Prenatal Diagnosis of a Fetal Intracranial Tumor

Susie N. Chung, MD, Richard L. Rosemond, MD, David Graham, MD

Neonatal intracranial tumors are rare and usually indicate a poor prognosis. The first case report of the identification of a fetal intracranial tumor by transabdominal ultrasonography was in 1980. Transvaginal ultrasonography and MR imaging have recently been employed to improve the imaging of fetal intracranial structures and abnormalities. Neonatal brain biopsy is still necessary, however, to assign a definitive diagnosis, since images of two histologically different tumors can appear the same.

Teratomas are the most frequent intracranial tumors found in the neonate, with meningeal sarcoma, craniopharyngioma, lipoma of the corpus callosum, and oligodendroglioma being found less commonly. We report a patient diagnosed prenatally as having an enlarging intracranial mass that proved to be a gangliocytoma.

CASE REPORT

A 27 year old gravida 2 para 1 woman at 24.7 weeks' gestational age was seen because of a fetal intracranial mass. Her prenatal course had been uncomplicated. The findings from two prior ultrasonograms performed at 7 and 12 weeks' gestation were normal. A 2.4 × 2.3 cm complex intracranial mass was visualized by ultrasonography. By 32 weeks' gestation, the fetus was in vertex presentation, and transvaginal ultrasonography using color Doppler energy revealed a suprasellar mass measuring 4.4 × 3.5 cm. The mass contained both cystic and solid components and caused marked displacement of the circle of Willis (Fig. 1). The lateral ventricles were mildly enlarged (10.3 mm), and the head circumference was typical of a fetus at 36 weeks' gestational age. No other anomalies were found. At 36 weeks' gestation the mass had enlarged to 6.3 × 6.4 cm, and an amniocentesis indicated fetal lung maturation. A repeat cesarean section was performed, and a 3100 g female infant with normal Apgar scores was delivered. The head circumference was noted to be 36 cm (> 90th percentile). Neurologic examination revealed an active infant, with strong sucking, fair grasp,
and weak Moro reflexes and slightly hypotonic lower extremities; otherwise, the infant appeared normal. A neonatal head MR imaging and CT scan (day 2) confirmed a $4 \times 5 \times 6$ cm suprasellar mass, with cystic components present bilaterally (Fig. 2). The brain stem was displaced posteriorly, and the lateral and third ventricles were mildly enlarged. The large complex mass involved the anterior one third of the third ventricle. No definite calcification or adipose tissue was noted in the mass. A needle biopsy of the mass and bilateral cyst aspiration was performed (day 3), and pathologic examination revealed a gangliocytoma.

Because of the slow growing nature of the tumor, the neurosurgeons elected to delay surgery. The infant did require a ventricular shunt at 4 months of age and did well until 22 months of age, when she was treated with chemotherapy in an effort to shrink the mass prior to surgery. Chemotherapy was unsuccessful, and at the time of surgery, only approximately half the mass was able to be resected. She is now 3 years of age, is mildly developmentally delayed in her speech and motor skills, and is currently being treated for hypothyroidism; otherwise, she is doing well.

**DISCUSSION**

Fetal intracranial tumors are very rare, but if detected, they are most commonly teratomas. Teratomas appear as masses with cystic and solid components on ultrasonography. Other possibilities include meningeal sarcoma, craniopharyngioma, lipoma of the corpus callosum, and oligodendroglioma. These tumors cannot be distinguished from each other until biopsy.

Gangliocytomas in the sellar region of the fetus have never been reported in the literature, as far as we know. These tumors are made up of mature neuronal elements and have proliferative potential. The growth of gangliocytomas is slow and nonaggressive. It is possible to have a good prognosis without recurrence if complete tumor resection is accomplished. The prevalence of gangliocytomas ranges between 0.4 and 3.8% of all intracranial tumors. These tumors tend to occur in children and in young adults without sex predilection. Gangliocytomas are more commonly found in the spinal cord (49%), the cerebral hemispheres (33%), and the brain stem (17%). It is rare to find these tumors in the sellar region, cerebellum, or pineal gland. Only 54 cases of sellar gangliocytomas have been published; the youngest symptomatic patient was 5 years old. In 65% of these patients, an additional pituitary adenoma, mostly growth hormone secreting, was also present.

Operative therapy, radiotherapy, and medical therapy are the three modes of treatment available. If operative therapy is chosen, it is mandatory to search for a pituitary adenoma. Because of the slow growing nature of gangliocytomas, it is not recommended to perform radical surgery and risk postoperative morbidity. Instead, multistaged operative therapy can be performed. Radiotherapy is effective against the hormonal overproduction syndrome; however, it is not effective in the prevention of recurrent growth of the gangliocytoma. The effect of medical therapy on the hormonal overproduction syndrome associated with gangliocytomas is not yet clear.
Because of their slow growth pattern without a tendency for developing into malignancy, the prognosis for sellar gangliocytomas is good. Unfortunately, 79% of patients with sellar gangliocytomas have suprasellar or parasellar extension, or both, making tumor removal very difficult. In addition, the prognosis is dependent on the ability to treat the hormonal hypersecretory syndrome.

In summary, this case report describes a case of a fetal sellar gangliocytoma. The growth, location, and extent of the mass were diagnosed with the aid of transvaginal ultrasonography with color Doppler energy, which allowed appropriate consultation to be obtained for delivery and immediate neonatal care planning.

REFERENCES