CASE REPORT

Congenital Cervical Teratoma

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Abstract

Congenital cervical teratoma is an extremely rare abnormality. Neonatal teratomas occur in about 1:20 000-40 000 live births and only 5% of them are located in the region of the neck. Although mostly benign, they are associated with a high mortality rate due to respiratory distress and require immediate surgical excision. Prenatal sonography is good at identifying the presence of tumors. Early diagnosis of a cervical teratoma can result in various obstetric measures, as far as abortion in the event of a hopeless prognosis. We present a congenital cervical teratoma case diagnosed in the first trimester to discuss prenatal diagnosis, perinatal and neonatal outcomes and the management strategies.

Keywords: congenital, cervical teratoma, pregnancy, management

Özet

Konjenital servikal teratom son derece nadir görülen bir anomalidir. Neonatal teratomlar tüm canlı doğumların 1:20 000-40 000'inde görülürler ve bunların sadece %5'i boyun bölgesindedir. Çoğunlukla benign olmalarına rağmen solunum yetmezliği ve acil cerrahi eksizyon ihtiyacı nedeniyle mortalite oranları yüksektir. Tümör tanısında prenatal ultrasonografi oldukça iyidir. Erken tanı prognozu umutsuz olanlarda, düşük dahil çeşitli obstetrik önlemlerin alınmasına imkan tanır. Biz bu makalede ilk trimesterde tanısı konan bir konjenital servikal teratom vakasını sunduk ve tedavi stratejilerini tartıştık.

Anahtar sözcükler: konjenital, servikal teratom, gebelik, tedavi

Introduction

Teratomas are tumors containing components of all three embryological germ layers. The pathogenesis of these tumors is unclear, but many believe they arise when germ cells escape the developmental control of primary organizers. Congenital teratomas occur in about 1:20 000-40 000 live births and only 5% of them are located in the region of the neck (1).

Prenatal sonography is good at identifying the presence of tumors. It is an essential and reliable method to detect cervicofacial tumors in utero. Because of its ability to characterize the composition of teratomas, as well as the relations of the mass to the large vessels of the neck, ultrasound is the main imaging method for diagnosis. Cervical teratomas are usually large and may lead to difficult labor and problems with the airway patency after birth. The prognosis is good if there is no respiratory compromise. Untreated cervical teratomas are associated with a mortality rate of 80% to almost 100%. On the other hand, it is also possible that the increasing rates of prenatally detected cervical teratomas are responsible for decreasing survival rates, as not all parents want to continue a high-risk pregnancy.

Case Report

A 24-year-old, gravida 2, para 1, pregnant woman referred to our hospital for prenatal control. According to her last menstrual period she was at 16 weeks gestation. The obstetric sonography revealed a 20x17 mm diameter mass on the anterior cervical region. The mass was composed of cystic and solid portions, also containing blood vessels. No other associated fetal anomaly was identified and the karyotype analysis showed a normal (46 XY) fetal karyotype.

After a follow-up period of four weeks, the mass reached to 66x47 mm in diameter and the development of a severe polyhydramnios was recorded (Figure 1). As the tumor showed rapid growth over this time period, the couple was counseled in a nondirective manner about the contingent agg-

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ressive nature of the mass and relatively early gestational age. The couple agreed to termination of the pregnancy at 20 weeks of gestation, which was achieved with intravaginal misoprostol administration.

Postmortem examination revealed a tumor located on the anterior cervical region of the fetus composed of both solid and cystic components. Final pathologic survey was consistent with an immature teratoma with no evidence of malignant elements (Figure 2).

Discussion

Teratomas are exceptionally rare malformations in the head and neck region. They are mostly benign but as a direct result of their rarity, most clinicians' experience of these tumors is very limited, and consequently most of the associated literature consists of single case reports (2). Generally germ cell tumors of the fetus and neonate are histologically benign and are classified as either mature or immature teratomas. Mature teratomas are comprised of well-differentiated tissues and may be pure or a histologic component of a mixed germ cell neoplasm. Immature teratomas may also be a component of a malignant germ cell tumor of mixed histology. The malignant component of mixed histology tumors that contain teratoma is usually of germ cell origin (yolk sac tumor, germinoma, embryonal carcinoma) and only rarely of somatic origin.

The differential diagnosis of prenatally diagnosed fetal neck masses are cystic hygroma, congenital goiter or other thyroid masses, thyroglossal duct, bronchial cleft cyst, cervical hemangioma and cervical lymphangioma. Prenatal sonography is good at identifying the present tumor (3). However, the differential diagnosis of these lesions is often difficult or impossible. If cystic components predominate, then it can be difficult to distinguish from cystic hygroma. Prenatal MRI is now recognized as a useful complementary tool in the assessment of congenital teratomas. The large field of view and better tissue contrast of MRI can provide detailed assessment of the appearance, positions and extent of the mass and invaluable information concerning airway involvement. Hubbard (4) in their review of three cases of giant fetal neck masses found that prenatal MRI more accurately distinguished cystic teratoma from cystic hygroma than prenatal ultrasound. Additional information from fetal MRI can directly affect pre and postnatal management, by confirming equivocal or further characterizing ultrasound-detected anomalies.

Survival with cervical teratoma depends on the size of the tumor and extent of the involved tissues, with respiratory compromise being the main cause of subsequent morbidity and mortality. Fetuses have a much lower survival rate than neonates, 23% versus 85%. Ten percent of the fetuses are stillborn (5). One third of prenatally diagnosed cervical teratomas are associated with maternal polyhydramnios, secondary to the inability of the fetus to swallow amniotic fluid, resulting from the tumor pressing on the esophagus (3,6).

The gold standard of treatment is complete *en bloc* excision of the tumor. The high mortality in neonates is due to inadequate airway control. It is vital that preparation should have been made to ensure adequate control of the airway during or immediately after delivery. Recent attempts at managing the fetal airway before delivery have focused on the surgical control of the airway before clamping the umbilical cord with the ex-utero intrapartum treatment (EXIT) procedure. A multidisciplinary approach to the delivery, with pediatric surgeons, intensivists in attendance is important, so that if endoscopically guided oro or nasotracheal intubation fails, emergency tracheostomy or neck dissection can be performed (7,8).

Unlike a conventional cesarean section, EXIT procedure is the delivery mode of choice when there is a concern for securing the fetal airway. By keeping the uterus relaxed via inhalation anesthetic, uteroplacental blood flow is continued; fetal patent *ductus arteriosus* and low pulmonary vascular resistance are maintained. Thus, continual oxygen is delivered to the patient allowing for up to 60 minutes of valuable additional time to control the airway. In the procedure, the fetal head and shoulders are delivered via hysterectomy; the remainder of the torso and umbilical cord remain in the uterus. Only after the baby is intubated the umbilical cord is clamped and the body delivered. Surgeons must also be prepared for emergent tracheostomy because of the possibility that the endotracheal intubation could fail. If the airway is secured and the tumor is removed in the immediate postnatal period, the outcome is excellent with reported survival rates as high as 85% (8,9). The most dangerous immediate complication at birth is airway obstruction and subsequent hypoxic injury to the newborn. They are usually benign and surgery can be curative.

Malignant change is extremely rare in the head and neck region. The outcome for patients with malignant elements treated with surgery and/or radiotherapy is generally poor. Currently, there are no definite chemotherapy guidelines for neonates with cervical teratomas. Azizkhan (10) recommended chemotherapy for only infants with disseminated metastases and those who have invasive tumors and residual tumor after resection. Long-term follow-up with imaging and α-fetoprotein determinations are additional recommendations.

In conclusion although congenital cervical teratomas are extremely rare neoplasms, its location might compromise the fetal physiology and might lead to fetal or neonatal death. Accurate prenatal diagnosis is crucial for treatment strategies, and the pregnant should be forwarded to tertiary obstetric units where a multidisciplinary team is available for appropriate care of the fetus and the neonate during and after delivery.

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