

Prenatal and postmortem imaging findings of fetal intracranial teratoma

Fetal intrakraniyal teratomun prenatal ve postmortem görüntüleme bulguları

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Abstract

Congenital intracranial teratoma is a well-recognized but infrequent entity. The prognosis is poor with death usually occurring shortly after birth. Teratomas are usually characterized by complete loss of the normal intracranial architecture. In this paper, we presented a case with a massive fetal intracranial teratoma that replaced the cerebral hemispheres and that was initially diagnosed by ultrasonography and further evaluated by prenatal magnetic resonance imaging.

Keywords: Intracranial teratoma, fetus, ultrasound, magnetic resonance imaging

Özet

Konjenital intrakraniyal teratom, iyi bilinen ancak nadir bir durumdur. Doğum sonrası kısa sürede meydana gelen ölüm nedeniyle prognozu kötüdür. Teratomlar, genellikle normal intrakraniyal yapının tam kaybı ile karakterizedir. Bu yazıda, ultrasonografi ve prenatal manyetik rezonans görüntüleme ile tanı konulan ve serebral hemisferlerde yer değiştirmeye neden olan masif fetal intrakraniyal teratomlu bir olgu tanımlandı.

Anahtar sözcükler: İntrakraniyal teratom, fetüs, ultrasonografi, manyetik rezonans görüntüleme

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Introduction

Ultrasonography (US) is widely used as a routine antenatal screening for fetal abnormalities. Although US reveals limited anatomical information in certain anomalies, magnetic resonance imaging (MRI) has found a more important role in prenatal imaging in recent years. Fetal MRI can play a significant diagnostic and prognostic role in the presence of complex and complicated abnormalities. Congenital intracranial teratoma is a very rare tumor. Fetal intracranial teratoma is often associated with macrocephaly, bulging anterior fontanel, hydrocephaly and polyhydramnios [1, 2]. In this presentation, we present a case of fetal intracranial teratoma diagnosed by US and further evaluated with prenatal MRI.

Case report

A 22-year-old, gravida 2, para 1, woman was referred for evaluation of the hydrocephaly that had been identified on US at 27 weeks' gestation. US (M.H.A.) showed a single male fetus (cephalic presentation, posterior placenta, normal amniotic fluid and three-vessel umbilical cord). Bilateral femur lengths and abdominal circumference measurements

were compatible with 27 weeks' of gestation. However, prenatal US at 27 weeks' gestation revealed a disproportionately enlarged head. US was performed with a Powervision 6000 ultrasound scanner (Toshiba Inc., Tokyo, Japan) and 4-MHz sector transducer. Serial examinations with US confirmed a mixed solid and cystic mass located centered on the supratentorial region and causing a severe obstructive hydrocephalus, with dimensions of approximately 8x4x5.5 cm (Figure 1). The lesion was seen as a highly echogenic mass. The midline structures and ventricles could not be discerned. The fetal brain was underdeveloped. Vascularization of the tumor was demonstrated for the basal parts using low pulse repetition frequency for low flow velocities at Doppler color flow mapping. Amniotic fluid volume was within normal limits. No additional fetal structural anomalies were visualized. There was no family history of congenital anomalies. The mother's antepartum course had been unremarkable.



Figure 1. Prenatal US at 27 weeks. An axial scan of the fetal brain shows a large heterogeneous echogenic mass with scattered calcifications and severe hydrocephalus.

The exact nature and origin of the mass was difficult to determine. Following informed consent, fetal MRI was performed at 29 weeks' gestation. We performed MRI examinations with a 1.5T scanner (Excelart; Toshiba Inc., Tokyo, Japan) using a body coil. The imaging sequence included a breath-hold single-shot true fast imaging with steady state precession (FISP) sequence (TR/TE/Nex = 1000/66/1, FOV = 340-400 mm, matrix = 275x400, slice thickness/gap = 3.5 mm/0 mm), with the planes oriented to demonstrate the pathology most satisfactorily. The fetus remained in head presentation. MRI showed a large heterogeneous supratentorial mass centered on the pineal region causing an obstructive hydrocephalus (Figure 2). The head was markedly enlarged by a soft tissue mass with cystic components and a large extra-axial fluid collection. Again, no normal brain tissue was detected.

The mass showed multiple cystic components, suggestive of a teratoma. The parents opted for termination of the pregnancy. Because of the severe macrocephaly, a cesarean section was performed, and a male infant was delivered at 30 weeks gestation. He had an Apgar score of 2 at 1 min and 2 at 5 min. At birth, his weight was 1750 g, length was 44 cm, and his head circumference was 48 cm (expected head circumference for 27 weeks' gestation is 25 cm) at birth. Biparietal diameter was 11.7 cm (expected biparietal diameter for 27 weeks' gestation is 6.7 cm). The newborn died two hours after delivery. Postmortem computed tomography (CT) examination of the fetus revealed a centrally located large, lobulated heterogeneous mass containing cystic/solid components with speckled calcification and severe hydrocephalus (Figure 3). The parents rejected the autopsy.

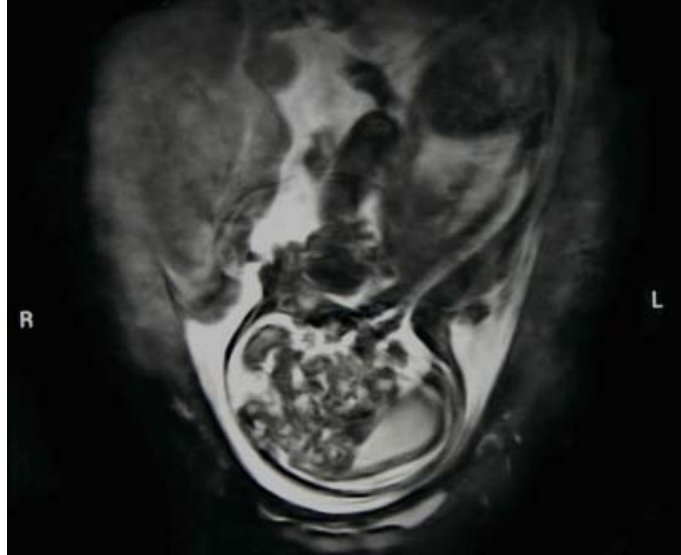


Figure 2. Sagittal true FISP magnetic resonance image showing a large heterogeneous mass centered on the supratentorial region and causing a severe obstructive hydrocephalus. In addition, the fetal brain was underdeveloped.

Discussion

Congenital brain tumors are rare entities which are nowadays often recognized during pregnancy by US and MRI [3]. In literature, first report of a massive congenital intracranial teratoma was published in 1864 by Breslau and Rindfleisch as cited by Washburne et al. [4]. Giant pediatric tumors may present with seizures due to irritation of the cortical gray matter. In more rapidly growing tumors, signs of increased intracranial pressure such as papilledema may occur. Bulging fontanel and macrocephaly may be evident in infants [5-8].

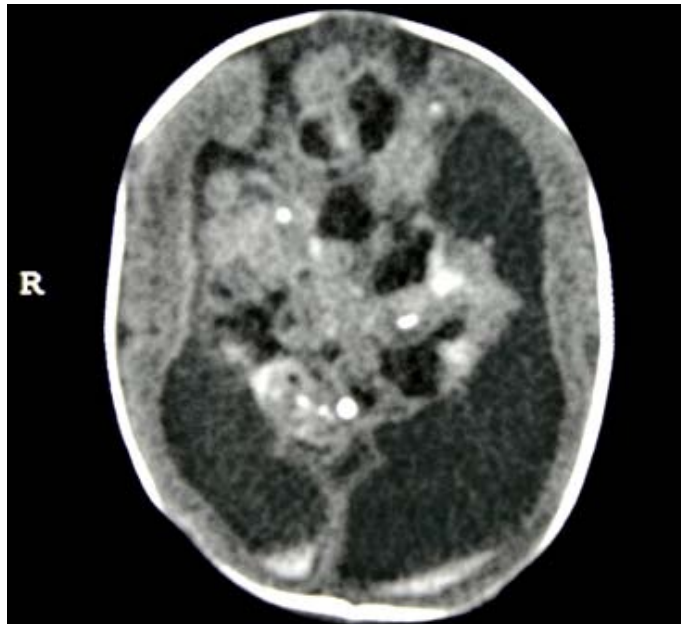


Figure 3. Postmortem unenhanced axial CT image demonstrates a large heterogeneous mass near totally replacing the intracranial content. The ventricles are enlarged (posterior horns of lateral ventricles). It has multiple low-density regions as well as numerous punctate high-density foci, which probably represent calcifications. In addition, CT scan shows blood in the lateral ventricles.

Teratoma is the most common congenital neoplasm composed of tissues originating from all three germinal layers, and may occur in a variety of locations. Teratoma is the most

common tumor in the neonatal period, representing one-third to one-half of all tumors. Previous reports have described several forms of congenital intracranial teratoma, including massive tumors replacing the intracranial contents, smaller tumors producing hydrocephalus, large intracranial tumors extending into the orbit or neck, and incidentally discovered tumors in stillborn infants [6]. Fetal intracranial tumors are rare, and their incidence is unknown. Fetal brain teratoma usually appears as a large, solid and/or cystic tumor, often replacing normal brain tissue and sometimes eroding the skull. Brain teratomas usually cause intrauterine or early neonatal death, and require cesarean section for delivery of an enlarged fetal head [8].

Fetal intracranial tumors are often associated with hydrocephalus, polyhydramnios, and macrocephaly [9, 10]. The initial US finding in our case was hydrocephalus. Intracranial teratoma has been associated with pulmonary hypoplasia and high output cardiac failure [9]. The use of US to assess fetal intracranial structure allows differentiation between tumors, hydrocephalus and other abnormalities. However, experience in the prenatal diagnosis of brain neoplasm is limited. Cystic tumors and teratomas are usually characterized by complete loss of the normal intracranial architecture [11]. Prenatal US has been used in diagnosing intracranial teratoma in more than 40 reported cases to date [1, 12]. The diagnosis is made between 20 and 40 weeks gestation and is usually suspected because of a sudden increase in uterine size resulting from tumor growth and polyhydramnios due to impaired fetal swallowing [7, 8]. The most common initial ultrasonographic finding in the fetus is macrocephaly, and additional features typically include gross distortion or replacement of normal brain tissue by an echogenic mass with multiple cystic components, as well as hydrocephalus secondary to obstruction [13]. At prenatal US, the diagnosis of teratoma should be considered for a complex intracranial mass with calcifications. However, the differential diagnosis for an ultrasonographically diagnosed intracranial mass also includes astrocytoma, ependymoma, craniopharyngioma, choroid plexus cyst, and hemorrhage. In the present case, prenatal US at 28 weeks gestation showed an ill-defined echogenic mass without an apparent cystic component.

Several recent case reports have described the use of fetal MRI between 25 and 36 weeks gestation in helping confirm the diagnosis of intracranial teratoma, and the typical appearance is a large, heterogeneous mass with cystic components on MRI [12, 14], with no apparent difference between mature and immature teratomas. Similar features have been described at postnatal MRI of mature and immature intracranial teratomas. Fetal MRI allows enhanced global imaging of these masses. Anatomical relationships and tissue characteristics are demonstrated by MRI with superior detail, except for calcifications (which are better noted with US).

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 [15, 16]. In our case, antenatal MRI performed at 29 weeks gestation showed an ill-defined supratentorial mass without discrete cystic elements.

In conclusion, US is still the first-line diagnostic examination for prenatal screening in fetal anomalies because of its safety, easy access, low cost, and real-time capability. However, recently, fetal MRI as a non-invasive, fast, highly informative examination has become a valuable adjunct to prenatal US, especially for those with obesity or oligohydramnios. To our opinion, the use of prenatal MRI in addition to US is a valuable tool in utero diagnosis and counseling for a large fetal intracranial mass.

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