Posterior fossa astrocytomas

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• Is it always benign?
• What is the prognosis?
• Surgical pitfalls
• Ethical considerations
Types

- Astrocytomas (location)
- Central neurocytoma
- Oligodendroglioma
- Epidymoma
- Subependymal giant cell astrocytoma (tuberous sclerosis)
- Optic Glioma
- Mixed glioma
WHO Grading of Astrocytomas

**Pathological features**
- Atypia
- Mitosis
- Endothelial proliferation
- Necrosis
- Immunohistochemistry staining

- **Grade I**: none of the features are present
- **Grade II**: one of these features is present
- **Grade III**: 2 of these features are present
- **Grade IV**: 3 or 4 features are present
Pilocytic astrocytoma

- WHO grade I.
- Common in children.
- The most common location is the cerebellum but they can be found in optic chiasm/ hypothalamus regions are often associated with neurofibromatosis 1.
- No infiltration to surrounding tissues
- Radiographically, typically cystic, with a mural nodule, and enhancing.
Pilomyxoid Astrocytoma

• Since 2000 was considered WHO Grade II
• Usually in infant or young children with a mean age 10 to 18 months.
• PMA most typically arises in the hypothalamus or optic chiasm but we had cases in post. fossa too
• Minimal nuclear atypia,
• infiltration to the surrounding tissues, are usually focal and minimal
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 monomorphous cells. Tumor cells are seen arranged radially around blood vessels.
Prognosis

Before PMA was recognized as a separate entity, these tumors were often considered as and treated as a variant of pilocytic astrocytoma. However, PMA has histological features that are quite unique and different from pilocytic astrocytomatas. Most importantly, PMA behaves in a more aggressive manner than pilocytic astrocytomatas. A local recurrence rate after surgery of 55-76%
United States
Approximately 5.4 cases per 100,000 population. Low-grade tumors make up approximately 10-20% of these tumors in adults and 25% in children

Saudi Arabia
12 cases / 100,1000 population
L G glioma 40.6 %
Clinical presentation

Due to the tumor location
- Cerbellar signs
- Visual deterioration
- Hypothalamic signs
- Motor or sensory deficit
- Upper motor neuron signs
- Epilepsy

Due to High ICP/hydrocephalus
- Headache
- Vomiting
- Loss of consciousness
- Increase HC
- Epilepsy
Diagnosis

- Clinical examination
- CT-Scan
- MRI
Differential diagnosis

**Intra axial**
- Astrocytoma
- Glioblastoma
- Brain stem tumors/ cavernous angioma
- Hemangioblastoma
- Ependymoma
- Choroid plexus papilloma
- Metastatic tumor
- Lymphoma
- TB/ abscess

**Extra axial**
- Cystic meningioma
- Epidremoid
- Arachnoid cyst
- Nerve sheath tumors/ Acoustic
- Lipoma
- Leptomeningeal disease
- Glomus Jugulare
Management

- Surgery
- The extend of surgery
- Pre operative preparation
- Anticonvulsant
- Diuretics,
- Steroids
- Postoperative care
- Depends on the extend of excision
- Steroids
• Chemotherapy
• Radiotherapy
• Watching and follow up
Management

• Chemotherapy Cisplatin- Etoposide regimen
• Cispolatin 30 mg/m²/day +
• Etoposide 150 mg/m²/day in ten three-day course
Cytogenetics

A potential cellular factor that may play a role in ONG growth is DNA topoisomerase IIa, an essential nuclear enzyme required for chromatin condensation and chromosome segregation during mitosis. Expression of this enzyme appears to correlate with tumor proliferation. On the other hand, OPGs associated with NF1 have been linked to a defective NF1 gene on chromosome 17q11.2. Neurofibromin is the protein product of the NF1 gene, and acts as a tumor suppressor gene.
PILOCYTIC ASTROCYTOMA
(WHO GRADE I)
• Section from the posterior fossa tumor shows proliferation of predominantly bland looking piloid cells in a fibrillary background.
• Foci showing biphasic pattern with cellular areas alternated by hypocellular areas & microcyst formation is identified.
• Rosenthal fibers, occasional hyaline globules & micro vessel proliferation is present.
• Small foci showing cellular pleomorphism & ischemic necrosis are also identified.
• No mitosis or pallisaded necrosis is seen in the material examined.
• Complex genetic alterations are indicative of a less favorable outcome in low-grade tumors.
• The authors corroborate that $c$-Myc amplification is a marker of poor prognosis in medulloblastomas should be considered as a marker for low grade glioma too!
Cystic walls

Remove or Not remove
Brainstem
High grade astrocytomas

- High-grade astrocytomas (anaplastic astrocytomas and glioblastomas) typically show substantial nuclear pleomorphism and other features of malignancy including increased mitotic activity, necrosis, and endothelial proliferation.
A case had taught me
Conclusion

• Total surgical removal should be the first option of treatment
• Low grade glioma is not always curable by surgery.
• It is very important to reach an accurate histopathological diagnosis
• Chemotherapy is a promising treatment for PMA (pilomyxoid astrocytoma)
• The wall of pilocytic astrocytoma does not contain tumor cells
• Surgery is rewarding for fibrillary astrocytoma in pontine medullary junction
Thank you