A rare case of large fetal intracranial tumor: Teratoma

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ABSTRACT

Introduction: Fetal intracranial tumors represent only 10% of all antenatal tumors; teratoma being the most common accounting for approximately half of all reported cases followed by astrocytoma, lipoma, choroid plexus papilloma, craniopharyngioma, and primitive neuroectodermal tumor. Usually these tumors clinically manifest in the third trimester of pregnancy, which otherwise nowadays often recognized earlier in pregnancy by ultrasonography and magnetic resonance imaging. Majority of fetal intracranial tumors are supratentorial, however in larger lesions imaging or even autopsy is frequently unable to determine the origin.

Case Report: Here we are presenting a rare case of large fetal intracranial teratoma with which the patient presented in the third trimester with fatal outcome.

Conclusion: Progress in technology has contributed to early diagnosis of congenital CNS tumors, but the same is not observed with fetal surgery, perhaps because the prognosis of fetal brain tumors remains poor. It is critical to have the most precise information regarding the tumoral nature and the extension of the lesion. Fetal medicine centers should be composed of a multidisciplinary team acting together to provide better assistance for fetuses with congenital CNS tumors and to develop new methods of treatment.

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CASE REPORT

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Keywords: Antenatal intracranial tumors, Immature teratoma

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INTRODUCTION

Diagnosis of any fetal abnormality is traumatic event for the family, the presence of a fetal intracranial tumor, along with mental trauma, carries with it additional diagnostic and therapeutic challenges. The prognosis is usually poor with few exceptions. Fetal intracranial

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tumors represent only 10% of all antenatal tumors, ranking behind extra cranial teratomas, neuroblastomas, and soft-tissue tumors. Most of the congenital brain tumors arise from the pineal gland, suprasellar area, or cerebral hemispheres. Intracranial teratoma is the most common fetal brain tumor, accounting for approximately half of all reported cases followed in frequency by astrocytomas of varying grades, lipomas, choroid plexus papillomas, craniopharyngiomas, and primitive neuroectodermal tumor [1, 2]. Teratoma is a neoplasm containing tissues foreign to the site of origin and containing more than one embryonic germ cell layer. These tumors most commonly become clinically manifest in the third trimester of pregnancy. Diagnosis earlier in pregnancy, particularly before 30 weeks gestation, portends a particularly deleterious prognosis, with reported mortality rates as high as 96.9% [3].

CASE REPORT

A 22-year-old female G1PoAoLo with 34 weeks gestational age (by LMP), presented to radiological department for routine antenatal ultrasound and ruling out any gross congenital malformation. As she belonged to rural background, she had not undergone any previous ultrasonography during the course of this pregnancy. On ultrasonography (Figure 1A–B), there was single live intrauterine fetus of 32 weeks gestational age with a large intracranial well defined lobulated heteroechoic lesion measuring 15.6(AP)x12.6(CC)x9.0(T) cm³ epicentered in midline supratentorial region with right sided extension causing significant dilatation of B/L lateral ventricles (R>L) with surrounding parenchymal thinning. The lesion was large enough for the origin to be exactly defined, however, was limited to supratentorial compartment only with infratentorial compartment and its contents absolutely normal. There were multiple variable sized internal cystic changes with few small calcifications noted within the lesion. No other fetal abnormality was seen. On MRI scan (Figure 2) the lesion was large well defined lobulated of approximately same dimensions as on USG, iso-hypointense on T1/T2 sequences with multiple T1 hypo and T2 hyperintense internal variable sized cystic changes with the status of obstructive hydrocephalus same as seen on ultrasonography. Tiny intralesional calcifications as seen on ultrasonography are not well appreciated on MRI scan. On the basis of above mentioned morphology of the lesion, antenatal diagnosis of fetal intracranial tumor possibly teratoma was given. With informed prognosis and proper counseling, termination of pregnancy was decided by parents. Cephalocentesis was performed (as the size of head was very large) followed by vaginal delivery. Parents refused for autopsy of delivered fetus and only the lesion extracted tissue was sent to pathology department for histopathological examination which confirmed the diagnosis of immature teratoma (Figure 3).

DISCUSSION

Fetal intracranial tumors though rare entity, nowadays are often recognized during antenatal period by ultrasound and magnetic resonance imaging, create significant medical and ethical dilemmas. An understanding of the different tumor types and their biologic behavior is necessary for appropriate counseling and care of these patients. Accurate diagnosis has important implications for fetal, maternal, and neonatal care [4]. Majority of fetal intracranial tumor are supratentorial in origin, arising mostly from pineal gland, suprasellar area, or cerebral hemispheres. Fetal intracranial tumors represent 10% of all antenatal tumors, teratoma being the most common accounts for approximately half of all reported cases [1, 2]. In literature, first report of a massive congenital intracranial teratoma was published in 1864 by Breslau and Rindfleisch [5]. Intracranial teratomas are generally large, complex, mixed cystic and solid masses with or without foci of calcification [1]. They typically arise in the midline, predominantly from the pineal gland. They may exhibit very rapid growth and reach massive proportions. Most fetuses die in utero or shortly after birth with only few exceptional long-term survivors [1–3]. They are most often diagnosed in the third trimester by ultrasonography and MRI scan. Often the masses are so large and locally invasive, replacing the intracranial contents, eroding the skull and extending into the mouth, orbit and neck that the precise anatomic point of origin cannot be determined [6].

At prenatal ultrasound, the diagnosis of teratoma should be considered for a complex intracranial mass with calcifications associated with gross distortion or replacement of normal brain tissue by the mass. The differential diagnosis for an ultrasonographically diagnosed large intracranial mass also includes astrocytoma, ependymoma, craniopharyngioma, choroid plexus cyst, and hemorrhage [7]. Intratumoral hemorrhage may also occur and thus in the setting of any fetal intracranial hemorrhage, underlying neoplasm should be considered. On color Doppler, there may be increased vascularity with low-resistance flow; leading to high-output cardiac failure and hydrops fetalis. Vascular flow is also a helpful finding to distinguish from hemorrhage. Computed tomography findings of intracranial teratoma include a large heterogeneous mass with coarse calcifications and hydrocephalus. Although the presence of calcifications on CT scan or ultrasound is a helpful diagnostic clue they are not frequently seen on MRI scan [8, 9]. On MRI, findings are highly variable, but often demonstrate a large lobulated multicystic heterogeneous signal mass. Magnetic resonance imaging scan is also helpful to determine the anatomic extent of the tumor, and to differentiate mass from intracranial hemorrhage [10 11]. Several recent case reports have described the use of fetal MRI scan between 25 and 36 weeks gestation in diagnosing intracranial teratoma, and the typical appearance is a large, heterogeneous

Figure 1: (A) Well defined large lobulated intracranial supratentorial heteroechoic lesion (red arrow) epicentered in midline with right sided extension causing significant dilatation of B/L lateral ventricles, (B) Intra-lesional variable sized internal cystic changes (blue arrow) and small calcifications (green arrow).

Figure 2: (A) T2 Axial with sagittal, and (B) T2 coronal shows well defined large lobulated intracranial supratentorial iso-hypoechoic lesion (red arrow) epicentered in midline with right sided extension causing significant dilatation of B/L lateral ventricles with intra-lesional variable sized T2 hyperintense internal cystic changes (blue arrow). Tiny calcifications are not well appreciated on magnetic resonance imaging.

Figure 3: (A) Microphotograph showing immature neural tissue (red arrow), keratin pearl (black arrow) and nodules of cartilage (blue arrow) (H&E stain, x400), and (B) Immature neuroepithelial tubules (green arrow), (H&E stain, x100). Diagnosis: Immature teratoma.

mass with cystic components on MRI scan, with no apparent difference between mature and immature teratomas [10]. Similar features have been described at postnatal MRI scan of mature and immature intracranial teratomas. Fetal MRI scan allows enhanced global imaging of these masses. Anatomical relationships and tissue characteristics are demonstrated by MRI scan with superior detail, except for calcifications (which are better noted with ultrasound and CT scan). As the MRI features of teratomas are relatively nonspecific, the differential diagnosis of congenital supratentorial tumors should also include primitive neuroectodermal tumor, astrocytoma, ependymoma, glioma, craniopharyngioma, and choroid plexus papilloma [12, 13].

The prognosis for congenital intracranial teratoma is extremely poor whether benign or malignant, with an overall mortality rate of 90% rising to 97% if diagnosed prior to 30 weeks gestation [8, 9]. Progress in technology has contributed to early diagnosis of congenital CNS tumors, but the same is not observed with fetal surgery, perhaps because the prognosis of fetal brain tumors remains poor. It is critical to have the most precise information regarding the tumoral nature and the extension of the lesion. In addition, even when the parents do not opt to terminate pregnancy, the precise diagnosis provided by antenatal imaging may help healthcare professionals prepare them for the neonatal outcome. Fetal medicine centers should be composed of a multidisciplinary team acting together to provide better assistance for fetuses with congenital Central nervous system tumors and to develop new methods of treatment. If vaginal delivery is planned, cephalocentesis may be necessary [1]. Caesarean section may be required to prevent dystocia or, in cases of fetal airway obstruction.

CONCLUSION

To conclude, congenital brain tumors represent a diverse group of benign and malignant masses that often have a devastating effect on the fetus and the pregnancy. Each tumor is unique in its natural history, histologic characteristics, anatomic location, and prognosis; and thorough understanding of all these features is necessary to assemble the appropriate multidisciplinary team and to guide patient care. Although rare, there is increasing trend of detecting fetal intracranial tumors prenatally with the ever-increasing use of antenatal ultrasonography in routine obstetric screening. Fetal magnetic resonance imaging is a non-invasive, fast, highly informative examination and problem solving tool and has become a valuable adjunct to prenatal ultrasound in suspected fetal abnormality, especially for those with obesity or oligohydramnios. To our opinion, the use of prenatal magnetic resonance imaging in addition to ultrasound is a valuable tool in utero diagnosis and counseling for a large fetal intracranial mass.

Author Contributions

Chiranjeev Kumar Gathwal – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Devender Kaur – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Kulvinder Singh – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Jyotsana Sen – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Monika B. Gathwal – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Parveen Rana Kundu – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Gaurav Malik – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Kalpna Beniwal – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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