Ganglioglioma with anaplastic transformation

Ganglioglioma com transformação anaplásica

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A nine year-old male had refractory seizures for four years. Diagnostic imaging (Fig 1A and B) demonstrated a cortico-subcortical focal lesion at the isthmus of the right cingulate gyrus. Biopsy yielded diagnosis of ganglioglioma (Fig 2A). Reoperation one year later showed similar features, but included atypical and multinucleated cells (Fig 2B).

At the age of 13, the patient had symptom recurrence. Diagnostic imaging (Fig 1C and D) demonstrated a heterogeneous enhanced lesion in the right parietal region, with necrosis. Histology (Fig 2C to F) revealed a malignant glial tumor with the appearance of glioblastoma multiforme. A few tumor cells were positive for chromogranin. A diagnosis of malignant transformation of ganglioglioma was made.

Gangliogliomas are rare tumors predominating in the early decades of life, with strong association with long term intractable epilepsy[1-3]. They are composed of variable proportions of glial (mainly astrocytic) cells and mature or dysplastic neurons[2]. Malignant change is a rare, but well recognized, complication. Transformation of the glial component from the low grade to a higher grade is observed in most cases. Also, there is a case of malignant transformation secondary to degeneration of the neuronal component into a neuroblastoma[4]. Some reports in literature[5] suggest that radiation may predispose to malignant degeneration. In this case, the patient did not receive postoperative radiation.

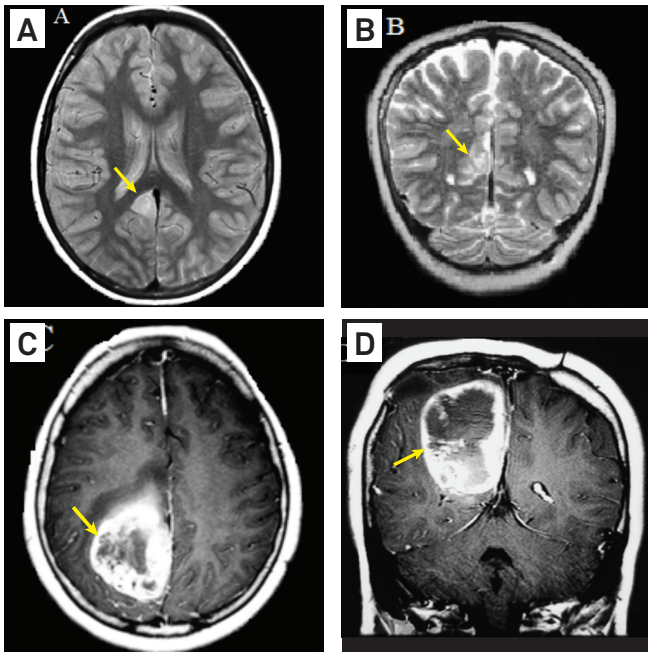

Fig 1. (A) Axial proton density: a small well circumscribed lesion in the right cingulus; (B) Coronal T2: a small lesion in the right cingulus, with high signal intensity in the mass. (C) and (D) Axial and coronal contrast-enhanced, 5 years after: a large mass in the right parietal region, with heterogeneous enhancement and component of necrosis.

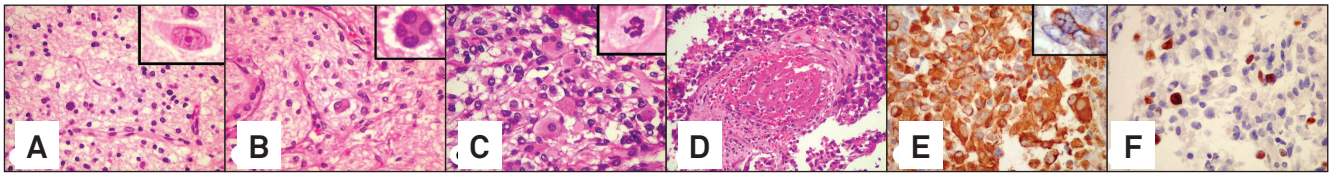

Fig 2. Neuropathology. (A) First biopsy. Moderately cellular low grade glial tumor with scanty atypical cells and thin capillaries. No mitotic figures or necrosis. HE x 100. Inset. Neuron in deep area of the tumor. HE x 400. (B) Second biopsy, one year later. It keeps same features as original specimen. At center, atypical neuron with eccentric nucleus. HE x 100. Inset. Aberrant cell with four nuclei, lineage uncertain. HE x 400. (C–F) Third biopsy. (C) Highly cellular tumor with moderate atypia. HE x 100. Inset. Atypical mitotic figure. HE x 400. (D) Abnormal vessel with thickened walls and occlusive thrombosis. HE x 100. Necrotic areas were present nearby. (E) Tumor cells are strongly positive for glial fibrillary acidic protein indicating astrocytic lineage. X 100. Inset. Isolated tumor cell positive for chromogranin (suggests neuronal differentiation). X 400. (F) About 10% of nuclei were marked by Ki-67 (mib1) antibody. X 100.

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