An Immature Teratoma of the Umbilical Cord: A Case Report and Review of the Literature

Van Keirsbilck Joachim¹*, Serkei Elvira¹, Vanwallegem Lieve², Vanderbeke Ilse³, De Catte Luc⁴, and Decaluwe Wim⁵

¹Institute of Obstetrics and Gynaecology, A.Z. Sint-Jan, Belgium
²Institute of Pathology, A.Z. Sint-Jan, Belgium
³Institute of Obstetrics and Gynaecology, Jan Iperman Hospital, Belgium
⁴Institute of Obstetrics and Gynaecology, University Hospitals Leuven, Belgium
⁵Institute of Pediatrics, Neonatology, A.Z. Sint-Jan, Belgium

Abstract
Teratoma is a rare tumor of the umbilical cord. Teratomas arise from totipotent embryonic cells from all three germinal layers. Teratomas are polymorphic in their presentation. Few have immature elements. Half of the cases present associated anomalies, with omphalocele being the most frequent one. Due to mechanic compression and/or change in fetal hemodynamics teratomas may lead to an increase in perinatal morbidity and mortality.

Our case presents a 30-year-old woman with a cystic mass of the umbilical cord of 4.7 cm at 13 weeks pregnancy. Serial high-resolution ultrasound examination and Color Doppler imaging was used to monitor the expanding mass until a maximal diameter of 21 x 20 x 17 centimeters were reached. Fetal development and well-being remained unremarkably. A cesarean section at 29 weeks and 6 days was imperative due to the worsening maternal condition related to the severely distended abdomen. In order to facilitate the delivery an in utero drainage of the cystic mass was performed. Pathological and histological examinations of the mass revealed an immature teratoma dominantly composed of neuroglial tissue.

ABBREVIATIONS
CM: Centimeter; PI: Pulsatility Index; MCA: Middle Cerebral Artery; MRI: Magnetic Resonance Imaging

INTRODUCTION
Teratomas in the umbilical cord are very rare and originate from totipotent germ cells derived from the three germinal layers into the umbilical cord. Besides from angiomas, they are the only true tumors occurring in the umbilical cord. Only 17 cases have been reported so far [1]. Therefore little is known on the change in fetal hemodynamics and the consequences upon the developing fetus.

We present a case of a teratoma in the umbilical cord, detected at 13 weeks pregnancy, presenting as a mass clearly separated from the fetus.

CASE PRESENTATION
A 30-year old gravida 2 para 1 was diagnosed with a multicystic mass in the umbilical cord with a maximum diameter of 4.7 cm at 13 weeks of gestation. All three umbilical vessels were running alongside the mass. A first trimester combined aneuploidy screening revealed a low aneuploidy risk. The couple was nonconsanguineous and both their medical history was unremarkable. Detailed ultrasound scan at 21 weeks showed a polymorphic heterogeneous mass measuring 11 x 10 x 7 cm, without fetal compression. Because of its characteristics and its large size, the diagnosis of cord teratoma was considered: multiple cystic loci and heterogeneous solid components without overwhelming vascularization in the tumor. The Doppler indices of the umbilical artery and the peak systolic velocity in the middle cerebral artery were all normal excluding compression of the cord vessels and a steel effect. There were no signs of placentaegaly, fetal hydrops or high-output cardiac failure. Ultrasound examination was performed weekly to assure fetal well-being. Corticosteroids were administered at 26 weeks to enhance fetal lung maturation. The fetal mass increased progressively in size from 16 x 14 x 14 cm at 27 weeks to 20 x 20 x 15 cm at 29 weeks of pregnancy.

At 29 weeks and 5 days the patient was admitted with increasing discomfort presented as regular contractions and shortness of breath due to the largely distended uterus. There was no vaginal bleeding, nor amnion leakage. Normal fetal activity was present. However, cardiotocography displayed two to four contractions every ten minutes but with normal reactive heart rate pattern. Ultrasound evaluation revealed an estimated birth weight of 1180 g (50th percentile) with a normal biophysical profile and normal Doppler indices (PI umbilical artery: 1,18; range 0,95-1,55); MCA peak systolic velocity 24,46 cm/sec; range 24,6-46,3); MCA PI 1,60; range 1,5-2,45...). The mass measured 21 x 20 x 17 cm (Figure 1). Arterial flow was present in the mass. The portion of the umbilical cord from the teratoma to the fetus presented two umbilical arteries and two umbilical veins. Both umbilical veins were present in the fetus, one of which connected

*Corresponding authors
Joachim Van Keirsbilck, Institute of Obstetrics and Gynaecology, A.Z.Sint-Jan Brugge, Ruddershove 10, 8000 Brugge, Belgium. Tel: 32-474459268; Fax: 32-50452749; Email: joachimvankeirsbilck@hotmail.com
Submitted: 12 June 2017
Accepted: 19 July 2017
Published: 22 July 2017
ISSN: 2333-6439

Keywords
• Teratoma
• Umbilical cord
• Fetal ultrasound
• Fetal MRI
to the ductus venosus, the other one seemed to drain in the inferior vena cava. MRI demonstrated an atypical mainly cystic though partly solid mass with internal septae and vessels coming from the umbilical cord. The placenta appeared hydropic (Figure 2). There was a normal fetal and placental implantation of the umbilical cord.

Worsening of the maternal condition led to a cesarean section at 29 6/7 weeks, after ultrasound guided aspiration of 820 milliliter from the largest cyst of the mass. A girl of 1335 grams was born with a 1 and 5 minutes Apgar scores of 7/8 and with normal umbilical blood gases.

Pathological examination showed a normalplacenta weighing 690 grams. The total length of the umbilical cord was 32 cm. At 16 cm from the placenta insertion there was the multiloculated mass measuring 22 x 20 x 10 cm and still weighing 725 grams (Figure 3).

On microscopic examination, three umbilical vessels (two venous and one arterial) were observed in the proximal part of the umbilical cord. Instead in the distal part (from the mass towards the fetus) an additional vein was present. The cystic mass was of a mixture of tissues originating from all three germinal layers, but the majority being of ectodermal origin, consisting mostly of glial tissue, but also including remnants of cerebral, retinal and plexus choroides tissue. Epithelial structures such as multilayered squamous epithelium with or without keratinization, hair and sebaceous glands were also present. The endodermal layer was represented by fragments of colon tissue, endo- and exocrine pancreas tissue and immature respiratory ciliary epithelium. As mesodermal structures mature and immature bone, cartilage, muscle, immature connective tissue, fat tissue and lymphoid tissue were shown. The final diagnosis was an immature teratoma of the umbilical cord with a dominant component of neuroglial tissue. The infant is thriving well one year after birth.

**DISCUSSION**

Polymorphic cystic masses of the umbilical cord are rare anomalies. Some are detected prenatally, but most of them are discovered at birth. Its origin, size and location determine the variable clinical impact of the mass. Their progressive growth can lead to vascular compression, hemodynamic changes in placenta and/or fetal circulation, impaired fetal growth and even fetal demise.

The first case of cord teratoma was described in 1887 [2]. It is the only true tumor of the umbilical cord, beside angiomas. Teratomas contain remnants from all thee germinal layers (ectoderm, endoderm and mesoderm), mostly in mature form. Immature derivatives, like in our case, have only been described in four additional cases [3-5]. The histogenetic origin of teratomas is still under review. Wagner et al suggested that the extragondal teratomas arise from pluripotent diploid precursor cells that have not yet undergone the first meiotic division, or from pluripotent embryonal or extra embryonal cells [6].

The presentation of an umbilical cord teratoma may vary considerably, not only in size -from a few centimeters to over 20 cm-, but also in appearance- cystic, solid or mixed-and location-the entire length of the umbilical cord [7].
Table 1: A review of the literature for umbilical cord teratomas.

<table>
<thead>
<tr>
<th>Author</th>
<th>Time of diagnosis</th>
<th>Gender</th>
<th>Placenta</th>
<th>Length of cord</th>
<th>Localization of the teratoma</th>
<th>Volume of the teratoma</th>
<th>Associated malformations</th>
<th>Delivery / Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Budin, 1878</td>
<td>At birth – term</td>
<td>Female</td>
<td>Unknown</td>
<td>Unknown</td>
<td>20 cm from the abdomen</td>
<td>Adult’s fist</td>
<td>Adult’s fist</td>
<td>vaginal birth, slight cyanosis, good</td>
</tr>
<tr>
<td>Haendly, 1923</td>
<td>At birth – term</td>
<td>Unknown</td>
<td>Normal</td>
<td>45 cm</td>
<td>10 cm from the abdomen</td>
<td>Child’s head</td>
<td>Umbilical hernia</td>
<td>vaginal birth, post-operative death</td>
</tr>
<tr>
<td>Hartzand van der Sar, 1945</td>
<td>At birth – term</td>
<td>Female</td>
<td>Unknown</td>
<td>Unknown</td>
<td>4 cm from the abdomen</td>
<td>Duck’s egg</td>
<td>None</td>
<td>vaginal birth, second tumor in umbilical region, neonatal death at 4 months</td>
</tr>
<tr>
<td>Krebsy, 1958</td>
<td>At birth – 8 months</td>
<td>Female</td>
<td>Unknown</td>
<td>36 cm</td>
<td>16 cm from placenta</td>
<td>Unknown</td>
<td>None</td>
<td>vaginal birth, stillbirth, small rupture of the cord near the placenta</td>
</tr>
<tr>
<td>Fujikuraan-Dwellings, 1964</td>
<td>At birth – 8 ½ months</td>
<td>Male</td>
<td>16 x 15 x 2,5 cm, 540 gr</td>
<td>34 cm - SUA from the placenta</td>
<td>1.5 cm from the placenta</td>
<td>3 x 1.4 x 1.2 cm</td>
<td>Poly malformation, hydrocephalus, myelomeningocele, absent right kidney, absence of right supraumbilical member</td>
<td>neonatal death at 1 month</td>
</tr>
<tr>
<td>Heckmann et al., 1972</td>
<td>At birth – term</td>
<td>Female</td>
<td>Unknown</td>
<td>20 cm diameter 550 gr</td>
<td>70 cm – SUA from the placenta</td>
<td>25 cm from the abdomen</td>
<td>9 x 7 cm</td>
<td>None, Good</td>
</tr>
<tr>
<td>Smith and Majmudar, 1985</td>
<td>At birth – term</td>
<td>Female</td>
<td>17 x 14 x 1.8 cm, 450 gr</td>
<td>40 cm</td>
<td>10 cm from the abdomen</td>
<td>1.8 x 0.6 cm</td>
<td>Bladder extrophy</td>
<td>Unknown</td>
</tr>
<tr>
<td>Bersch et al, 1985</td>
<td>At birth – term</td>
<td>Unknown</td>
<td>20 x 18 x 3 cm</td>
<td>15 cm</td>
<td>1 cm from the placenta</td>
<td>10 x 6 x 3 cm</td>
<td>None</td>
<td>Good</td>
</tr>
<tr>
<td>Wagner et al, 1993</td>
<td>At birth – 34 w</td>
<td>Female</td>
<td>14 x 10 x 3.5 cm, 300 gr</td>
<td>Unknown</td>
<td>0.5 cm from the placenta</td>
<td>2.5 cm diameter</td>
<td>None</td>
<td>Hypotrophic fetus, Good</td>
</tr>
<tr>
<td>Kreczy et al, 1994</td>
<td>20 w</td>
<td>Female</td>
<td>Unknown</td>
<td>Unknown</td>
<td>2 cm from the abdomen</td>
<td>10 x 7 x 5, 210 gr</td>
<td>Small omphalocele</td>
<td>vaginal birth at 38 w, Good</td>
</tr>
<tr>
<td>Satgé et al, 2001</td>
<td>12 w</td>
<td>Male</td>
<td>70 gr</td>
<td>18 cm</td>
<td>1 cm from the abdomen</td>
<td>10 x 7.5 x 4 cm, 112 gr</td>
<td>Omphalocele</td>
<td>termination at 17 w – vaginal birth</td>
</tr>
<tr>
<td>Hargitai et al, 2005</td>
<td>16 w</td>
<td>Female</td>
<td>Normal</td>
<td>16 cm</td>
<td>At the wall of the omphalocele (5.5 x 6 x 6 cm)</td>
<td>3 x 3.5 x 4 cm</td>
<td>Omphalocele</td>
<td>Termination at 17 w – vaginal birth, trisomy 13</td>
</tr>
<tr>
<td>Del Sordo et al, 2006</td>
<td>At birth</td>
<td>Female</td>
<td>15.5 x 14.3 cm, 540 gr</td>
<td>40 cm</td>
<td>1.5 cm from the placenta</td>
<td>3.6 x 1.9 x 0.8 cm</td>
<td>None</td>
<td>Good – vaginal birth at 40 w</td>
</tr>
<tr>
<td>Crashes M et al, 2013</td>
<td>18 w</td>
<td>Female</td>
<td>Normal</td>
<td>66 cm</td>
<td>8 cm from the abdomen – 58 cm from the placenta</td>
<td>23 x 16 x 15 cm, 2515 gr</td>
<td>Omphalocele, atrioventricular canal defect, bowel dilatation</td>
<td>cesarean at 37 w, malrotation, intestinal duplication, 2680 gr</td>
</tr>
<tr>
<td>Keene et al, 2013</td>
<td>20 w</td>
<td>Female</td>
<td>Unknown</td>
<td>Unknown</td>
<td>At the wall of the omphalocele</td>
<td>8.5 x 5.5 x 4.5 cm</td>
<td>Omphalocele</td>
<td>vaginal birth at 38 w, small bowel resection, small bowel dilatation, good, 3763 gr</td>
</tr>
<tr>
<td>Chawali et al, 2014</td>
<td>20 w</td>
<td>Female</td>
<td>Unknown</td>
<td>Unknown</td>
<td>At the wall of the omphalocele (11.2 x 8.0 x 5.8 cm)</td>
<td>10 x 7 x 5 cm</td>
<td>Omphalocele – duodenal atresia</td>
<td>vaginal birth at term neonatal death 2 days after surgery</td>
</tr>
<tr>
<td>Van Keirsbilck et al, 2016</td>
<td>13 w</td>
<td>Female</td>
<td>22 x 23 x 1.8 cm, 690 gr</td>
<td>32 cm – additional vein from the placenta</td>
<td>16 cm from the placenta</td>
<td>22 x 20 x 10 cm, 725 after drainage</td>
<td>None</td>
<td>Cesarean at 29 wks, 6 days, good, 1335 gr</td>
</tr>
</tbody>
</table>

Abbreviations: CM: centimeter; GR: grams, SUA: single umbilical artery, W: weeks, D: days
an increased perinatal morbidity and mortality. As in teratomas, mechanical compression of adjacent blood vessels can have an impact on the fetal hemodynamic state [13]. Hemangiomas of the umbilical cord originate mostly from the umbilical artery, rarely from the vein. They consist of angiomatous nodules with degenerated Wharton’s jelly and edema. Mostly located near the placental end of the cord, they easily can be detected by ultrasound imaging and by the presence of blood flow on Color Doppler. Their high complication rate of 35% is associated with the presence of co-existing factors like non-immune hydrops fetalis, intrauterine growth retardation, fetal hemorrhage, intrauterine fetal death or maternal obstetrical complications [8].

Our case is the earliest detected (at 13 weeks pregnancy) umbilical cord teratoma of the 17 cases documented in the literature (Table 1). The wide range in presentation has been repeatedly described. There is a pre-dominance for female fetuses (13/17). Half of the cases are associated with additional anomalies; most frequently omphaloceles, but also an umbilical hernia, bladder extrophy and myelomeningocele with hydrocephalus [8]. One case with an associated omphalocele revealed a trisomy 13 [14]. A teratoma in the umbilical cord also has to be distinguished from a holoaracoid amorphous, which is a compilation of monochorionic twinning. Due to the reversed arterial perfusion of the co-twin by the pump twin, the hypoxemia results in a variable degree of skeletal development and umbilical cord formation [15].

Teratomas may be associated with increased perinatal morbidity and mortality. The management of those pregnancies is not clearly defined yet, due to the rare occurrence and to the delay in diagnosis. In our case, the early detection ensured regular monitoring of the growth and vascular behaviour of the mass, profound scanning for associated anomalies and monitor the overall well-being. Compromise of the fetal hemodynamic state was anticipated because of mechanic compression of the umbilical vessels, and the potential of secondary thrombosis or arteriovenous fistula formation, all of which were absent in our case. Fetal MRI may be of added value in the differential diagnosis. We performed a cesarean section because of deteriorating maternal condition and preterm contractions due to the rapidly expanding umbilical cord mass. In utero drainage of the intra-cystic fluid, helps to decompress the mass and facilitate the delivery. However, it carries a small risk of haemorrhage into the tumor and subsequent fetal jeopardy. Vaginal delivery should be discouraged in large and predominantly cystic lesions because of the risks of dystocia, sudden rupture of the cystic mass and the surrounding blood vessels.

Histopathological investigation is essential to establish a definitive diagnosis.

CONCLUSION

Teratomas in the umbilical cord are rare tumors which can be detected prenatally as mixed solid-cystic masses. Serial ultrasound and Doppler examinations are used to monitor their size and the overall fetal well-being. Their prognosis is largely determined by associated anomalies and the hemodynamic changes in the fetus.

REFERENCES