Case report

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Prenatal diagnosis of a giant epignathus teratoma in the third trimester of pregnancy using three-dimensional ultrasound and magnetic resonance imaging. Case report.

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Abstract

A 20-year-old nulliparous woman was referred due a cervical mass in the fetus in an ultrasound examination performed in the 25th week of pregnancy. The exam revealed an irregular, solid-cystic heterogeneous mass measuring 75x54 mm that came to the exterior through the mouth of the fetus. Three-dimensional ultrasound and magnetic resonance imaging confirmed the diagnosis of epignathus teratoma and the normal finding of the central nervous system. The patient was admitted at 28 weeks, in premature labor. Tocolysis, corticosteroid and amniotic fluid drainage were programmed to be performed before conducting ex utero intrapartum treatment (EXIT). However, there was premature rupture of membranes and the EXIT procedure was brought forward. After premature placental abruption, the newborn's birth was concluded. Tracheostomy was performed, but the newborn's condition progressed to bradycardia and death in a few minutes.

Keywords: prenatal diagnosis, epignathus teratoma, 3D-ultrasound, magnetic resonance imaging

Introduction

Epignathus is a teratoma located in the oral cavity that arises from the base of the skull. Its incidence ranges from 1 in 35,000 to 1 in 200,000 live births and it usually affects the posterior nasopharynx, sphenoid or hard palate [1]. It was first described by Kang et al [2] and is a rare and isolated condition. In rare cases, it is associated with chromosomal syndromes. The prenatal diagnosis is made, in most cases, in the last two trimesters and termination of pregnancy occurs infrequently [3,4]. Two and three-dimensional ultrasound in association with magnetic resonance imaging (MRI), allows early diagnosis with appropriate genetic counseling, provision of information to the couple and preparation of the multidisciplinary team.

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Case report

The patient was a 20-year-old nulliparous woman in her third pregnancy who was admitted to the obstetrics service of Hospital São Paulo, Federal University of São Paulo (UNIFESP) in the 25th week of pregnancy due to a cervical mass in the fetus, for clarification. Two-dimensional ultrasound revealed an irregular, solid-cystic and voluminous heterogenous mass measuring 75 x 54 mm that came to the exterior through the mouth of the fetus and was difficult to evaluate. It seemed not to compromise the central nervous system (fig 1). The fetal weight was below the first percentile and the amniotic fluid index was at the 96th percentile. At the beginning of hospitalization the patient was already showing uterine height

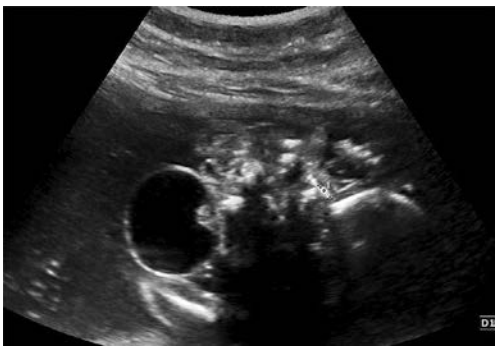

Fig 1. Midsagittal view of the fetal face from two-dimensional ultrasound using a transabdominal approach. An irregular, solid-cystic and voluminous heterogenous mass can be seen, which comes to the exterior through the mouth of the fetus.

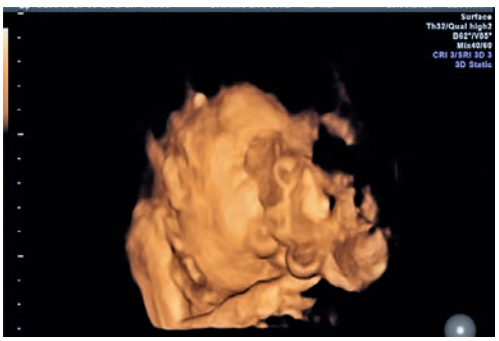

Fig 2. Three-dimensional ultrasound in rendering mode, on the fetal face from an abdominal approach. The mass can clearly be seen projecting through the oral region and occupying the face, with overlapping in the nasal and ocular regions.

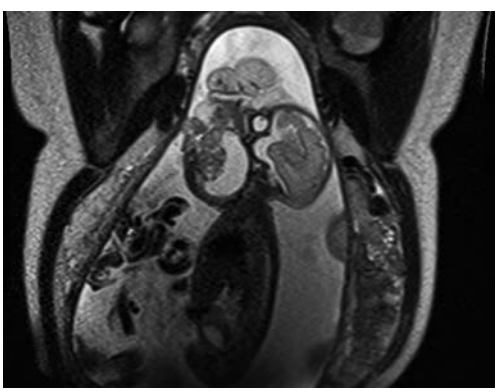

Fig 3. Fetal T2 magnetic resonance imaging in sagittal incidence, showing a mass of large proportions with a solid-cystic component. Note that the eyeball is apparently preserved and the central nervous system has no direct involvement with the lesion.

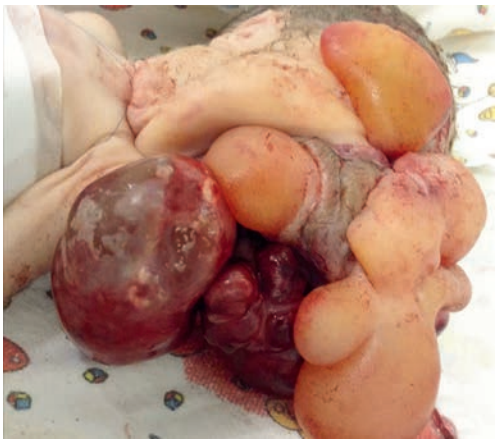

Fig 4. Image of the newborn in the delivery room, showing mass of large volume with a cystic component of large proportions at the lower end. It is not possible to identify the nose and eyes.

greater than would be expected for the gestational age, and she progressed with epigastric and breathing discomfort. It was decided to drain the amniotic fluid: 1360 mL of amniotic liquid was aspirated and specialist prenatal monitoring was provided. In the 28th week of pregnancy, the patient was readmitted due to preterm labor. Three-dimensional ultrasound in rendering mode (fig 2) was performed in order to make an adequate diagnosis. This showed a multilobulated mass, of large proportions and imprecise limits, which came to the exterior at its base through the mouth of the fetus and occupied the nasal and ocular area. Through fetal magnetic resonance imaging (fig 3) with T2 sagittal and coronal incidences, we were able to make a good evaluation of the lesion characteristics, highlighting the solid-cystic formations and the absence of central nervous system involvement. It was also possible to observe the presence of the eyeball just below the mass, apparently without alterations. Because polyhydramnios had recurred, tocolysis, steroids and new amniotic fluid drainage were proposed so that the multidisciplinary team would be able to perform ex utero intrapartum treatment (EXIT) in a timely manner. However, the patient developed premature rupture of the ovular membranes and quickly underwent EXIT. During the procedure, premature placental abruption occurred and the delivery was concluded immediately. The newborn was handed over alive to the neonatology, pediatric surgery and anesthesiology teams for immediate support. The male newborn weighed 1520 g with Apgar scores of 1 in the first minute and 1 in the fifth minute. The umbilical cord pH was 7.069. He was promptly handed over to the neonatology, pediatric surgery and anesthe-

siology teams, and underwent immediate tracheostomy, which was successfully performed in less than 5 minutes. During resuscitation, the newborn's heart rate remained below 100 bpm, despite resuscitation maneuvers. The patient died within a few minutes after birth (fig 4). The mother required transfusion of two units of red blood cells during the operation but remained hemodynamically stable. She was discharged from hospital after 48 hours, without any complications.

Discussion

In addition to epignathus teratoma, the differential diagnoses for masses in the fetal face include hemangioma, congenital epulis (a rare benign tumor of the oral cavity that originates from the anterior alveolar ridge), frontal encephalocele, dermoid cyst, lymphatic malformation and teratoma of the tongue [5]. Some cases of parasitic twin syndrome have also been described in this location [6,7].

Teratomas are tumors derived from pluripotent stem cells and consist of various tissue elements derived from the endoderm, mesoderm and ectoderm. The tissues that are often found in these tumors consist of brain, cartilage, bronchial epithelium and cystic tissues. Epignathus teratoma is a rare form and it is linked to the base of the skull, usually to the hard palate or the mandible [8].

Two-dimensional ultrasound allows appropriate and early identification of the lesion, even at a gestational age of 15 to 17 weeks [9,10]. However, it can be seen from the literature that most of the reports are descriptions from the second and third trimesters of pregnancy [11], thus suggesting that these tumors may present delayed development during pregnancy [12]. Three-dimensional ultrasound in rendering mode is very useful as a method that is complementary to two-dimensional ultrasound, because it allows spatial evaluation of the lesion, as well as assessment of its relationships with the anatomical structures of the face and head. Furthermore, it allows better comprehension of the disease by the parents, thereby facilitating counseling and understanding of the prognosis [13]. Magnetic resonance imaging (MRI) is of importance in the diagnostic detailing, especially in confirming whether the central nervous system is involved, as well as in properly planning for cases in which the fetus will undergo the EXIT procedure [14].

Regarding the imaging differential diagnosis, it is known that oral teratomas present heterogeneity of MRI signals because of their variable contents, which include cysts, fat and calcifications. Lymphatic malformations and encephalocele exhibit cystic characteristics. Dermoid cysts show an intense signal from preserved fat and hemangiomas are represented by imaging of a mass of solid content with diffuse vascularization and high-flow vessels, with intense enhancement through contrast [15].

As well as being infrequent, teratoma epignathus has a poor prognosis and mortality occurs in nearly 100% of the cases. Even with the possibility of surgical removal, death is an expected event for the vast majority of cases, because of the airway obstruction caused by the effect of the mass. The progression of the pregnancy is often complicated by polyhydramnios resulting from extrinsic compression and from the fetus's difficulty in swallowing [16].

Langer et al [17] reported a case similar to ours, in which the fetus underwent tracheostomy for the EXIT procedure before clamping the cord, in the 26th week of pregnancy, due to a teratoma that had been diagnosed through prenatal ultrasound. Although that case evolved to death due to pharyngeal hemorrhage, it illustrates the importance of bringing forward the surgical team's actions in order to treat the patient.

Studies indicate that 60 minutes is the limit for placental support while performing the EXIT procedure [18]. This technique enables maintenance of fetal oxygenation so that there is a secure airway, for proper resection of the tumor. Hirose et al [19] found encouraging results when using this technique for mass resection with prior ventilatory safety. Liecthy et al [20] published a study in 2006, on 23 fetuses that underwent the EXIT procedure, and concluded that diagnostic ultrasound is essential for surgical planning, since many cases may be unfeasible, due to extreme compression of the airways by mass in the face and neck.

In summary, we can conclude that, even though epignathus teratoma is rare condition, it needs to be diagnosed in the fetus as soon as possible. The combination of three-dimensional ultrasound and magnetic resonance imaging provides unquestionable benefits, not only for detailing and confirming the anomaly, but also for indicating the cases that actually will benefit from the EXIT technique, in which surgical correction would be feasible. Given the high complexity of this condition, treatment must be conducted in a tertiary-level referral center, under an experienced multidisciplinary team.

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