Xantoastrocitoma pleomórfico (XAP)

Es un tumor de estirpe astrocitaria considerado grado II de la OMS, excepto en los casos con alto índice mitótico o necrosis.

De acuerdo a la nueva clasificación de la OMS de los tumores cerebrales, el XAP pertenece al grupo de los astrocitomas circunscritos (localizados o no infiltrativos), junto con el astrocitoma pilocítico y el de astrocitoma subependimario de células gigantes.

Existe la variante de Xantoastrocitoma pleomórfico anaplásico.

Historia

Descrita por primera vez en 1979, del que han sido descrito 100 casos aproximadamente.

Epidemiología

Muy poco frecuente que afecta con mayor frecuencia a adultos jóvenes.

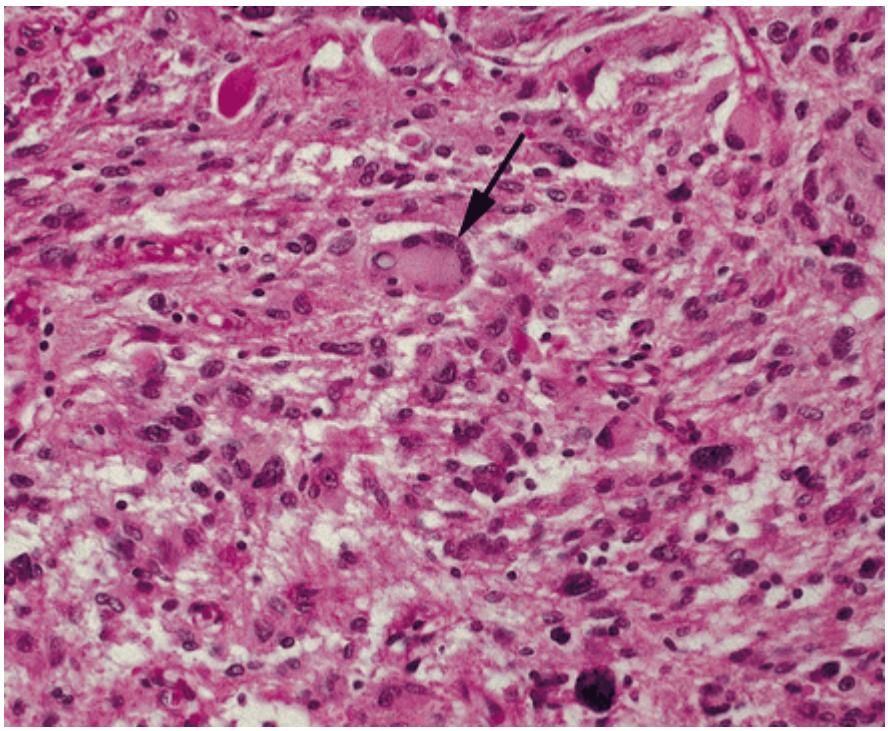
Constituye menos del 1% de todos los astrocitomas cerebrales y en el 98% están en situación supratentorial. Parecen originarse de astrocitos subpiales o de células precursoras multipotenciales neuroectodérmicas.

Surgen con mayor frecuencia a nivel supratentorial en los lóbulos temporales y parietales de los niños y adultos jóvenes con historia clínica de crisis convulsivas, aunque hay reportes de localizaciones poco frecuentes como el cerebelo y la médula espinal (Mano 2009).

Solo existe un caso publicado a nivel pineal (Thakar y col., 2011).

Anatomía patológica

Macroscópicamente se caracterizan por ser sólidos o quísticos con nódulo mural.



El cuadro histológico puede no presentar abundancia de xantomización citoplásmica, y combina pleomorfismo e hipercelularidad con escasez de mitosis y ausencia de necrosis. Aunque bien vascularizada, la neoplasia no presenta proliferación endotelial glomeruloide.

Neoplasia bien circunscrita que está constituida por células astrogliales pleomórficas que ocasionalmente presentan xantomización citoplásmica. Aparece un río entramado vascular de paredes escleróticas con infiltrados inflamatorios cónicos inespecíficos. En múltiples zonas del tumor, las células adoptan un patrón de crecimiento algo fusiforme con aislados elementos de apariencia histiocitaria. A pesar de la atipia, las mitosis son escasas y no se observa necrosis. Se observa positividad intensa para la proteínas glial fibrilar ácida y S-100, y focal para la sinaptofisina.

Microscópicamente está constituido por células heterogéneas, que varían de células poligonales sin procesos citoplásmicos, células fusiformes, células gigantes multinucleadas y células lipidizadas. Un pequeño número de casos ha mostrado células ganglionares atípicas, aunque no está claro si se trata de tumores de colisión o componente astrocítico en gangliogliomas (Sugita 2009).

Se cree que los xantoastrocitomas pleomórficos derivan de los astrocitos subpiales, ya que los estudios ultraestructurales han demostrado la presencia de una membrana basal que rodea las células neoplásicas y la abundancia de fibras de reticulina.

Clínica

La sospecha de encontrarnos ante esta entidad empieza en la historia clínica del paciente, puesto que se trata de pacientes jóvenes, que refieren crisis focales, y que radiológicamente presentan tumores típicamente periféricos, a veces quísticos, temporales o parietales (Okazaki 2009).

Diagnóstico

Las características de los estudios de imagen son las de una masa intracortical hemisférica en

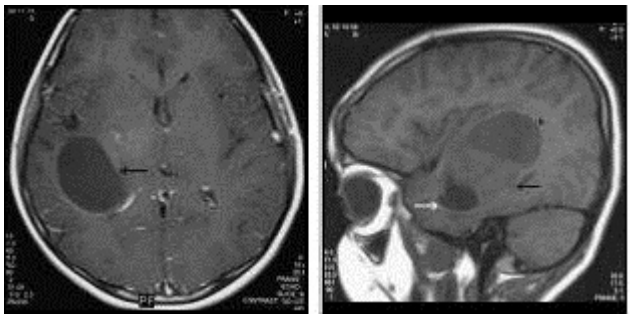
situación periférica, bien delimitada, con un componente quístico y de nódulos murales, generalmente protruyendo en la meninge.

El componente quístico es hipointenso en T1 e hiperintenso en T2 y FLAIR.

El componente sólido tiene generalmente una señal intermedia en todas las secuencias con un realce moderado a intenso tras el contraste.

La masa puede deformar las meninges adyacentes que se realzan de manera intensa (similar a la cola dural). Puede asociarse displasia cortical. En el diagnóstico diferencial debe incluirse el ganglioglioma, meningioma y astrocitoma tanto de bajo grado como anaplásico (Fu, Yong-Juan 2009).

Los gangliogliomas tienen nódulos murales similares a los XAP, pero no se encuentran adyacentes a las meninges y no exhiben una reacción de estas similar a la de la cola dural.



1.Imagen sagital spin-eco potenciada en T1 que muestra una masa circunscrita en situación témporo-insular derecha con un componente quístico hipointenso

2.Imagen axial spin-eco potenciada en T1 con contraste que muestra el componente quístico de la masa con un moderado realce periférico (flecha).

Tratamiento

Resección máxima posible.

En caso de grado III RT o QT.

Pronóstico

Los XAP son tumores generalmente bien definidos y de crecimiento lento.

La infiltración parenquimatosa y de los espacios de Virchow-Robin no están relacionados con un peor pronóstico, aunque la presencia de hiperplasia endotelial, mitosis, atipia y necrosis indican progresión anaplásica y una conducta biológica agresiva.

Es importante reconocerlo porque se trata de un tumor menos agresivo de lo esperado teniendo en cuenta su agresivo aspecto histológico.

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