November 2003: Neonatal female with congenital brain tumor

Contributed by: L. F. Bleggi-Torres, MD, PhD, MIAC[1,2,3]; L. Z. B. Pope, MD[1]; A. Koerbel, MD[4]; C. V. Grande, MD[4]; J. C. Araújo, MD[4]

[1] Department of Pathology (Neuropathology), Universidade Federal do Paraná, Curitiba, Brasil.
[2] Laboratory of Pathology, Hospital Nossa Senhora das Graças (HNSG),Curitiba, Brazil.
[3] Laboratory of Experimental Pathology, PUC-PR, Curitiba, Brazil.
[4] Neurosurgical Unit, HNSG, Curitiba, Brazil.

CLINICAL HISTORY

A 33-year-old pregnant female patient was admitted to hospital with a history of 32 weeks gestation. There were signs of severe fetal suffering and an unusually large fetal skull was detected in ultrasound examination. A cesarean section was performed and a baby girl was delivered with cyanosis, vomiting and irritability. After basic neonatal procedures, neurosurgical evaluation revealed a large skull with congested and prominent superficial vasculature, prominent eyes, and a bulging anterior fontanel with wide open sutures. CT scan showed a large partially cystic mass that occupied approximately half of the intracranial space. Marked hydrocephalus was also noted. A neurosurgical procedure was performed and several fragments of the tumor were submitted for neuropathological examination. The baby expired at age 2 months and an autopsy examination was not performed.

MICROSCOPIC DESCRIPTION

Histologic sections showed a tumor composed by a uniform population of large cells with minimal cell-to-cell variability immersed in a fibrillary background (Figure 1). The cells showed glassy, eosinophilic cytoplasm (Figure 2). Nuclei were usually eccentric with small nucleoli (Figure 3). Mitotic figures, necrosis, vascular proliferation, and perivascular lymphocyte cuffing were not seen. Immunohistochemical staining for GFAP (Figure 4) and vimentin (Figure 5) were strongly and diffusely positive. Immunostains for EMA, actin (HHF-35) and cytokeratin were negative.

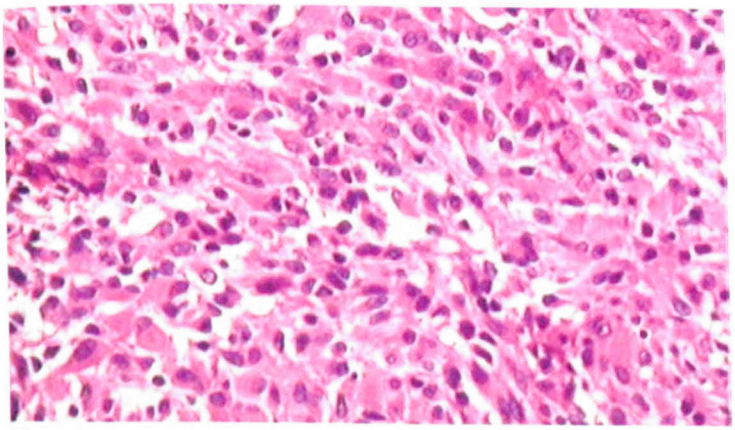

Figure 1.

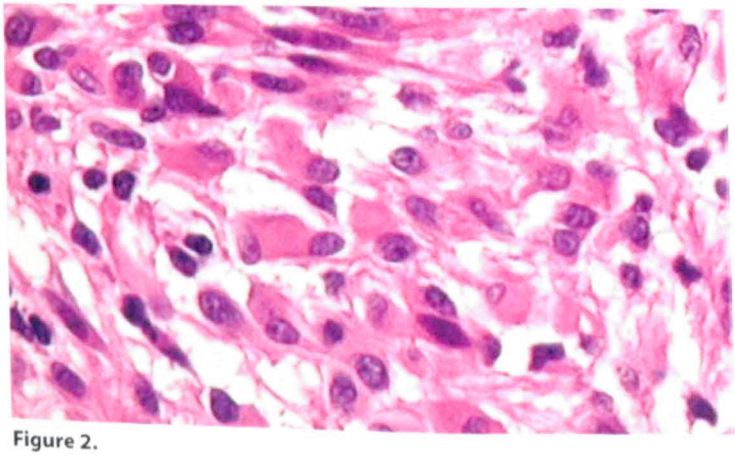

Figure 2.

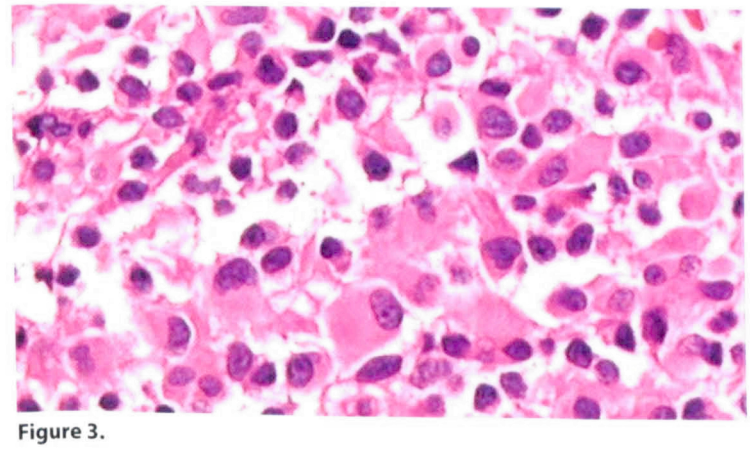

Figure 3.

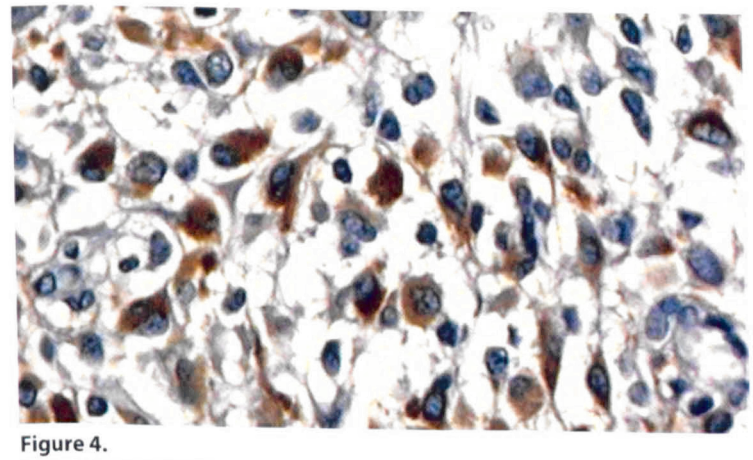

Figure 4.

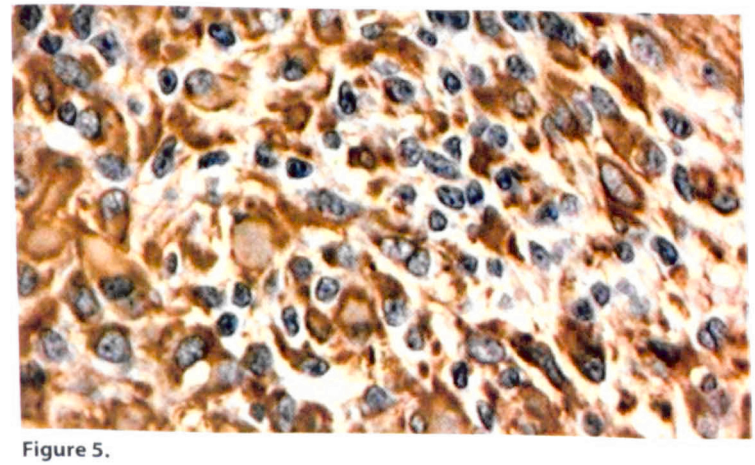

Figure 5.

DIAGNOSIS

Congenital gemistocytic astrocytoma.

DISCUSSION

Intracranial congenital tumors (ICT) are rare. They comprise only about 0.5% to 1.5% of those detected during childhood (7). Arnstein et al (1) first reviewed brain tumors of the newborn in 1951, and they defined neonatal brain tumors as those that developed up to 60 days after birth. Since then, many authors reported such cases according to this definition (1, 2, 4, 7).

Dystocia secondary to the increased head size often results in cesarian sections in these cases. The infants may show vomiting, seizures, and irritability. ITC are most commonly supratentorial, as seen in this case.

The most common ICT are teratomas, followed by medulloblastomas, astrocytomas, choroid plexus papillomas, ependymomas and rarely ependymoblastomas.

The prognosis is poor in most ICT due to the large size of the tumor, especially teratomas, as well as the poor general conditions of the baby at presentation. Histologic malignancy is also assumed to be another complicating factor (4, 7). Our case had a large size and the histologic diagnosis was gemistocytic astrocytoma (grade 2: WHO). Gemistocytic astrocytomas are particularly prone to progress to anaplastic astrocytoma and even glioblastoma (5), although such changes were not detected in our case. Gemistocytic astrocytomas account for 9% to 24% of all astrocytomas (3, 7) and affect mainly adult patients. To date this seems to be the first report of congenital gemistocytic astrocytoma.

ACKNOWLEDGMENTS

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