Pilocytic astrocytoma

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Key concepts

● a subgroup of astrocytomas (WHO grade I) with better prognosis (10-year survival: > 95%) than infiltrating fibrillary astrocytoma or diffuse astrocytomas

● the most common astrocytic tumor in children

● the average age is lower than for typical astrocytomas (75% of patients are ≤ 20 yrs)

● common locations: cerebellar hemisphere, optic nerve/chiasm, hypothalamus

● CT/MRI appearance: discrete appearing, contrast-enhancing lesion. Cerebellar pilocytic astrocytomas are classically cystic with a mural nodule

● the principal CNS tumor associated with NF1

● pathology: biphasic. 1) Compacted, 2) loose textured astrocytes with Rosenthal fibers and/or eosinophilic granular bodies

● the danger of over grading and overtreating if not recognized. Histology alone may be inadequate for diagnosis; knowledge of patient age & radiographic appearance is critical

● complete surgical resection, when possible, is usually curative. For cystic PCAs with enhancing mural nodule, only the nodule needs to be resected (the cyst wall is not neoplastic). XRT is used post-op only for nonresectable recurrence or malignant degeneration.

Background and terminology

Pilocytic astrocytoma is typically grouped under the circumscribed astrocytic glioma in the World Health Organization Classification of Tumors of the Central Nervous System 2021. The distinction of pilocytic astrocytoma from diffuse glioma is fundamental as it could be the difference between CNS WHO grade 1 and grade 4 glioma [1].

Pilocytic astrocytoma (PCA) is the currently recommended nomenclature for these tumors previously referred to variously as cystic cerebellar astrocytomas and juvenile pilocytic astrocytomas (JPA), among others. Based on location, subtypes include optic gliomas, hypothalamic gliomas, and cerebellar pilocytic astrocytomas. Since treatment decisions vary based on location and neural involvement, it is helpful to discuss differences in the management of these subtypes. PCAs differ markedly from infiltrating fibrillary astrocytomas in terms of their reduced tendency to invade tissue and their very low propensity for malignant degeneration.

Epidemiology

Pilocytic astrocytoma epidemiology.

Etiology

It is driven by aberrant mitogen-activated protein kinase signaling most commonly caused by BRAF gene fusions or activating mutations

It is most frequently caused by KIAA1549:BRAF fusions and leads to oncogene-induced senescence (OIS). OIS is thought to be a major reason for the growth arrest of PA cells in vitro and in vivo, preventing the establishment of PA cultures [2].

Molecular heterogeneity

For tumors originating in the supra-or infratentorial location, a different molecular background was suggested, but plausible correlations between the transcriptional profile and radiological features and/or clinical course are still undefined.

It is a clinically and molecularly heterogeneous disease that occurs most often in the cerebellum and hypothalamic and chiasmatic regions. Classically, pilocytic astrocytomas are driven by the mitogen-activated protein kinase/extracellular signal-regulated kinase pathway.

Genetic aberrations involving this pathway are critical for tumorigenesis. Tandem duplication of 7q34 encodes BRAF and produces several KIAA1549-BRAF novel oncogenic fusions. Activating point mutations of BRAF, such as BRAF (V600E), also lead to pilocytic astrocytoma. Loss of the NF1 gene allows hyperactivation of the oncogene KRAS.

A better understanding of the evolving molecular heterogeneity of pilocytic astrocytomas offers hope for developing molecularly targeted therapeutic armamentariums [3].

Pilocytic astrocytomas of different locations can be precisely differentiated on the basis of their gene expression level, but their transcriptional profiles does not strongly reflect the radiological appearance of the tumor or the course of the disease [4].

Classification

see Infratentorial pilocytic astrocytoma.

Pilocytic astrocytomas in the supratentorial compartment make up 20 % of all brain tumours in children with only 5 % of these arising in the suprasellar region.

Suprasellar pilocytic astrocytomas are challenging to manage surgically with high morbidity rates from surgical resection.

Recent studies suggest that the behavior and biology of WHO grade I pilocytic astrocytomas (PAs) in adults is different than that associated with grade I PAs in children.

see Anaplastic pilocytic astrocytoma.

Clinical features

The presentation depends on the location. In the posterior fossa tumors, there is predominantly a mass effect with signs of raised intracranial pressure, especially when hydrocephalus is present. Bulbar symptoms or cerebellar symptoms may also be present.

see Cerebellar pilocytic astrocytoma.

Diagnosis

Pilocytic Astrocytoma Diagnosis.

Pilocytic Astrocytoma Differential Diagnosis

Pilocytic Astrocytoma Differential Diagnosis.

Treatment

Pilocytic astrocytoma treatment.

Outcome

Pilocytic astrocytoma is a benign low grade tumor with a favorable prognosis.

It is mostly excellent if gross total resection can be achieved, with 10-year survival rates of up to 95%.

see Pilocytic astrocytoma recurrence.

Case series

Pilocytic astrocytoma case series.

Case reports

A previously healthy 10-year-old girl presented with a loss of consciousness following a sudden headache and vomiting. A non-contrast brain computed tomography (CT) scan revealed a massive cerebellar hemorrhage with obstructive hydrocephalus; however, subsequent CT angiography (CTA) showed no vascular abnormalities. An emergency craniotomy was performed to evacuate the hematoma, and histological analysis of the specimen obtained from the tissue surrounding the hematoma revealed a pilocytic astrocytoma (PA). Six months after the ictus, her recovery was scored at 2 on the modified Rankin Scale.

PA can be a cause of critical cerebellar hemorrhage. In this case of a life-threatening massive hematoma, CTA was useful to exclude a major vascular pathology and to save time [5].

References

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