CASE REPORTS

Prenatal Sonographic Evaluation of Two Intracranial Teratomas

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Prenatal sonographic evaluation is of utmost importance in the detection of congenital anomalies. At the Ultrasound Section of our Radiology service, we incidentally detected fetal intracranial neoplasms in two different patients using non-invasive imaging. Our presumptive diagnosis in both cases was a teratoma, the most common brain tumor in the perinatal period. Subsequent confirmation was provided with pathological samples obtained at autopsy in both patients, revealing immature teratomas. Fetal intracranial tumors are so rare and incredibly, these two unique cases presented in our section within just a few months of one another.

Key words:Prenatal sonography, Immature teratoma, Fetal brain.

Intracranial tumors of early childhood are rare but even rarer is their presentation in the fetus. Of all the childhood brain tumors, the most common to present at birth is the teratoma. We wish to present two cases of intracranial, immature teratomas detected prenatally using sonographic imaging.

The first case was that of a 15 year old, primigravida, who arrived at our institution at 25 weeks gestational age for her first sonographic evaluation. The biparietal diameter (BPD) was 7 cm and the head circumference (HC) was 26.9 cm, both of which correlated with 29 weeks of gestational age (GA), while the remaining fetal parameters (femoral length and abdominal circumference) corresponded to 25 weeks of GA. The normal brain parenchyma was completely distorted revealing an intracranial mass, which is depicted in Figures 1-3.

The working diagnosis of this brain neoplasm was of a teratoma, as it accounts for more than half the intracerebral neoplasms in this age group (1). The fetus was still viable after sonographic evaluation and the mother continued with the pregnancy despite a dismal prognosis rate for this mass. A baby girl was delivered by cesarean section at 32 2/7 weeks due to an enlarged fetal head. The neonate had Apgar scores of 8 at one minute and 9 at five minutes. Six days after birth, the neonate's condition began to

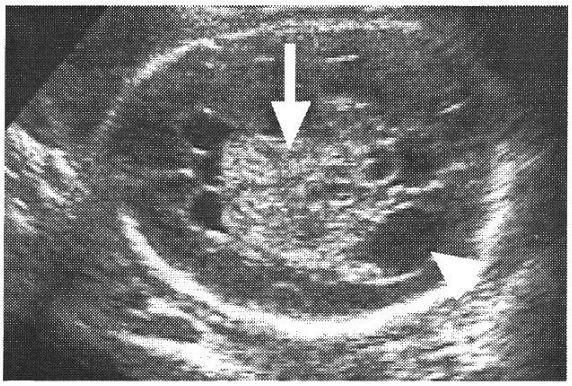

Figure 1. Coronal view of fetal head shows a large, complex, predominantly echogenic intracranial mass (arrow) extending from midline towards one of the hemispheres and associated with ventriculomegaly (dilated ventricles) arrowhead.

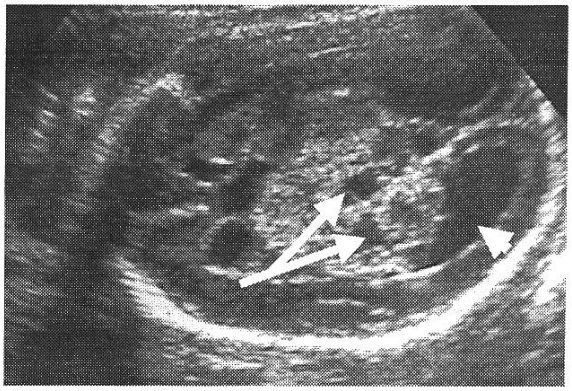

Figure 2. Another view of the intracranial mass showing sonolucent spaces, (arrows) in favor of regions of hemorrhage or necrosis. Also, the visualized ventricles are dilated (arrowhead).

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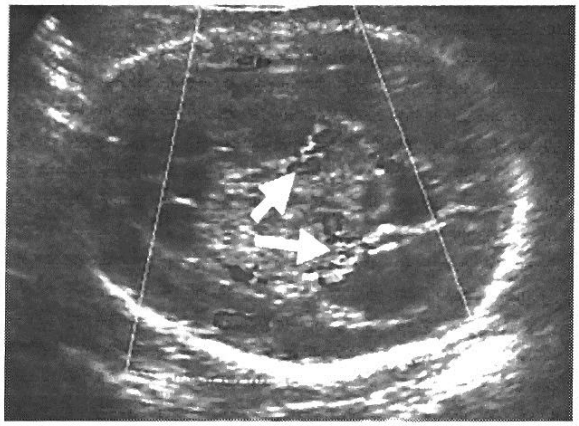

Figure 3. Doppler imaging demonstrating vascularity within the intracranial mass (arrows).

deteriorate. She developed hyperbilirubinemia, which was treated with phototherapy. Later the same day, the patient's oxygen level and heart rate fell and, despite cardiopulmonary resuscitation, she died of respiratory failure.

Autopsy confirmed the diagnosis of intracranial teratoma; figures 4-6 show the gross pathology of the lesion: the brain was markedly softened but the gyral pattern was adequate for the patient's gestational age. The cerebral mass weighed 540 g after fixation (expected weight was 198g +/- 48g). The cerebral hemispheres were asymmetrical, the left side being larger and having a firmer consistency than the right. An indurated mass was palpable on the left side, measuring 10 cm x 10 cm x 6 cm, involving the entire ipsilateral cerebral hemisphere. The mass extended to the right hemisphere with effacement of

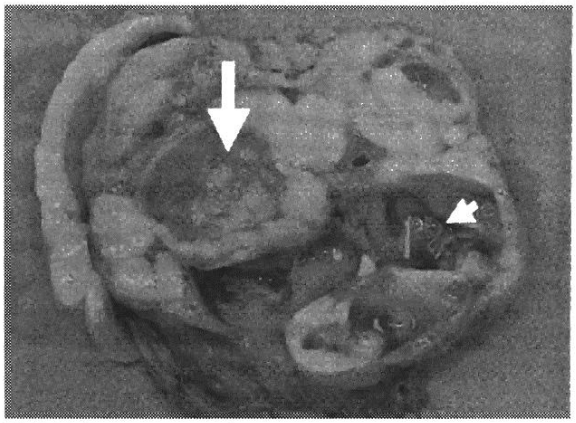

Figure 4. Coronal section of the intracranial mass showing almost complete replacement of the brain parenchyma by a variegated tumor with solid encephaloid areas, (arrow) cystic spaces and hemorrhage (arrowhead).

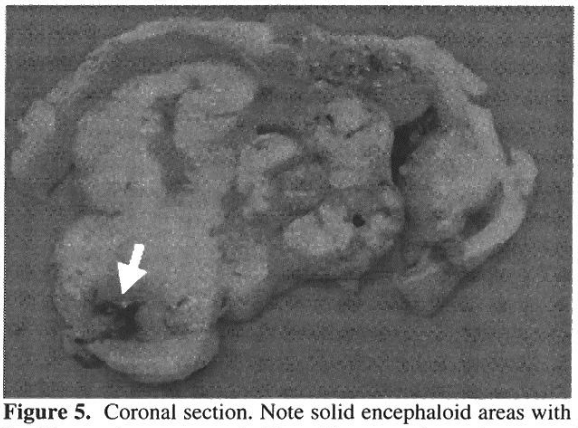

Figure 5. Coronal section. Note solid encephaloid areas with focal hemorrhage surrounded by a thin cortex (arrow).

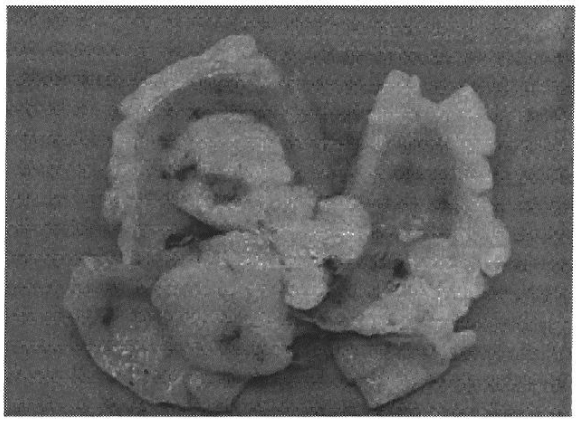

Figure 6. Coronal section, frontal area. There is thinning of the cortex and ventricular dilatation.

major cerebral structures such as the basal ganglia, thalamus, midbrain and hippocampi. Histological evaluation revealed extensive encephaloid and cystic areas containing serous fluid and hemorrhage as well. The ventricular system appeared markedly dilated. The cerebellum was grossly unremarkable. An immature teratoma was microscopically demonstrated with extensive areas of immature neuroepithelium and foci of cartilage. Also seen were pancreatic and liver tissue, muscle, respiratory epithelium, mucous secreting glands and dilated cystic glands.

The second case, which was detected just a few months later, was that of a 23 year old primigravida who came to our hospital for her first prenatal evaluation at 24 weeks gestation. The BPD was 6.5 cm and HC was 24 cm, both of which correlated with 26 weeks of GA, while the femoral length and abdominal circumference parameters corresponded to a GA of 22 weeks. The sonographic images also revealed an intracranial mass as shown in Figures 7-8.

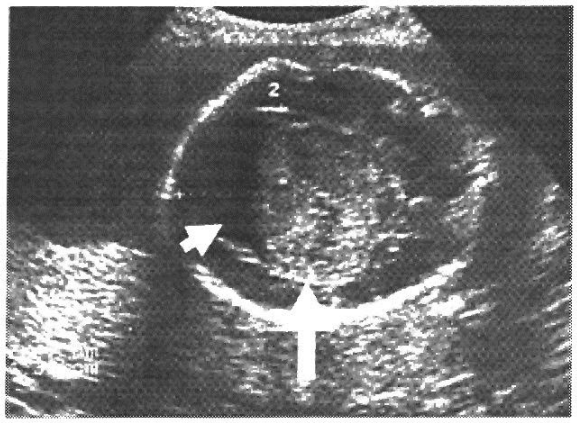

Figure 7. Sagittal view of fetal head shows a large echogenic intracranial mass, (arrow) with associated peripheral cystic components (arrowhead) causing distortion of the expected normal anatomy.

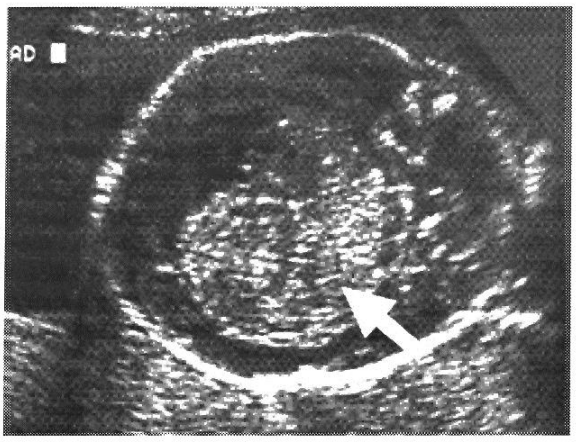

Figure 8. Another view of a predominantly echogenic, solid intracranial mass arising from midline and extending towards lateral hemisphere (arrow).

In this case, the mother requested a therapeutic abortion, and it was performed. Autopsy revealed an immature intracranial teratoma consisting mainly of immature neuroepithelium. Unfortunately, correlation between the sonographic and gross pathological findings could not be achieved due to friability of the tissues.

Discussion

It is known that congenital intracranial tumors are rare and only account for 0.5-1.5% of all childhood brain tumors (2). The sonographic presentation of this lesion is cranial enlargement as a result of an heterogeneous intracerebral mass having both solid and cystic components and occasional areas of calcifications (1). Normal brain architecture is distorted. The teratoma has increased vascularity with low resistance flow, the ventricular system is markedly dilated (3).

Early diagnosis is of utmost importance. It is well known that the prognosis of a congenital intracerebral teratoma is generally very poor. With the aid of sonographic evaluation in the early prenatal period, diagnosis of an extremely rare entity can be established and managed accordingly.

Resumen

La sonografia prenatal es muy valiosa en la detección de anomalías congenitas. En la sección de Sonografía, dentro del servicio de Radiología Diagnóstica, incidentalmente detectamos dos casos de tumor fetal intracraneal haciendo uso de esta modalidad no invasiva.

Nuestra impresión diagnóstica fue de teratoma intracraneal en ambos casos, ya que es el tumor cerebral mas común en la edad perinatal. Esto fue confirmado patológicamente, luego de la autopsia de ambos fetos, siendo el diagnóstico histológico el de teratomas inmaduros.

Los tumores fetales intracraneales son muy raros, sin embargo, estos dos casos excepcionales fueron vistos en nuestra sección de sonografia, en un corto intérvalo de tiempo.

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