Pathological Case of the Month

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This male infant was born at 41 weeks' gestation to a 20-year-old, gravida 3, para 2 woman with an unremarkable medical history, normal prenatal test results, and no smoking, drug use, or other harmful habits. Pregnancy was complicated by decreased fetal movements during the last week of gestation. An ultrasound performed the morning before delivery revealed severe hydrocephalus. A previous ultrasound performed at the 20th week showed no abnormalities. The infant was delivered by emergency cesarean section without complications or need for resuscitation. Apgar scores were 7 and 9 at 1 and 5 minutes, respectively. His birth weight was 4054 g (>95th percentile for age), length was 51 cm (75th percentile), and head circumference was 40 cm (>95th percentile). He was transferred to the neonatal intensive care unit for further evaluation and management.

On physical examination the infant had normal vital signs. He had marked macrocephaly, a full anterior fontanel measuring 5 × 4 cm and a full posterior fontanel measuring 5 × 3 cm. Results of the remainder of the examination were normal. Complete blood cell count, blood chemistry analysis, and results of coagulation studies were normal. Serum and cerebrospinal α-fetoprotein levels were markedly elevated at 7555 µg/L and 1028 µg/L, respectively. Serum quantitative human chorionic gonadotropin levels were in the normal range. Magnetic resonance imaging of the head (Figure 1) showed a multicystic mass located within the third ventricle.

The patient developed seizures on the third day of life and underwent radical resection of the intracranial mass on the following day. A 4-cm, grayish, heterogeneous tumor growing into the cavum septum pellucidum and third ventricle was resected. The surgical specimen consisted of 3 pieces of soft, tan and maroon, gelatinous tissue. Microscopic sections of the tumor are shown in Figure 2 and Figure 3.
Intracranial Immature Teratoma

In general, intracranial teratomas are extremely uncommon. Most are located in the pineal region, but about 20% are located in the suprasellar or infrasellar regions. Histologically they are classified as mature, immature, or teratoma with malignant components based on the amount of tissue differentiation. They contain structures derived from all 3 germ cell layers. Immature teratomas may also contain primitive neural tissue. They may produce plasma or cerebrospinal fluid markers such as α-fetoprotein and human chorionic gonadotrophic hormone. Prenatal diagnosis has increased with the use of ultrasonography.

Congenital intracranial teratomas have a poor prognosis. There are several cases reported in the literature of stillborn infants presenting with severe macrocephaly, history of polyhydramnios, and even skull rupture at delivery. Most of these infants die within the first few months of life and the longest survival reported has been 1.5 years.

Treatment includes radical surgical resection of the tumor, followed by either radiotherapy and/or chemotherapy. The extent of surgical resection is an important prognostic factor. If the patient survives, radiotherapy is usually postponed until 24 to 36 months of age to avoid complications such as stunted growth, endocrine disturbances, and neuropsychologic problems.

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REFERENCES