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To cite this article: Heron Werner, Carolina Mocarzel, Renato Augusto Sá, Gabriele Tonni, Victoria Arruga Novoa y Novoa, Elyzabeth Avvad-Portari, Paola Bonasoni & Edward Araujo Júnior (2016): Antenatal Diagnosis of a Large Immature Abdominal Wall Teratoma by 2D-3D Ultrasound Using HDlive and Magnetic Resonance Imaging, Fetal and Pediatric Pathology, DOI: 10.1080/15513815.2016.1214199

To link to this article: http://dx.doi.org/10.1080/15513815.2016.1214199

Published online: 25 Aug 2016.
CASE REPORT

Antenatal Diagnosis of a Large Immature Abdominal Wall Teratoma by 2D-3D Ultrasound Using HDlive and Magnetic Resonance Imaging

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ABSTRACT

We describe the first case of prenatally detected teratoma of the fetal abdomen wall using ultrasound and fetal magnetic resonance imaging (MRI). A heterogeneous mass, partly solid and cystic, originating from the anterior abdominal wall of the fetus close to an omphalocele sac was detected by means of 2D/3D ultrasound and MRI. Amniodrainage was performed and due to sign of impending fetal risk, an emergency Cesarean section was performed. A bulky, crumbly and bleeding tumoral mass was confirmed at delivery. Ligation of the supplying artery to the tumor was complicated by uncontrollable hemorrhage and early neonatal death. Pathology identified the tumor as an immature teratoma of the anterior fetal abdominal wall. 2D/3D ultrasound, especially using HDlive application and MRI demonstrated accurate detection and characterization of this congenital tumor.

ARTICLE HISTORY
Received 19 April 2016
Revised 27 June 2016
Accepted 5 July 2016

KEYWORDS
Anterior abdominal wall; prenatal diagnosis; immature teratoma; three-dimensional ultrasound; magnetic resonance imaging; HDlive rendering

Introduction

Teratomas are heterogeneous tumors that may be congenital or may develop later in childhood. Most cases of teratomas are seen in the sacrococcygeal region [1], in the gonadal apparatus [2] or they may originate from rare locations such as the anterior mediastinum [3–5], the retroperitoneum [6] or the anterior abdominal wall. Leva et al. [3] described a mature thoracic-abdominal teratoma, diagnosed in utero that underwent successful postnatal surgical treatment using mini-invasive approach (thoracoscopy and laparoscopy). Fetal teratomas located in the mediastinum can produce a mass location effect causing pulmonary hypoplasia and heart failure, non-immune hydrops as well as fetal esophageal and airway compression causing late-gestation polyhydramnios and preterm labor [5,7].

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Prenatal diagnostic cluster of teratoma relies, by ultrasound, upon detection of a heterogeneous partly solid and cystic tumor mass [2]. Moreover, ultrasound is a useful method to monitor tumor growth pattern and to detect sonographic changes within the tumoral mass and has shown to be accurate even in retroperitoneal immature teratomas in the third trimester of pregnancy [8]. HDlive is a 3D/4D ultrasound application that provides realistic images by means of propagation of light through fetal skin and tissues allowing better visualization of fetal anatomy and has been applied to assess several fetal malformations [9].

To the best of our knowledge, this is the first case describing a large fetal anterior abdominal wall teratoma diagnosed prenatally by means of 2D-3D ultrasound with HDlive rendering and MRI.

Case report

A 25-years-old pregnant woman, gravida 2, with no family history of genetic syndromes, negative serologic tests for TORCH (toxoplasmosis, other, rubella, cytomegalovirus and herpes virus) and no use of medications in the preconceptional period underwent routine ultrasound examination. First trimester screening for common trisomies (combined test) was performed using a Voluson E8 apparatus (GE, Milwaukee, WI) equipped with a transabdominal volumetric RAB 4–8D probe and showed a fetus with a crown-rump length (CRL) of 78.3 mm and a nuchal translucency measurement of 2.7 mm (>95th percentile for gestational age, according to a Brazilian reference range) [10]. Biochemical analysis was assessed using Krypto (BRAHMS, Germany) and showed a \( \beta \)-hCG of 17.90 IU/l (0.513 MoM) and a PAPP-A of 6,580 IU/l (1.570 MoM). Nasal bone was seen. Doppler ultrasound documented a reversed A-wave and a pulsatility index (PI) of 2.70 at the level of the ductus venosus. An omphalocele was detected at first trimester scan. No other structural fetal malformations were seen. After genetic counseling, chorionic villus sampling demonstrated a 46, XX karyotype. Follow-up scan at 22 weeks of gestation revealed polyhydramnios (amniotic fluid index - AF = 26 cm), and confirmed the presence of an omphalocele containing loops of small bowel and liver. Fetal echocardiography was impaired due to presence of the tumor mass that was adjacent to the fetal thorax and face. Clinical and ultrasound controls showed a significant increase in maternal fundal height and considerable growth of the fetal abdominal mass. At 26 weeks and 2 days of gestation, the patient had a fundal height of 36 cm and complained of mild and irregular uterine activity. Cervix was assessed by transvaginal scan and measured 30 mm long with no funneling.

MRI was arranged (1.5-Tesla Magnetom Avanto, Siemens, Erlangen, Germany) and documented an expansive and exophytic mass, heterogeneous in type (solid and cystic), lobulated and with irregular contours in direct communication with the anterior abdominal wall of the fetus at T2-weighted images. The tumor extended from the mesogastric region (immediately above the umbilical cord) to the thoracoabdominal transition, measuring \( 140 \times 98 \times 87 \text{ mm} \) (volume, 620 ml). The mass was partially defined and showed cleavage plane with liver surface and a normal parenchyma signal intensity of the liver. Other abdominal and pelvic structures showed normal appearance (Fig. 1).

Two dimensional ultrasound performed one week later showed slight growth of the tumor size (146 mm \( \times \) 105 mm \( \times \) 104 mm, volume 843 ml) (Fig. 2). Doppler ultrasound documented a poor peripheral vascualrization to the tumor and an umbilical artery PI Doppler of 1.50, a middle cerebral artery Doppler PI of 2.30, a peak systolic velocity (PSV) = 47 cm/s, a ductus
Figure 1. Fetal magnetic resonance imaging (MRI) with T2-weighted sequence in sagittal (A) and axial (B) planes showing a large, heterogeneous mass originating from the anterior abdominal wall (thick arrow) wand extending from the mesogastrium to the fetal face (thin arrow). The tumoral mass was close to the origin of the umbilical cord at the level of the omphalocele sac (star).

venous (DV) PI Doppler of 0.90. 3D ultrasound with HDlive rendering (Zipf, Austria) was applied and provided a realistic imaging of the anterior fetal abdominal wall tumor (Fig. 3). Polyhydramnios (AFI = 33cm) was an associated finding and was treated by amniodrainage of 2500 ml of brownish amniotic fluid. The woman was kept hospitalized on tocolysis regimen (Atosiban infusion for three hours 300 μg/min and 100 μg/min for three hours and 30 minutes after). Doppler ultrasound examination three days after amniodrainage detected an absent diastole with periods of absent/reverse flow in end-diastole and a reversed A-wave (PI above the 95th percentile for gestational age) at the level of the DV. Emergent Cesarean delivery was performed at 28 weeks and 1 day of gestation and a female newborn 1782 g, with Apgar scores at 1- and 5-min of 3 and 5, respectively, was delivered. The hemoglobin (Hb) and hematocrit (Hct) levels in the newborn were 8.7 g/dl and 26%, respectively, after birth. A large exophytic mass originating from the anterior abdominal wall close to the origin of the omphalocele sac (containing loops of small bowel and liver) was confirmed. There was extensive bleeding by the tumor, leading to hemodynamic instability,

Figure 2. Two-dimensional (2D) ultrasound in axial plane in B-mode (A) showing a large, heterogeneous and lobed tumoral mass (partly solid and cystic) coming off the anterior abdominal wall. The tumor showed poor peripheral vascularization and branching at color Doppler ultrasound (B).
which responded poorly to resuscitation maneuvers. Emergency operation led to ligation of the supplying artery to the tumor (in proximity to the omphalocele sac) followed by excision of the mass. Surgery was complicated by hemorrhage at ligation site after 14 hours, and the newborn died due to hypovolemic shock and disseminated intravascular coagulation (Fig. 4). The parents did not give the consent for a full autopsy of the newborn; so only the tumor and the placenta were sent for histopathologic examination.

The tumor originated from the anterior abdominal wall. It weighed 672 g and measured 20 cm × 15 cm × 8 cm. On cut section the mass was brownish, partially cystic, with multiple foci of hemorrhage. Histology showed a mixture of different tissues consistent with congenital teratoma. Immature elements as primitive neuroepithelium with ependymal rosettes were abundant and widely represented. Mature tissues of mesenchymal origin included islands of cartilage, bone and glial differentiation (Figs. 5 and 6). Despite ample sampling, no malignant component was detected.

Placenta was large and heavy, weighing 1,096 g and measuring 27 cm × 20 cm × 4 cm. Grossly, the parenchyma was diffusely pale. Histology showed widespread immaturity with mesenchymal and hydropic villi. Erythroblastosis was seen suggesting intrauterine hemorrhage into the tumor.
Figure 5. Congenital teratoma (Hematoxylin and Eosin, 100X): The image shows a mixture of tissue components from different origin and differentiation: mature glial tissue (thicker arrow), mature cartilage (star), and immature neuroepithelial rosettes (thinner arrow).

Figure 6. Congenital teratoma (Hematoxylin and Eosin, 200X): Immature component composed of neuroepithelial ependymal-like rosettes.
Discussion

Here, we have presented the first case of a third trimester fetus with a tumor originating from the anterior abdominal wall detected prenatally by means of 2D/3D ultrasound using HDlive rendering mode and MRI. Anterior abdominal wall teratomas are rare diseases and prenatal diagnosis of such lesions has been scarcely reported in the medical literature. However, prenatal imaging studies can identify these lesions also when originating from unusual locations [3–5]. Rarely, teratomas may arise from the retropleural space where the tumor can mimic an abdominal mass [11] or from pelvic and ovarian mature teratomas alongside malignant mixed germ cell tumor in the anterior abdominal wall [12]. Although conventional 2D ultrasound is diagnostic in assessing a heterogeneous mass, partly solid and partly cystic, 3D ultrasound may enhance characterization of the tumor tissues as it enables to capture clear-cut realistic anatomical details of the lesions, enhancing image quality, especially using HDlive application. MRI is an important diagnostic tool that may be added to 2D/3D ultrasound to improve detection of tumor edges and its relationship with the surrounding structures. MRI should thus be included in the diagnostic workflow as an implementation of antenatal ultrasound imaging. Nevertheless, Doppler ultrasound may add further information about tumoral vascularization pattern and vascular branching of the tumor. In case of large fetal mediastinal teratomas, Doppler ultrasound can detect a decrease in cardiac output, prolonged acceleration time in the pulmonary artery and an increase in the umbilical artery S/D ratio [13].

Regarding histological examination, pathology revealed a three-germ-layer tumor with immature neuroepithelium consistent with a congenital immature teratoma. Although the mass was widely sampled, no malignant component was detected. The placenta was hydropic, with villi more immature than expected, with erythroblastosis, a finding that suggests ongoing hemorrhage into the tumor while in utero.

Intrauterine as well as perinatal prognosis are dependent upon size and location of the tumor, rapid rate of tumor growth, associated polyhydramnios, and/or degree of intracranial spread, especially in cases of teratomas originating from the oropharyngeal cavity [14,15]. Indicators of poor prognosis on prenatal ultrasound include tumors of large size (>5 cm), polyhydramnios and fetal hydrops [16–19]. Nonetheless, only a previous publication has reported an umbilical cord hernia mimicking a cord teratoma [20] and only in 2007, Sheil and Collins [21] described an early neonatal death due to birth trauma in an undiagnosed intraabdominal immature teratoma. Although conventional 2D ultrasound is diagnostic in assessing a heterogeneous mass, partly solid and partly cystic, 3D ultrasound may enhance characterization of the tumor tissues as it enables to capture clear-cut realistic anatomical details of the lesions, enhancing image quality, especially using HDlive application. MRI is an important diagnostic tool that may be added to 2D/3D ultrasound to improve detection of tumor edges and its relationship with the surrounding structures. MRI should thus be included in the diagnostic workflow as an implementation of antenatal ultrasound imaging. Nevertheless, Doppler ultrasound may add further information about tumoral vascularization pattern and vascular branching of the tumor.

Werner et al. [22] have demonstrated that 3D ultrasound and MRI volumes can be used to produce 3D virtual physical models of different type of congenital anomalies, particularly useful in cases of giant cervical teratoma or congenital neck masses [23]. In such cases, the application of this advanced imaging technique has provided to be of value in the evaluation of the fetal upper airways tract. In addition, 3D virtual physical models have demonstrated to improve genetic counseling and planning accurate perinatal management [24,25].
Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

References


