Giant intracranial teratoma and lack of cortical development in a fetus

Case report

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Microneursurg (Pediatrics 2) 103:180–183, 2005

Antenatal diagnosis of an intracranial neoplasm is extremely rare. The authors describe a case in which a 21-week-old fetus was found, by using fetal ultrasonography, to have a large intracranial mass. Fetal magnetic resonance (MR) images, obtained at 21 and 25 weeks’ gestation, supported the diagnosis of a teratoma. As the tumor increased in size, near-complete brain atrophy ensued. Premature labor was induced, and a nonviable fetus died within minutes of delivery. Postmortem analysis confirmed a teratoma occupying a major portion of the intracranial space. In cases in which abnormal brain development is suspected in a fetus, the use of fetal MR imaging can give a clearer picture of the pathological entity, which may allow for a more accurate diagnosis. The usefulness of fetal MR imaging in monitoring brain development and tumor growth during treatment planning is discussed.

Key Words • brain tumor • fetus • magnetic resonance imaging • teratoma • pediatric neurosurgery

A ntentatal diagnosis of a brain tumor in a fetus occurs rarely. We present an unusual case of a giant intracranial teratoma diagnosed antenatally by using fetal ultrasonography in the initial stages. The presence of progressive brain atrophy and marked tumor growth was documented in serial ultrafast MR images, which were invaluable to the multispeciality team and family in treatment planning.

Case Report

This 22-year-old primagravida presented at 21 weeks’ gestational age. Fetal ultrasonography revealed a large mass within the interhemispheric region. The mass consisted of solid components with some cystic components and appeared to extend into the oropharynx (Fig. 1). The ventricles were dilated, and visualization of normal brain matter was difficult. After a multispeciality conference, the team agreed that more information was needed before definitive treatment planning could occur. Fast MR images of the fetus were obtained using a 1.5-tesla Siemens Symphony unit (Erlangen, Germany). A T2-weighted half-acquisition single-shot turbo–spin echo sequence was performed, with a mean sequence acquisition time of 15 seconds. No fetal sedation was used. The sequences were repeated if fetal movement occurred. Sagittal and axial images of the fetal head (Fig. 2 upper and center) were obtained, and an intracranial teratoma was suspected. Because the fetus still had extensive normal brain matter surrounding the tumor (Fig. 2 lower), the team elected to repeat the MR imaging sequence 4 weeks later. These images demonstrated unequivocal growth of the tumor, worsened hydrocephalus, and a substantial loss of brain matter (Fig. 3). Following these findings, labor was induced, and a nonviable fetus was delivered, living only a few minutes. Complete obstruction of the oropharynx by a tumor was present. Postmortem analysis confirmed a teratoma that occupied approximately 50% of the intracranial space.

Discussion

Prior to new advances in ultrafast MR imaging, diagnosis of fetal abnormalities was bound by the limitations of ultrasonography. In the rare cases of a brain tumor being diagnosed antenatally by using ultrasonography, clear definition of the tumor was limited by maternal fat, position of the fetus within the pelvis, and reverberative artifacts of the bony calvarium, especially after 32 weeks of pregna-
Magnetic resonance imaging is not limited by these factors. With the increasing popularity of fetal surgery, a concurrent increase in the use of fetal MR imaging has occurred. Previous limitations of fetal MR imaging have been overcome by the development of ultrafast sequence imaging, which obviates the need for sedation of the fetus by way of an invasive procedure requiring a pancuronium injection into the umbilical cord.\(^\text{1,15}\) This procedure involves umbilical puncture, which carries the risk of hematoma and infection. The ultrafast MR imaging techniques allow clear, precise images of normal and abnormal fetal central nervous systems without the risks involved in fetal or maternal sedation.\(^\text{2,4–6,9}\) In this case, the multispecialty team used these imaging advances to correctly diagnose the fetal intracranial mass as a teratoma.

Intracranial teratomas are extremely rare lesions that usually occur in infancy or childhood, representing nearly 0.5% of all intracranial tumors.\(^\text{3}\) Histologically, they are one of the most common neonatal brain tumors.\(^\text{1,13}\) Teratomas are composed of tissue representing the three germ layers and can greatly vary in their level of maturity.\(^\text{11}\) As in the case presented, teratomas most often affect the pineal region\(^\text{3,8}\) and are located in the midline. They have varying degrees of calcification and cyst formation within the

![Fig. 1. Transuterine fetal ultrasonography images obtained at 21 weeks’ gestational age. Upper: Sagittal image revealing a solid intracranial mass with questionable extension into the oropharynx. Lower: Axial image demonstrating a cystic structure within the intracranial space and poor differentiation between brain tissue and abnormal tissue.](image1)

![Fig. 2. Half-acquisition single-shot turbo–spin echo T2-weighted MR imaging sequences at 21 weeks’ gestational age. Upper: Sagittal image demonstrating a normal fetal brain matter above the large midline intracranial mass, which extends into the oropharynx. Center: Axial image demonstrating a normal cortical mantle on the left side with hydrocephalus and a solid mass with a cystic component on right hemisphere. Lower: Far-lateral sagittal image demonstrating normal fetal brain with an abundance of cerebral spinal fluid in the ventricles and anterior to the cerebrum.](image2)
tumor mass and may show the presence of fat or cartilage formations.\textsuperscript{11}

Some neonatal teratomas are so massive that the normal posterior fossa anatomy is obscured, or the brainstem may be replaced completely by the teratoma.\textsuperscript{7} Resection is the treatment of choice for teratomas, with successful resection providing the best prognosis possible in these difficult cases.\textsuperscript{14} Because teratomas are often large and located in the midline, however, resection is often hazardous.\textsuperscript{11} Another concern in dealing with neonatal teratomas is the vascular nature and the limited blood supply of an infant.\textsuperscript{14} Whereas benign teratomas actually have an excellent prognosis following resection, malignant ones have a poor prognosis\textsuperscript{14} and carry a particularly high mortality rate.\textsuperscript{2,12} Infants with malignant teratomas have only a 7\% 1-year survival rate.\textsuperscript{13}

In the case presented, resection was not feasible because of the large size of the teratoma and its effects on the normal cortical matter.

In the case of this unfortunate infant, the second set of fetal MR images, obtained 4 weeks after the first, revealed a noticeable increase in the size of the intracranial tumor, concurrent with less cerebral tissue. We speculate that tumor growth occurred at the expense of normal brain development. This sequence could be secondary to compressive effects on the brain, with occlusion of developing brain vessels by the tumor. It could also be secondary to a nutrient redirection phenomenon, arising from angiogenesis within the tumor. Although significant hydrocephalus was apparent, it did not compress the tumor, possibly because tissue within the tumor was firmer than that of the surrounding cerebral tissue. We suspect that redirection of blood flow caused the normal cerebral tissues to have insufficient oxygen and nutrients necessary for growth and development. Slagle, et al.,\textsuperscript{10} showed that infants with insults such as parenchymal hemorrhage, cystic periventricular leukomalacia, or intraventricular hemorrhage had local delays in convolutional maturation. They also hypothesized that alterations in local nutrients and associated decreased cellular proliferation can result in delayed sulcal development.

Previous studies have shown that fetuses with cerebral anomalies have wide variations in cortical development. Levine, et al.,\textsuperscript{5} demonstrated that fetuses with abnormal brains exhibit a 2-week lag in cortical development in comparison with the cortical development of normal fetuses. This case of neoplasm presented a more dramatic abnormality and thus may explain why the decrease in normal cortical growth was so extreme and eventually led to deterioration of previously established cortical matter.

Conclusions

In the case presented, fast MR imaging confirmed the devastating diagnosis of teratoma. In addition, it allowed the physicians to track normal cortical growth as well as the growth of the teratoma. The images provided a measure of confidence that was valuable in counseling the family and in allowing them to make an informed decision. Based on the findings of this case, we recommend the use of ultrafast MR imaging in cases in which fetal abnormalities are suspected. Early diagnosis is especially important because medical advances offer a variety of treatment options.

References

2. Levine D, Barnes PD: Cortical maturation in normal and abnormal brains exhibit a 2-week lag in cortical development in comparison with the cortical development of normal fetuses. Radiology 210: 751–758, 1999
Giant intracranial teratoma in a fetus


Manuscript received August 12, 2004. Accepted in final form January 27, 2005.

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