

CASE OF THE MONTH: January 1999 - Fetus with Echogenic Mass in Third Ventricle

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Clinical History and Radiology

This was a 29-week gestational age newborn male born after prolonged premature rupture of membranes. Two weeks before birth, a routine prenatal ultrasound had shown moderate hydrocephalus and distorted intracranial anatomy, with an echogenic mass in the third ventricle. Organized thrombus and tumor were considered in the differential diagnosis. At delivery he was limp, bradycardic, and had Apgar scores of 3 and 5 at one and five minutes. A CT scan of the head revealed extensive hemorrhage, parenchymal infarction, and numerous calcifications (Figure 1). He was intubated and placed on mechanical ventilation. Multiple decompressive ventricular taps showed hemorrhagic cerebrospinal fluid (CSF) with no evidence of infection. No malignant cells were detected on CSF smears. Toxoplasma and RPR serologies were negative. Despite supportive treatment, his condition progressively deteriorated and he died four days after birth.

Gross and microscopic description

At postmortem examination the head was enlarged (head circumference, 42 cm). The brain weighed 315 gm (expected for body size, 180 +/- 34 gm) and had marked distortion of the architecture. There was severe hydrocephalus and the cerebral hemispheres were massively dilated, with only a thin rim of apparent cortical tissue (Figure 2). Beneath this rim was a mass composed of blot clot and soft, pale tissue measuring approximately 10 x 6 x 3 cm. The exact site of origin of this tumor

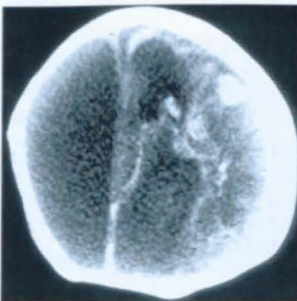


Figure 1.

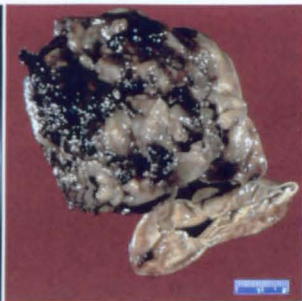


Figure 2.

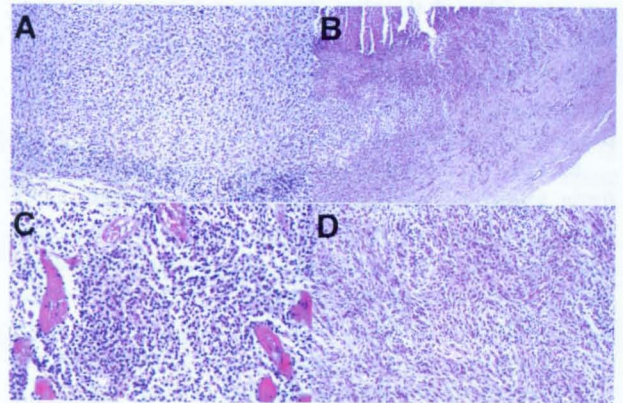


Figure 3.

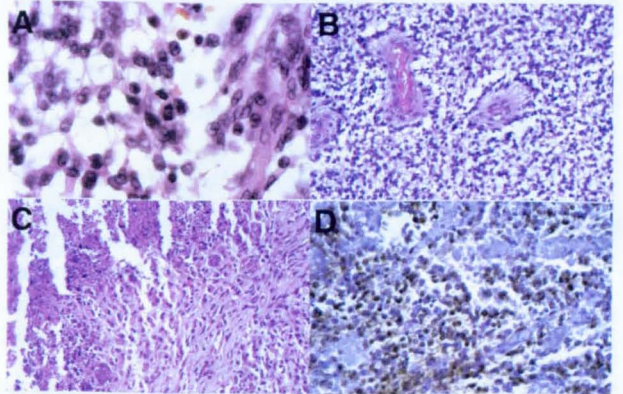


Figure 4.

was unclear due to extensive destruction of adjacent structures. The brainstem and cerebellum appeared orange-stained, rubbery and firm.

Histologic examination showed neoplastic tissue involving lateral and third ventricles, supratentorial brain parenchyma (Figure 3a) and meninges (Figure 3b). The neoplasm was highly cellular, composed of small round to ovoid cells with hyperchromatic nuclei and little cytoplasm (Figure 3c). Some areas exhibited more compact spindle cells (Figure 3d). Mitotic figures and pleomorphism were not prominent (Figure 4a). Occasional pseudorosettes were seen (Figure 4b). There was extensive hemorrhage with areas of necrosis surrounded by multinucleated giant cells (Figure 4c). The tumor cells expressed S-100 (Figure 4d) and focally, synaptophysin and GFAP. Cerebellum was not involved by tumor.

Diagnosis

Congenital Cerebral Primitive Neuroectodermal Tumor (PNET)

Discussion

Congenital neoplasms of the central nervous system are rare, although they have been reported since the mid-19th century (1). They represent only 0.5-1.9% of all pediatric brain tumors (2). In recent years, modern imaging studies such as MRI and CT have allowed early diagnosis of this group of brain tumors.

Review of the literature revealed that medulloblastoma to be the most common type of primary CNS tumor in infants and stillborn fetuses (3), although some series have reported teratoma as the predominant tumor in the fetal and perinatal period (4, 5, 6). Congenital PNETs are rare in the fetal and perinatal period. Most arise from the cerebellar midline, and if large enough, they can extend into the supratentorial compartment (7). The same infratentorial distribution has been reported in older children and with other tumors (8).

Radiologically, the tumors are seen as solid or heterogeneous masses with associated hemorrhage and hydrocephalus. Calcification can be identified by CT scan (2). The presence of hyperechoic areas by ultrasound in suggestive of necrosis (7).

The pathologic findings in this case were indicative of a poorly differentiated congenital neoplasm consistent with a PNET, involving lateral ventricles, brain parenchyma, and meninges with extensive hemorrhage. The strong positive immunohistochemical staining for S-100 and focal expression of GFAP and synaptophysin support multipotential differentiation.

This case is unusual in the supratentorial location of the tumor as well as its early detection in utero. The exact site of origin is still unclear due to the extensive destruction of the CNS tissue.

References

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Case Abstract

A 29-week gestational age newborn male infant was found to have an echogenic mass in the 3rd ventricle by prenatal ultrasound 2 weeks prior to delivery. At delivery he was poorly responsive and had hydrocephalus and ascites. A CT scan after birth showed cerebral infarction, amorphous tissue in the left hemisphere and numerous calcifications. Despite supportive treatment he died 4 days after birth. Postmortem examination of the brain revealed marked distortion of the architecture and a supratentorial undifferentiated neoplasm consistent with a PNET. The tumor showed extensive areas of hemorrhage and necrosis and involvement of lateral and third ventricles, brain parenchyma, and meninges.

For a more complete discussion of this case, additional micrographs and information regarding submission of cases, please access the WWW at: <http://brainpathology.upmc.edu>. We welcome comments about these or similar cases our readers may have encountered. Email: bpath@np.awing.upmc.edu