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Images in Neuroscience: Question

Macrocephaly and right arm disuse in a 7-month-old boy

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1. Clinical background

A 7-month-old boy presented with macrocephaly (head circumference of 52 cm, 2.5 standard deviations above the 98th percentile). An examination of his cranial contour demonstrated prominent dilated scalp vessels, an elevated and full anterior fontanelle, and mild splaying of the coronal and sagittal sutures. He maintained his right hand in a fist position, and would not reach towards objects or faces with his right hand; his right upper extremity was also significantly hypertonic. A neurological examination of his cranial nerves and other limbs was otherwise normal. His medical history was unremarkable. A cranial ultrasound scan demonstrated a markedly enlarged left lateral ventricle with a round, well-defined hyperechoic structure (Fig. 1A). Contrast-enhanced MRI was then performed (Fig. 1B, D–G).

2. The most likely diagnosis is

- A. Atypical teratoid/rhabdoid tumor
- B. Desmoplastic infantile ganglioglioma
- C. Supratentorial primitive neuroectodermal tumor
- D. Teratoma

Answer on page 304.

Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

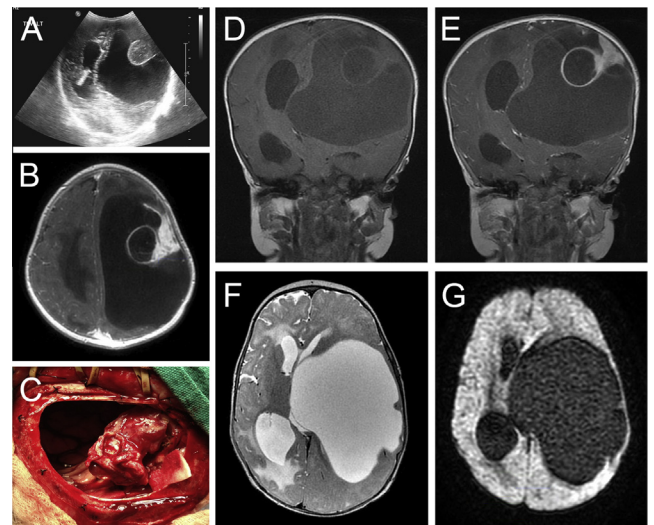


Fig. 1. (A) Ultrasound imaging demonstrates a markedly enlarged left lateral ventricle with a well-defined hyperechoic nodular lesion. (B) Axial and (D, E) coronal T1-weighted MRI demonstrates a large complex cyst within the left cerebral hemisphere. (F) This cyst is isointense compared to cerebrospinal fluid on T2-weighted MRI, and (G) Diffusion-weighted imaging does not demonstrate restricted diffusion. There is an avidly enhancing lesion attached to the dura with an additional cystic component: (D) pre-contrast, (B, E) post-contrast, (C) intraoperative visualization following large cyst drainage.

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