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Congenital Intracranial Teratoma: Management Challenges!

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Abstract

Congenital brain tumors are extremely rare. Among them teratomas are the most common. They make up one-third of all congenital brain tumors. Late diagnosis, poor prognosis, and majority being delivered by cesarean section (subjecting the mother to increased morbidity) add to the complexities and challenges of the case. We report a rare case of a massive congenital brain tumor diagnosed prenatally on ultrasound. Because of the poor prognosis and associated craniomegaly, pregnancy termination was contemplated at 28 weeks aiming at vaginal delivery, despite a high probability of an eventual cesarean delivery due to the associated macrocrania. Intrapartum second stage of labor was protracted due to nondescent of the large head; however, providentially there was timely skull rupture of the fetus facilitating a vaginal delivery. The cerebral tumor was confirmed at autopsy to be an immature teratoma. There were no other associated malformations. Ultrasound has emerged as major modality in prenatal diagnosis aiding the antenatal preparation of the prospective parents and the multidisciplinary team for the poor outcome. The knowledge gained and anticipated prognosis help immensely in planning the management, thereby avoiding subjecting the woman to cesarean section and its morbidity.

Keywords

- skull rupture
- brain
- congenital
- intracranial
- massive
- teratoma

Introduction

Congenital brain tumors are extremely rare and comprise 0.5 to 1.9%[1] of all childhood brain tumors. Among these, teratomas are the most common and constitute one-third[2,3] of all such cases.

As prognosis of intracranial teratoma is poor,[4] the prenatal diagnosis and obstetric management present a great challenge for planning and follow-up and treatment of the disease in accordance to preferences of parents.[5]

Case Report

A 29-year-old lady, G5P4L2D1, attended antenatal care in Wattayah ObGyn clinic at 23 weeks of gestation. She had previous normal deliveries. Apart from mild gestational diabetes in the present pregnancy, there were no other comorbidities.

This was a nonconsanguineous marriage. She had two live healthy children. Her last pregnancy had a fetus with renal agenesis and ended in intrauterine fetal death.

Anomaly scan at 23 weeks gestation showed that the fetus had a brain anomaly with ventriculomegaly—holoprosencephaly. There was already craniomegaly and advanced biparietal diameter corresponding to 28 weeks. This was followed by a detailed anomaly scan by a fetal medicine consultant who reported a large intracranial tumor. The scan at 27 weeks showed a large for gestational age fetus with a weight of 2 kilos. Macrocephaly was noted with a biparietal diameter measuring 12.24 cm and head circumference measuring

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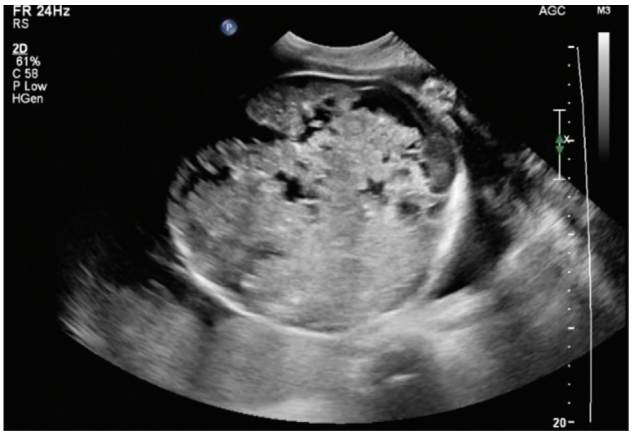

Fig. 1 Ultrasound showing massive intracranial tumor with solid and cystic areas replacing the entire brain tissue and disrupting the anatomy.

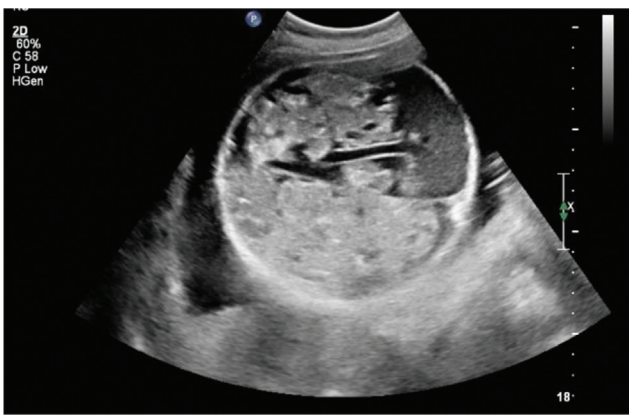

Fig. 2 Ultrasound showing the fetal head with no discernable anatomy.

40.13 cm. Abdominal circumference was also advanced and measured 27.2 cm corresponding to 31 weeks gestation. All long bones corresponded to 26 to 27 weeks gestation. Associated polyhydramnios was noted. Fetal head was grossly enlarged. The entire brain was replaced by large irregular solid and cystic areas with no normal tissue (►**Figs. 1**–**3**).

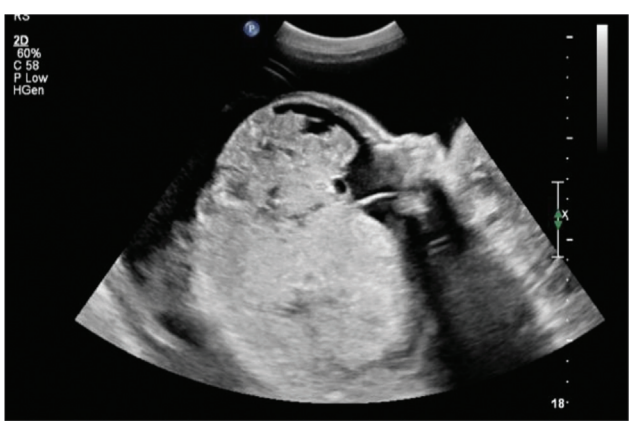

Fig. 3 Ultrasound showing profile view of the fetal head showing distorted anatomy due to the massive intracranial tumor entire brain tissue and disrupting the anatomy.

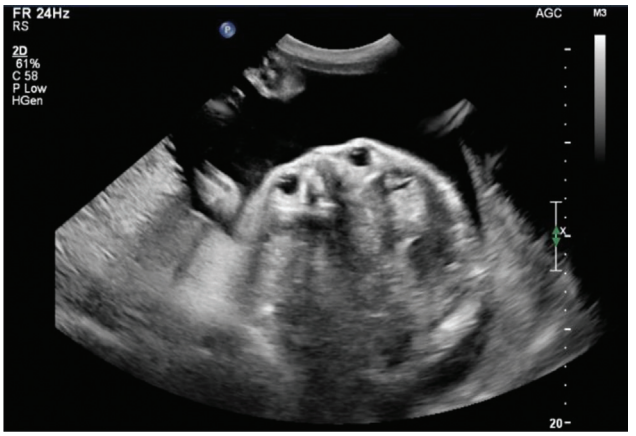

Fig. 4 Ultrasound showing hypertelorism.

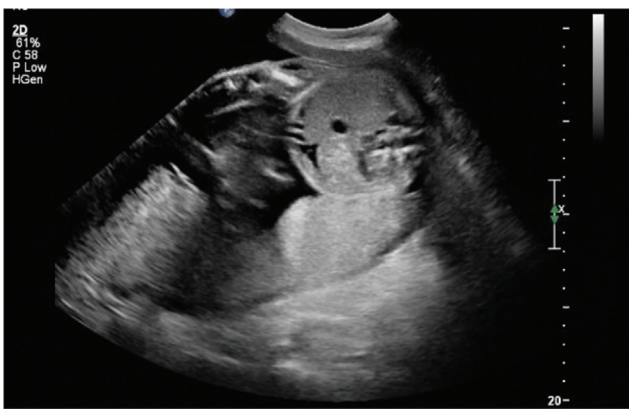

Fig. 5 Ultrasound showing mild ascites: consistent with features of nonimmune hydrops.

The fetus also showed frontal bossing and hypertelorism (►**Fig. 4**). Mild ascites (►**Fig. 5**), pericardial effusion (►**Fig. 6**), and cardiomegaly were noted. There was mild tricuspid regurgitation but outflow tracts were normal. Stomach bubble was small and collapsed. The spine, kidneys and urinary bladder were normal. Impression was of a brain tumor.

Couple was counseled about the poor prognosis. As fetus already had macrocrania, delivery was planned at 28 weeks

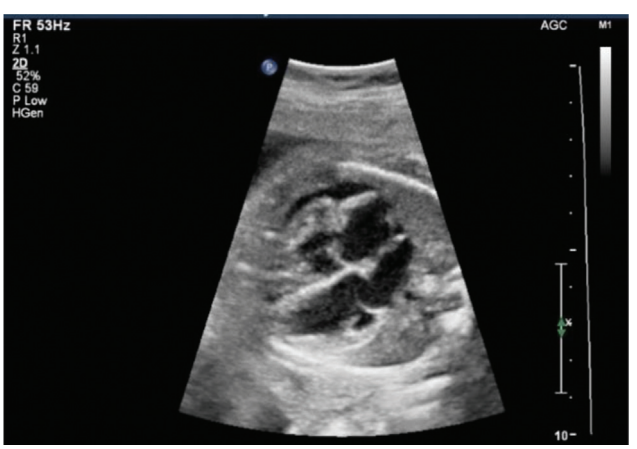

Fig. 6 Ultrasound showing mild pericardial effusion, consistent with features of nonimmune hydrops.

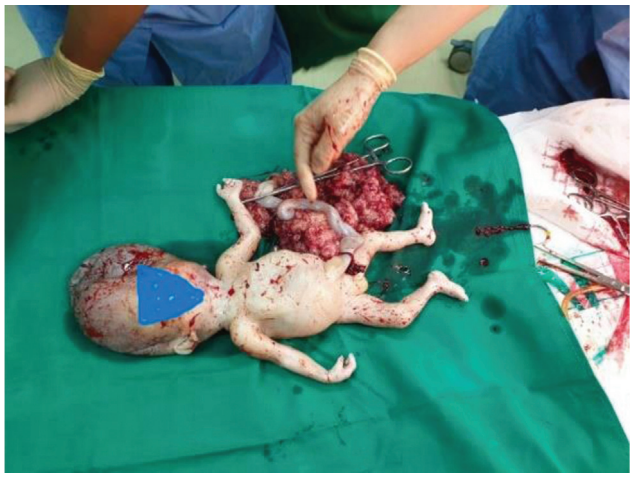

Fig. 7 Stillborn baby with decompressed cranium and massive variegated intracranial tumor by the side.

with the aim of avoiding cesarean section for a large head. The aim was to achieve vaginal delivery.

Labor was induced. The fetus was in cephalic presentation. Second stage of labor was protracted as the head remained above pelvic brim and there was no descent. Cephalocentesis did not seem a viable option as there was more of solid tissue within the fetal cranium rather than fluid that could be tapped. We were contemplating cesarean section for nondescent of the fetal head but providentially there was timely spontaneous rupture of the fetal head with the decompression of fetal skull and brain tumor tissue from the vagina, facilitating the delivery of the head followed by the body (►**Fig. 7**). At birth, the neonate was seen by the pediatrician. There were no signs of life and it was declared as a fresh stillbirth. On gross examination, the brain tissue was disordered, hyperplastic and had areas of hemorrhage, cystic, and necrotic changes. Brain tissue was sent for histopathological examination and was reported as congenital intracranial immature teratoma.

Due to the anticipated diagnosis and poor prognosis of the fetus by prenatal scan, the couple and medical team were prepared for the birth of anomalous baby with guarded viability. There was great relief, both from the obstetrician and the couple, at avoiding a cesarean scar with its associated morbidity.

Discussion

Teratomas contain tissues from all the three embryonic germ layers.[6] Though teratomas are the commonest among congenital intracranial tumors, there are only few mentioned in literature that reached massive proportions[2,7] replacing the entire cerebral hemisphere of the fetus, as was seen in our case.

These are usually diagnosed in the third trimester and carry an extremely poor prognosis[5] irrespective of the malignant potential, especially if diagnosed before 30 weeks[6] of gestation. Mortality rate is quoted as high as 90 to 97%.[6]

The differentials of a large intracranial lesion include hydrocephalus, holoprosencephaly, astrocytoma, ependymal cyst, craniopharyngioma, choroid plexus cyst, hemorrhage, arachnoid cyst, vein of Galen aneurysm, and porencephaly.[8]

Since it usually has a sporadic occurrence, karyotyping is not necessary[5,9] unless accompanied by other malformations.

Precise determination of extent of tumor involvement of adjacent structures is vital to determine the prognosis, amenability for resection and possible sequelae of surgery.[9]

Ultrasound has emerged as a major valuable tool[2,3] in assessing intracranial structures and differentiates between the tumor and other abnormalities. Ultrasound and magnetic resonance imaging (MRI) appearance of intracranial teratoma is usually of an irregular heterogeneous mass with hyperechoic and hypoechoic features with both solid and cystic areas as well as calcified components with distorted brain anatomy.[2,9–11] MRI can help in determining the remaining brain structures, exact location of tumor, and differentiating from hemorrhages.[12] Both ultrasound and MRI are complementary[2] and together allow more precise diagnosis of these tumors prenatally.

Fetal MRI is not freely available at many centers. However, due to the reliability and accuracy of information provided by ultrasound alone, we may resort to it as our guiding modality. In our center, we were able to diagnose, predict the poor prognosis, and manage this case solely on the basis of ultrasound expertise.

Maternal presentation is with rapidly increasing fundal height and polyhydramnios in 50%[2,3] cases. Cephalopelvic disproportion can occur very early, even in the second trimester.[2,13] Babies present with an intracranial mass, macrocrania, hydrocephalus, and signs of nonimmune hydrops that can be attributed to cardiac failure secondary to vascular shunt in tumor[2]. Poor prognosis of intracranial tumor at overall survival rate of 7.8 to 10%[10,13] has been quoted in literature.

In our case, ultrasound reliably demonstrated gross macrocephaly with biparietal diameter measuring as big as 12.24 cm and head circumference 40.13 cm (at 27 weeks of gestation), loss of recognizable anatomic features due to replacement of entire brain by large irregular solid, and cystic areas with no normal tissue, features of fetal hydrops and polyhydramnios. Color doppler ruled out vascular tumor (►**Fig. 8**). All the above features helped in predicting poor fetal prognosis.

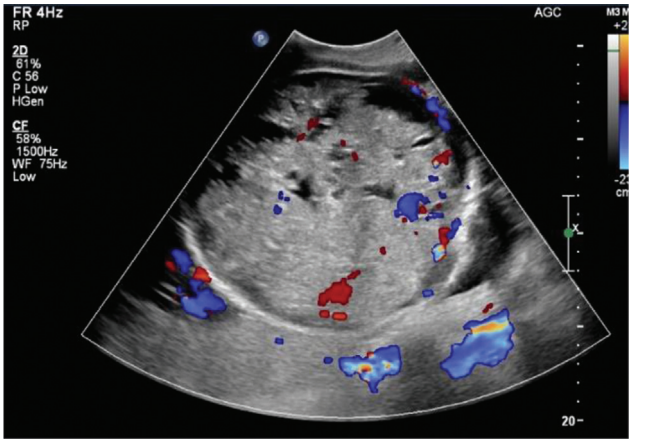

Fig. 8 Ultrasound image with unamplified color Doppler signals over the intracranial mass ruling out vascular tumor.

Definitive diagnosis was made by histology after birth.[1]

Causes of death due to intracranial teratomas are brain effacement by the tumor, dystocia, and cranial rupture during delivery.[2,14]

During the fetal period, the skull has the remarkable ability to expand, so some congenital central nervous system tumors can expand enormously leading to dystocia, hydrops, and stillbirth. Large tumors may necessitate decompression of the cranium to permit vaginal delivery.[14–16]

Cesarean section rate is alarmingly high almost 60%[13] in such cases.

Classic teratomas are associated with high death rates and should be conservatively managed. If possible, medical termination of pregnancy (MTP) can be offered before viability. Vaginal delivery should be the aim,[16] far as possible.

There are a few case reports in literature, similar to our report, wherein there have been intracranial tumors with an exponential growth[7,17] with extremely poor prognosis.

Isaac[17] has reported a rare presentation of an extremely uncommon tumor associated with massive growth, high output failure, and skull rupture. In a case report by Washburne et al,[7] cesarean section was performed at 30 weeks of gestation for an 18 years old due to severe hydrocephalus. Spontaneous rupture of the skull at cesarean section led to delivery of a stillborn infant, and a congenital teratoma was found.[7]

Isaac[17] reported that of 49 cases of fetal and newborn intracranial tumors, 16 of these fetuses died in utero and seven were stillborn. Case survival ranged from 9.5 hours to 2 years. Only three remained alive. Location and resectability of the tumor determine long-term survival.[17]

In another review by Robles Fradejas et al,[5] a total of 49 cases of congenital intracranial teratomas were reviewed. Among them, electively 12 cases underwent early medical termination with misoprostol. Twenty-eight cases were delivered by cesarean section and only nine delivered vaginally. All died in the neonatal period except three cases, where there was intrauterine demise after diagnosis.[5]

There have been reports in literature, whereby in majority of the cases, cesarean section had been performed because of the large fetal head circumference.[7] Cephalocentesis does not outwardly appear to be a possible option due to the solid nature of the intracranial tumor. But multiple reports have confirmed intrapartum skull rupture indicating the fragile nature of skull that may be amenable to cephalocentesis[16] and helpful in skull decompression facilitating vaginal delivery. This feature of fetal skull rupture at delivery makes consideration for vaginal delivery a viable option even in cases of massive brain tumors.

Due to the solid nature of the tumor that was indicated by ultrasound in our case, we did not resort to cephalocentesis as it seemed less likely to help in shrinking the macrocephalic head. Cesarean section was being contemplated for nondescent of the fetal head but providentially skull ruptured with immediate decompression, thereby making vaginal delivery possible even with the huge fetal head! This incident helped in strengthening our confidence to consider cephalocentesis or other methods to decompress the fragile skull in fetuses with intracranial tumors, to maximize efforts at vaginal delivery. Given the escalating rate of cesarean section worldwide, similar decisions will help in curtailing the cesarean section rate and decreasing maternal morbidity, especially where the fetus is known to have a poor prognosis.

Due to the poor prognosis, it becomes a great obstetric challenge to manage, plan, follow-up, and treat the disease according to parental preferences.[5] Advancement in technology has greatly improved early diagnosis of congenital intracranial tumors, but there has not been much headway in fetal surgery, perhaps because prognosis for brain tumors remains poor. It is vital to have utmost precise information regarding the tumor, nature, and extension of lesion. In addition, even when parents do not opt for termination, the precise diagnosis provided by antenatal imaging may help healthcare professional prepare the parents for the neonatal outcome. Fetal medicine centers should be well equipped with multidisciplinary teams acting, in unison, to provide better perinatal management for the prospective parents and care for fetuses with congenital intracranial tumors.[6,9]

Conclusion

Ultrasound is majorly a useful modality for early and accurate opinion of congenital intracranial tumors. Given the poor prognosis for congenital intracranial teratomas, the information provided is essential in timing delivery, reducing maternal morbidity by promoting vaginal delivery and deferring cesarean section. It also enables multidisciplinary management in counseling and emotional preparedness of the parents.

Authors' Contributions

SS and MV conceptualized the study. MV helped in methodology and data collection.

SS was involved in writing, editing, review, and supervision of the study.

All authors have read and agreed to the published version of the manuscript.

Statement Conforming to the Declaration of Helsinki

The case report is in accordance with the Declaration of Helsinki.

Patient Permission

Identity of the patient is not revealed in the case report and part of the image is obscured so as to conceal the identity.

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None.

Conflict of Interest

The authors declare no conflict of interest.

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