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Supratentorial primitive neuroectodermal tumor (PNET)

An uncommon location

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A 10-year-old girl with headache. Diagnostic imaging demonstrated a right frontotemporal solid-cystic lesion, in the cortex and white matter, with a component with decrease in diffusion, attributed to high cellularity and nuclear-to-cytoplasmic ratio, which may be seen in PNET or lymphoma[1,2]. A high signal component on T1 and on T2 was observed (extracellular methaemoglobin), often seen in PNET.

Histological analysis led to the diagnosis of PNET .

Supratentorial PNETs are very rare[2]. More than 50% occur in the first 5 years of life[2]. Our case also merits attention because of the late age at presentation.

REFERENCES

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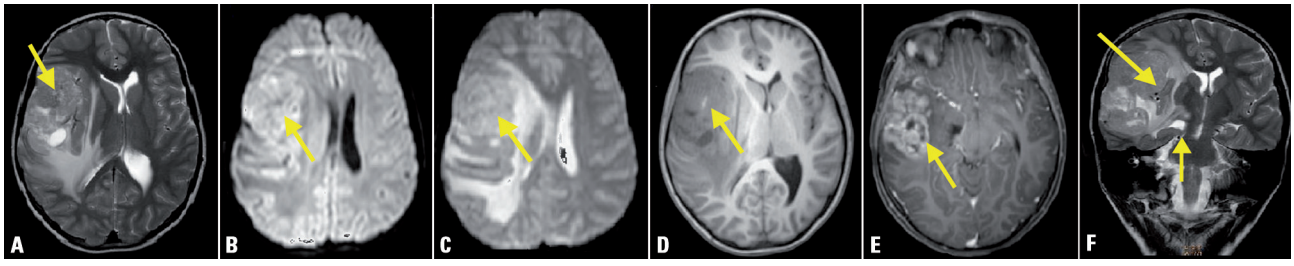

Fig 1. [A] Axial T2 weighted image (WI) demonstrates a large right frontotemporal solid-cystic mass. On DWI [B] and ADC map [C] there is decreased diffusion in the lesion (arrows) (bright on DWI and low intensity in the ADC map). Axial T1 precontrast [D] showed a heterogeneous mass, with a focal high signal (arrow) component (methaemoglobin) and [E] postcontrast T1 shows a heterogeneous enhancing mass. [F] Coronal T2 WI demonstrates heterogeneously low signal (arrow) and there is also right uncal herniation (small arrow).

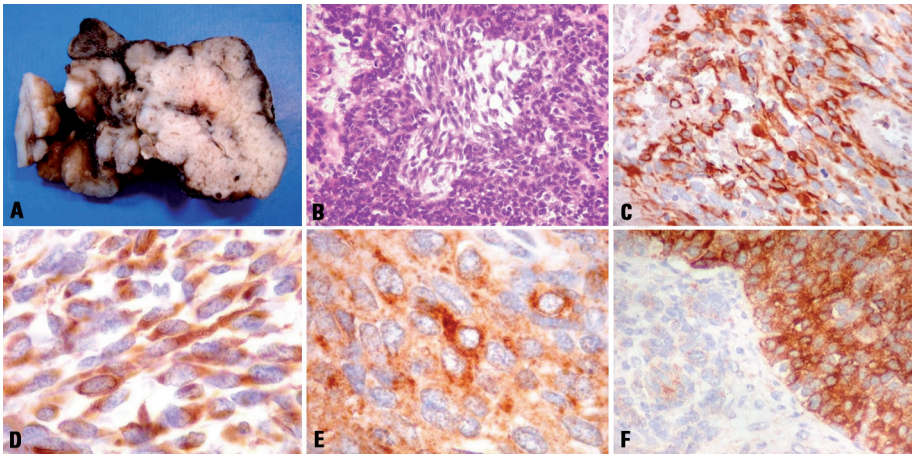

Fig 2. Neuropathology. A primitive-looking neoplasm with immunohistochemical evidence of differentiation along various neuroectodermal cell lines was consistent with the diagnosis of PNET (primitive neuroectodermal tumor). [A] Surgical specimen measured 8 × 4 × 4 cm and weighed 75 g. Well delimited, lobulated firm mass, with a pale pink cut surface. [B] On HE stain, tissue was composed of small undifferentiated cells in solid lobules, with scattered islets of loosely arranged cells, creating a vague resemblance to desmoplastic medulloblastoma. [C] GFAP- positive cells were interspersed with negative elements. [D] Cytoplasmic staining for nestin, an intermediate filament expressed by immature cells, witnessed the primitive nature of the tumor. [E] Positivity for synaptophysin in the cytoplasm of several cells, some displaying cytoplasmic processes reminiscent of neurites, indicated early neuronal differention. [F] Areas of compactly arranged cells were strongly marked for EMA (epithelial membrane antigen), a common feature of ependymal cells. Adjoining loose cells were negative, creating an organoid pattern.

TUMOR NEUROECTODÉRMICO PRIMITIVO SUPRATENTORIAL: UMA LOCALIZAÇÃO INCOMUM

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