Astrocitoma pilocítico

Tumor neuroepitelial circunscrito, grado 1 en la clasificación de la OMS.

Epidemiología

Es un tipo común de tumor cerebral en la población pediátrica, ya que forman un 19,1% de todos los tumores cerebrales pediátricos en el grupo de 0 - 14 años. [Last accessed on 2012 Mar 23]. Available from: http://www.cbtrus.org/reports/reports.html, y son los tumores de cerebelo más frecuentes en niños (Burger y col., 2000; Farwell y col., 1977).

Pueden afectar a todo el neuroeje, pero el cerebelo es el sitio más común de origen.

El 15 % de los pacientes con neurofibromatosis tipo 1 desarrollan un astrocitoma pilocítico.

Localización

85 % se encuentran infratentoriales y 15 % supratentoriales.

Si bien esta neoplasia puede localizarse en cualquier topografía del neuroeje, ocurre más comunmente en el cerebelo, nervio óptico, la región hipotálamo-quiasmática, mesencefálica.

La localización en ganglios basales es menos habitual.

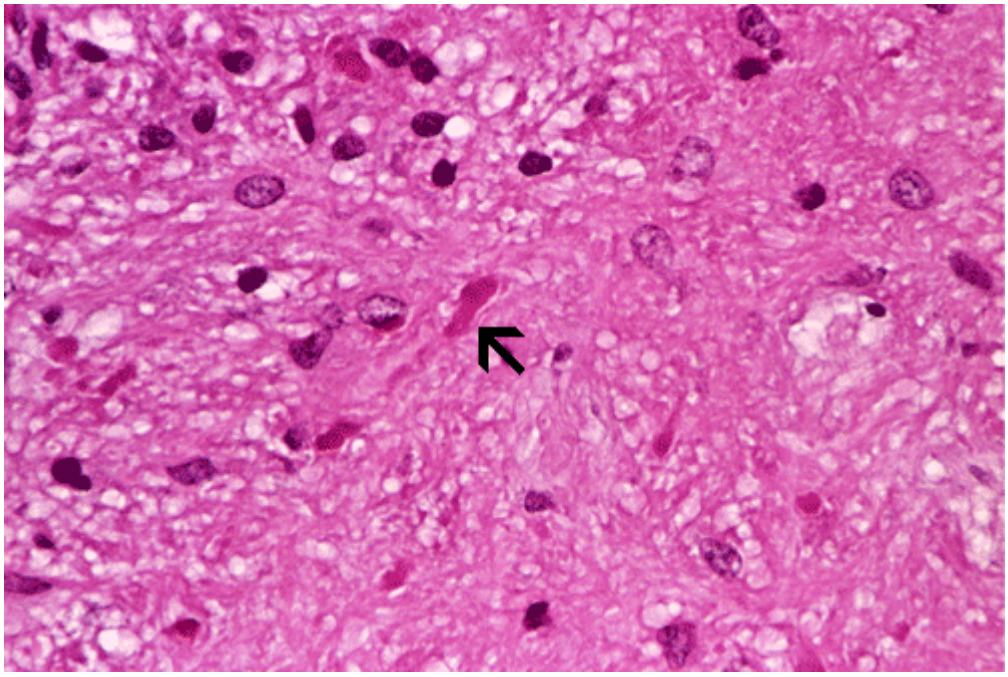
Astrocitoma pilocítico de cerebelo.

Astrocitoma pilocítico de nervio óptico.

Astrocitoma pilocítico hipotalámico.

Astrocitoma pilocítico medular.

Anatomía Patológica



El estudio de estos tumores plantea la dificultad de poder definir los criterios mínimos diagnósticos para el diagnóstico de la entidad.

Varios grupos han reportado una serie de tumores astrocíticos con características comunes a los astrocitomas pilocíticos pero con algunas notables diferencias que justifican su análisis por separado.

Esencialmente, estas neoplasias exhiben una patente monomorfa que ha sido llamada pilomixoide y predomina una densa trama de fibrillas gliales con abundantes fibras de Rosenthal.

La presencia de proliferación vascular en la histopatología son hallazgos posibles en el astrocitoma pilocítico, y al contrario que en otros gliomas, no son indicativos de malignidad.

Se ha descrito un Glioma Metacrónico, multicéntrico de astrocitoma pilocítico con componente de oligodendroglioma a través de distintas aberraciones genéticas (Kanoke y col., 2012).

Se sospecha que puede haber casos previamente diagnosticados de astrocitoma pilocítico y que en realidad son tumores neuroepiteliales disembrioplásicos y en cualquier caso, en presencia de un tumor cerebeloso con características de astrocitoma pilocítico, la posibilidad de una variante compleja de tumor neuroepitelial disembrioplásico se debería tener en cuenta (Vaquero y col., 2012).

Heterogeneidad molecular

La vía de señalización de las proteínas quinasas activadas por mitógenos MAPK, es crítica para su formación.

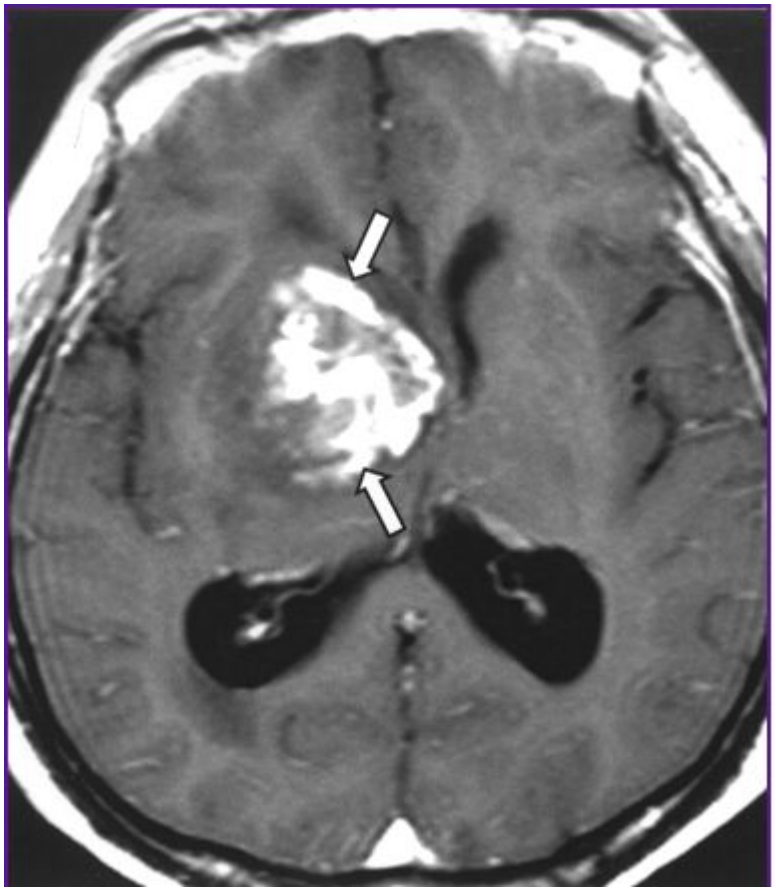
El gen BRAF se encuentra situado en el brazo largo del cromosoma 7 (7q34) y codifica una quinasa serina/treonina citoplasmática de la familia RAF que, al igual que RAS, RET y TRK es miembro esencial de la ruta de señalización de proteínas quinasas activadas por mitógenos (MAPK).

La pérdida del gen NF1 permite la hiperactivación del oncogén K-RAS (Sadighi y col., 2013).

Clínica

Tiene un curso clínico relativamente indolente.

Diagnóstico



En la resonancia magnética, es habitualmente un tumor sólido-quístico bien delimitado, iso-hipointensas en ponderación T1 e hiperintensas en ponderación T2.

Tienen realce intenso pero heterogéneo.

Diagnóstico diferencial

El astrocitoma pilocítico puede tener un componente de like-oligodendroglioma, sin embargo, el diagnóstico diferencial de los oligodendrogliomas puede ser difícil cuando este componente es mayoritario en el tumor. El astrocitoma pilocítico es immunoreactivo a la GFAP y Olig2, pero no a la Sinaptofisina, EMA, o IDH 1(Utsuki y col., 2011).

Tratamiento

Según los casos, el seguimiento clínico y radiológico de la lesión puede ser razonable. La cirugía puede ser curativa si se consigue una resección completa del tumor.

La radioterapia adyuvante después de cirugía puede ser necesaria en los pacientes de más edad, con resecciones incompletas o con otros factores de riesgo.

Pronóstico

Existen reportes recientes que mencionan la diseminación por el líquido cefaloraquídeo (LCR) implantes espinales y evolución anaplásica.

Sin embargo, estos desarrollos son excepcionales y el pronóstico global de los pacientes es bueno con sobrevidas del 80 al 100% a los 10 años.

De los análisis preliminares de sobrevida surge la impresión que estos tumores tienen una tasa de recurrencia y diseminación por el LCR mayor que la de los astrocitomas pilocíticos convencionales.

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