

# Prenatal Ultrasonographic Diagnosis of Congenital Intracranial Teratoma

## KONJENİTAL İNTRAKRANYAL TERATOMUN PRENATAL ULTRASONOGRAFİK TANISI

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### Abstract

Congenital intracranial teratomas are rare tumors presenting with macrocephaly, bulging anterior fontanel, hydrocephaly and polyhydramnios. The prognosis in these cases has been poor, with stillbirth or immediate postpartum death as the usual outcome. They are usually diagnosed by routine antenatal ultrasound examination. We present a case of immature intracranial teratoma detected sonographically at 28 weeks of gestation and confirmed by autopsy.

**Key Words:** Congenital intracranial teratoma,  
ultrasonography, prenatal diagnosis

### Özet

Konjenital intrakranyal teratomlar makrosefali, ön fontanel bombeleşmesi, hidrosefali ve polihidroamnios ile kendisini gösteren nadir görülen tümörlerdir. Bu tür vakalarda prognoz kötüdür, ölü doğum veya ani postpartum ölüm beklenen bir sonuçtur. Çoğu zaman rutin antenatal ultrason muayenesi ile tanı konmaktadır. Biz bu çalışmada sonografik olarak 28. gebelik haftasında tespit edilen ve tanısı otopsi ile teyit edilen immatür intrakranyal teratom vakasını sunuyoruz.

**Anahtar Kelimeler:** Konjenital intrakranyal teratom,  
ultrasonografi, prenatal tanı

Türkiye Klinikleri J Gynecol Obst 2004, 14:332-335

Congenital brain tumors were defined as tumors presenting within 60 days of birth and accounts for 0,5-1,5% of all brain tumors encountered during childhood.<sup>1-3</sup> The most common intracranial neoplasm in prenatal period are teratomas (28,8-50%) and gliomas (25%).<sup>2-4</sup> Teratomas are derived from more than one germ layer and 90% of these from all germ layers.<sup>5</sup>

Fetal intracranial teratoma is often associated with macrocephaly bulging anterior fontanel, hydrocephaly and polyhydramnios. Intracranial teratomas are usually diagnosed by routine antenatal ultrasound examination. Ultrasonographic examination performed because of large fundal height. Magnetic Resonance Imaging (MRI) or Computed Tomography (CT) may be indicated in differential

diagnosis of other intracranial lesions. We report a case of immature teratoma detected sonographically at 28 weeks of gestation and confirmed by autopsy.

### Case Report

A 25- year- old gravida 2, para 1 woman was applied to our perinatology department at 28 weeks of gestation with preterm labour. Her last menstrual period was unknown. A previous transabdominal scan at 20 weeks was reported to be normal. Our ultrasound (US) examination at 28 weeks revealed polyhydramnios and an enlarged fetal head with breech presentation. The biparietal diameter (BPD) was 12.2 cm, corresponding to a gestational age of over 40 weeks, whereas the other fetometric parameters, such as abdominal circumference.

(AC) or femoral length (FL) were consistent with 28 weeks of gestation. Polyhydramnios with an amniotic fluid index of 25.9 cm was also detected. Inside the fetal head, normal brain tissue

Geliş Tarihi/Received: 26.02.2004 Kabul Tarihi/Accepted: 30.04.2004

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was replaced by a large brain tumor with solid and cystic areas (Figure 1). There was no recognizable brain parenchyma. Inside the tumor, calcifications were present. No other fetal anomalies were detected. By these findings, ultrasonographic diagnosis was large intracranial tumor which destroyed normal intracranial structures.

Because of the preterm labour and obvious ultrasonographic imaging of brain tumor destructing normal intracranial structures, MRI was not ordered. Ethic committee decided not to arrest preterm labour because of the poor nature of prognosis of these tumors. Parents were informed. Their consent was also obtained. The delivery was performed by cesarean section because of breech presentation and dystocia due to large fetal head.

The male infant (Figure 2) weighed 2480 gm and had Apgar score of 1 and 2 at 1 and 5 min, respectively. Head circumference was measured as 42 cm (>95 th percentile). The infant died at five hours of age. The mother's postoperative course was normal.

At autopsy the brain weighed 532 gm. When skull was opened in macroscopic evaluation, normal brain parenchyma was replaced by 17x15x7 cm sized tumoral tissue. The maximal width of the brain parenchyma was 1 cm. The tumor was greyish-white, irregular shaped, predominantly solid with cystic areas. The skull bones were extremely



**Figure 1.** A prenatal ultrasonographic imaging of fetal head demonstrating deterioration of normal intracranial anatomy and tumor including cystic and solid structures.



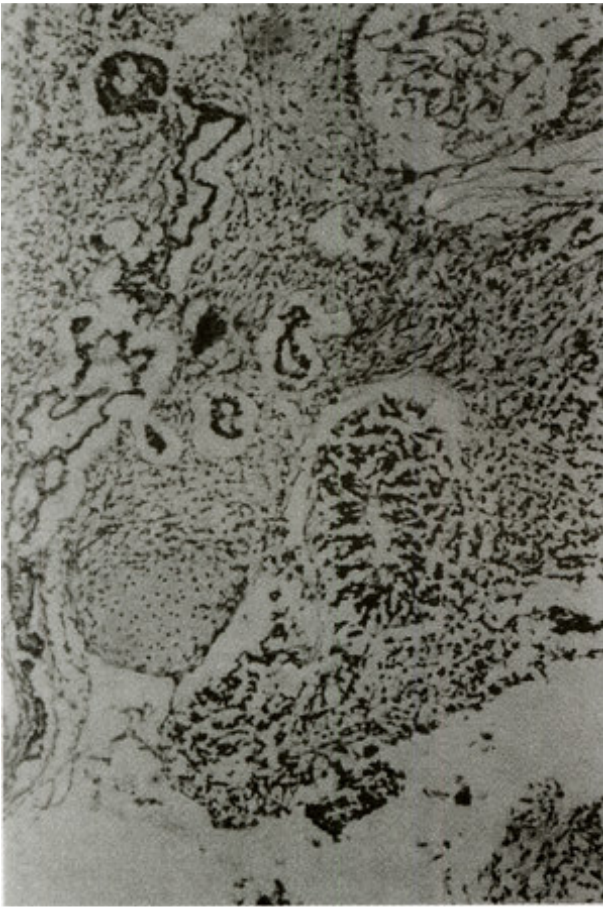
**Figure 2.** Neonate with massive craniomegaly. Head circumference was 42 cm in 28<sup>th</sup> weeks gestation.

thin. The microscopic evaluation revealed immature intracranial teratoma with mature components (Figure 3).

## Discussion

Congenital intracranial teratoma is a rare disease. The first reported case of massive congenital intracranial teratoma was by Bresland in 1964.<sup>6</sup> Weyers et al reviewed 20 cases diagnosed in utero but only one survived with severe intellectual impairment after complete resection. The tumor mass is usually huge and distorts the brain anatomy beyond recognition as in our case.

Up to 14% of children with congenital brain tumors, associated anomalies such as cleft palate, cardiac and urinary tract malformations or polyhydramnios are described.<sup>7-11</sup> This may reflect in



**Figure 3.** Immature teratoma. Immature neural tissue consists of neuroepithelial tubules and cellular glial tissue (top right). Nests of fetal-type cartilage and epithelium-lined tubules or glands are at the bottom.

some cases genetic defects have some roles in pathogenesis of cranial teratomas. Except polyhydramnios these malformations were not present in our case. Because of the size of most tumors it is often impossible to locate the exact site of origin. It's thought that intracranial teratomas originate in the vicinity of pineal gland, quadrigeminal plate or the walls of the 3rd ventricle in a median position. In our case we could not determine the origin of the tumor because of the large destructive nature of the tumor and little residual normal brain tissue. Routine prenatal US can detect fetal cranial tumors in early prenatal period. MRI may be performed when

more accurate anatomic details are required or when ultrasonographic diagnosis of the intracranial lesion is doubtful or limited. In contrary, ultrasonography has the advantage of determining cystic structures and calcifications where MRI is insufficient.<sup>12</sup> The prognosis of congenital teratoma is poor (usually fatal) because of the rapid or invasive growth of the tumors and the destruction of regular cerebral structures.<sup>13</sup> As it is in our case, in most of the cases a normal brain structures are hardly seen or cannot be seen. Surgical excision where possible can be curative but it is only limited to the smaller benign intracranial teratoma. In review of the literature unless the tumor is small, prognosis is poor, mostly lethal. So in our opinion in large destructive tumors like in this case the parents should be informed about the prognosis and termination of pregnancy should be treatment of choice regardless of gestational age. Unless the teratoma is small, the delivery route preferred is cesarean section to avoid dystocia or avulsion of the tumor.<sup>14</sup>

If the intracranial tumor is small or fetus with demonstrable cerebral parenchyma may have the potential to improve following early management of hydrocephalus. So-Hyang proposed that successful resection may be possible in some cases.<sup>15</sup> Although some intracranial teratomas are associated with pulmonary hypoplasia or high output cardiac failure,<sup>16</sup> most are isolated diseases without associated anomalies.

Fetuses with teratomas usually display sudden onset of large cranium. The rapid growth of these tumors is reported by Crade, as in our case, in the 20th gestational week cerebral structures were evaluated as normal. In the 28th gestational week the tumor replaced nearly the whole brain tissue.<sup>17</sup> In differential diagnosis meningoencephalocele should be in consideration. These tumors also enlarge the cranium, mass may extend outside the head. But it is usually possible to identify the midline skull surface and sulci in the mass. Intracranial calcifications may aid the diagnosis of the teratoma which contain teeth or bone tissue. They might be present also in intrauterine infections, but in the

latter hemispheric and ventricular architecture are generally preserved. The other lesions may mimic a teratoma are exencephaly and ischemic brain necrosis. But in all of the patients with teratoma calvaries could be identified. Ultrasound is an extremely useful tool in prenatal diagnosis of fetal intracranial teratomas.

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