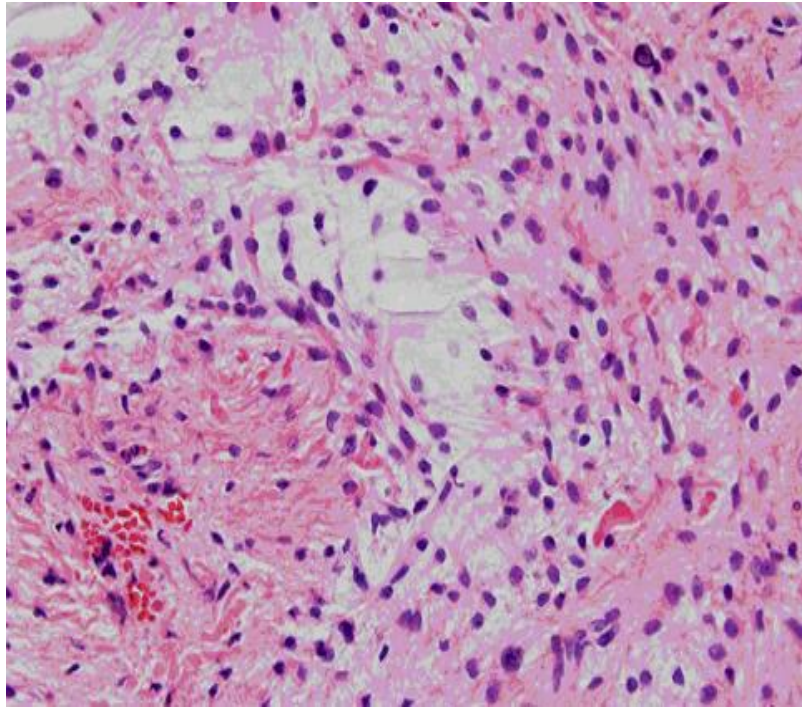
Typical Pilomyxoid and Pilocytic Histologic Features

Feature	Pilomyxoid	Pilocytic
<i>Architecture</i>	Monophasic	Biphasic
<i>Protoplasmic cells</i>	Rare	Present
<i>Rosenthal fibers</i>	Absent	Present
<i>Myxoid background</i>	Predominant	Infrequent
<i>Calcification</i>	Uncommon	Occasional
<i>Eosinophilic granular bodies</i>	Absent	Present
<i>Angiocentric pattern</i>	Frequent	Rare

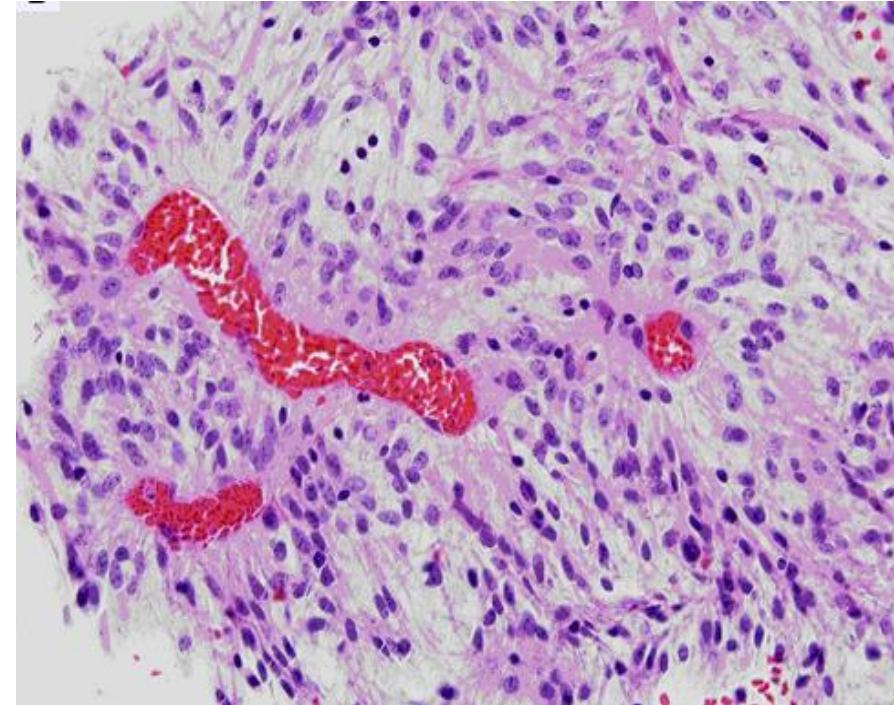
Source: Neurosurg Focus © 2005 American Association of Neurological Surgeons

- Komotar RJ, et al: Pilocytic and pilomyxoid hypothalamic/chiasmatic astrocytomas. Neurosurgery 54:728-30, 2004

PMA Histologic Features



Classic appearance of juvenile pilocytic astrocytoma with characteristic compact and biphasic architecture with densely cellular areas alternating with loose cystic regions with characteristic Rosenthal fibers and eosinophilic granular bodies



The pilomyxoid variant differs in that there is a characteristic myxoid matrix with small compact piloid and highly monomorphic cells. Tumor cells are seen arranged radially around blood vessels.



PMA Symptoms

- Focal neurological symptoms such as visual disturbances and endocrine dysfunction.
- Failure to thrive, developmental delay, altered level of consciousness, vomiting, feeding difficulties, and generalized weakness.
- If it involves the posterior fossa: gait abnormalities, dysmetria, and nystagmus may be present, with tumor extension into the foramen magnum leading to neck stiffness or head tilt.



PMA Radiological Findings

- Commonly isointense on T_1 -weighted sequences, hyperintense on T_2 -weighted ones, and exhibit variable enhancement on MR images with addition of Gd.
- May exhibit peritumoral edema, mass effect, and necrosis
- Less frequently cystic. More often hemorrhagic.
- proton MR spectroscopy in PMA revealed decreased concentrations of total choline, creatine, and N-acetylaspartate. In PA revealed elevated choline and decreased creatine and N-acetylaspartate signals.
- Arterial spin labeling (ASL) perfusion imaging has high accuracy in differentiating PMA from PA

PMA Prognosis



- 33% of patients with PMA died of their disease, compared with 17% of those with PA.
- A local recurrence rate after surgery of 55-76%

PMA Treatment

- Complete resection if possible.
- Chemotherapy. When?
- Radiation?
- Follow up MRI after 6 month





References:

- **Pilomyxoid Astrocytoma: Diagnosis, Prognosis, and Management;** *Ricardo J. Komotar, M.D.; J Mocco, M.D.; Jess E. Jones, B.A.; Brad E. Zacharia, B.S.; Tarik Tihan, M.D., Ph.D.; Neil A. Feldstein, M.D.; Richard C. E. Anderson, M.D.*
- **Differential imaging characteristics and dissemination potential of pilomyxoid astrocytomas versus pilocytic astrocytomas.**
Alkonyi B¹, Nowak J, Gnekow AK, Pietsch T, Warmuth-Metz M.
- **Pilomyxoid astrocytoma in an adult woman: Case report.**
Kibola AH, McClelland Iii S¹, Hlavin J, Friedman JA.
- **Pediatric astrocytomas with monomorphous pilomyxoid features and a less favorable outcome.**
Tihan T¹, Fisher PG, Kepner JL, Godfraind C, McComb RD, Goldthwaite PT, Burger PC.

Thanks



"Whoa! *That* was a good one! Try it, Hobbs — just poke his brain right where my finger is."