

Prenatal Ultrasound Diagnosis of Cervical Teratoma

- case report -

Abstract

The fetal face and neck represents one of the key anatomic regions in ultrasound investigation from a psychological and clinical standpoint point of view. Although the sonographic appearance of some masses is characteristic, a substantial overlap exist in echotexture of the masses of the face and neck. In addition to analysing the echotexture of the mass, attempts should be made to determine: the site of origin, the presence of distortion of surrounding anatomy, the presence or absence of other malformations and polyhydramnios. Cervical teratomas are very rare and account for 5% of all teratomas. The majority of them are benign, often large and produce airway obstruction. We present a rare case of a very large cervical teratoma diagnosed at a late gestational age.

Keywords: cervical teratoma, sonographic diagnosis, polyhydramnios

Crîngu Ionescu¹,
Horațiu
Haradja²,
Diana
Gheorghiu²,
Teodora
Vlădescu³,
Irina Pacu¹,
Mihai Dimitriu¹

1. University of Medicine
and Pharmacy "Carol Davila",
Department of Obstetrics
and Gynaecology, Emergency
Clinical Hospital
"St. Pantelimon" Bucharest
2. Department of Obstetrics and
Gynaecology,
Emergency Clinical Hospital
"St. Pantelimon" Bucharest
3. Department of Pathology,
Emergency Clinical Hospital
"St. Pantelimon" Bucharest

Correspondence:
Crîngu Ionescu
e-mail: antoniugineco@
yahoo.com

Introduction

Cervical fetal abnormalities are rare. Among these, the most important cervical tumors are: cystic hygroma, occipital encephalocele, occipital myelomeningocele, cervical teratoma. Hemangioma, lymphangioma or thyroid goiter are rarely detected. Ultrasound examination of the neck must include: the occipital region, the cervical spine, the oropharynx and the neck vessels.

Case study

The patient DM aged 35, presented to the ER on September 13th 2010 for rhythmic uterine contractions that lasted 25-30 seconds and were 2-3 minutes apart, normal uterine tonus, FHR 130-135/min. Admission diagnosis was 30 weeks pregnant, live fetus, intact membranes, vertex presentation, normal maternal pelvic anatomy, polyhydramnios, labor. At the physical exam we found an abdominal circumference of 154 cm and an uterine height of 45 cm. Local exam showed an abundant yellowish vaginal discharge, the cervix was 6 cm dilated, the amniotic fluid sac was intact, fetal head at -3 station, normal pelvic anatomy. The lab works showed slight anemia with leukocytosis and lymphopenia, cervical presence of *Escherichia Coli*. The patient has had 2 vaginal live births in vertex presentation (3400g, 3450g) and 1 spontaneous abortion. LMP was around February 15th 2010. During the respective pregnancy she had only once visited the General Practitioner in Lehliu Hospital at 22 weeks of pregnancy. The blood work has shown anemia with leukocytosis and lymphopenia. She has not followed the instructions for fetal ultrasound, nor has she returned for follow-up visits.

The ultrasound examination at admission showed: severe polyhydramnios (AFI = 39 cm) (figure 1); at cervical level, between the mandible and anterior

chest wall, a 13/9 cm hyperechogenous tumor, with intense hyperechogenous areas (calcifications), with relatively net contour (figure 2), 2-3 septa delineating some small transonic areas, poorly perfused (figure 3), intense hyperechogenous lung parenchyma, abnormal crossing of the blood vessels emerging from the heart.

The differential diagnosis between a cervical teratoma, a cystic hygroma or thyroid goiter was brought to question.

Cesarean delivery was performed considering the size of the tumor and a 1500 g male fetus was born. He could not breathe on his own nor could he be intubated due to compression exerted by the tumor (figure 4).

Gross pathology exam showed a 15/10/12 cm anterior cervical tumor (figure 5), communicating with the mediastinum, total primitive pulmonary atelectasis (figure 6), and transposition of the great vessels.

The microscopic studies showed the tumor to be composed of varying proportions of immature and mature tissues of ecto, endo and mesoderm origin predominantly represented by glial type neural tissue with neuroectodermal areas and presence of adipose, muscular and cartilaginous tissue (figure 7).

The histopathological conclusion was: immature teratoma.

Comment

Cystic hygroma is the most common fetal cervical tumor. However there are other rare anatomical entities which may originate at this level such as anterior cervical teratoma or thyroid goiter.

Cervical teratomas are infrequent lesions, which from a histological point of view show a predominance of neural cells of ectodermal origin. Sacro-coccygeal teratomas are the most frequent ones, about 50% of neonatal teratomas,



Figure 1. Polyhydramnios



Figure 2. Hyperechoic neck tumour with neck extension



Figure 3. Minimal tumor vascularisation



Figure 4. Macroscopic appearance immediately after birth



Figure 5. Appearance and macroscopic tumor invasion of the mediastinum and pulmonary atelectasis



Figure 6. Appearance and macroscopic tumor invasion of the mediastinum and pulmonary atelectasis

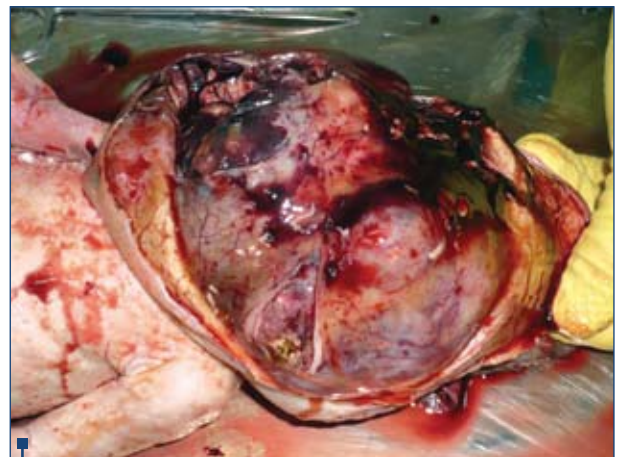


Figure 7. Cervical teratoma

while 5% of them are cervical or orofacial⁽¹⁾. The starting point of the latter can be the cervical region, the nose or the palatine area. Intracranial teratomas can extend to the base of the skull, oropharynx, or face.

The ultrasound diagnosis of cervical teratoma is not very difficult. The tumor can be identified on the midsagittal section with the fetal head in slight hyperextension. The cervical level axial section iden-

tifies the tumor, and sometimes its relationship with other anatomical structures of the neck.

Cervical teratomas are variable in size and have a hyperechoic echostructure (solid) or partially cystic with areas of calcification^(2,3). Teratomas arising from the oropharynx (epignathus) usually occupy the oral cavity, are anteriorly located and can protrude when the fetus opens the mouth. Cervical teratomas

are located anterolaterally, have a greater size and a large base of implantation, usually intersect the medial line, causing hyperextension of the head. Cervical teratomas extend to the face, shifting the respective structures (nose, ear) or to the thorax. Some may present a rich vascularization identified using color Doppler⁽⁴⁾.

Differential ultrasound diagnosis of cervical teratomas is difficult especially when the lesion is large⁽⁵⁾. The differential diagnosis must include: cystic hygroma, branchial cyst, thyroglossal duct cyst, goiter, hemangioma, lymphangioma.

Cystic hygroma is the main differential diagnosis, but sometimes ultrasound differentiation is impossible due to similarities regarding size, location and ultrasound aspect. Ventral developed cystic hygroma is multicystic, with multiple septa of variable thickness. Sometimes it can have a solid appearance because the septa are thick and the spaces between them are small. It should nevertheless be stressed out that the cervical cystic hygroma's edge is not as clearly defined as the teratoma's⁽⁵⁾.

Branchial cysts and thyroglossal duct cysts have an ultrasonographic appearance characterized by larger cystic (transonic) areas or by the multicystic aspect⁽⁶⁾.

The thyroid goiter has a homogeneous ultrasound aspect, increased echogenicity and often a paramedian location. It has clear, well defined limits.

The hemangioma has a homogeneous ultrasound aspect, increased echogenicity, but with increased vascularization that can be shown using color Doppler.

The 3D ultrasound may be useful in establishing the position of the tumor and its relations to the fetal face.

The risk of association between cervical teratoma and chromosomal anomalies or non-chromosomal syndromes is low.

Although the cervical teratoma is a benign lesion, fetal prognosis depends on the following factors: tumor size, location and extension, presence or absence of polyhydramnios. Polyhydramnios association is due to compression on the fetal esophagus and is a negative prognostic factor. If the tumor is small

the prognosis is relatively good, the teratoma being surgically removed after birth. The prognosis is grim if the tumor is large due to secondary compression of the respiratory tract.

Obstetrical attitude in these situations is extremely complex. Cesarean delivery is recommended in the presence of polyhydramnios. Dealing with a compression on the esophagus and upper airways (CHAOS, congenital high airway obstruct syndrome) the neonatal mortality is almost 100%. In these circumstances the only procedure that can be attempted is the EXIT procedure (Ex-utero Intrapartum Treatment)^(7,8): after opening the abdominal cavity and hysterotomy, the membranes are cut, the fetal head is delivered and fetal intubation is performed immediately; only after intubation is achieved, the rest of the fetus is extracted and the umbilical cord is cut. The success of this extremely laborious and difficult procedure depends on the tumor's size and position.

Cesarean delivery is also recommended if polyhydramnios is absent for secondary dystocia due to the presence of the tumor, especially in large tumors.

Surgical removal is the definitive treatment of teratomas. Immediate surgical excision is recommended if the tumor determines the deformation of the neck or deglutition difficulties. If the teratoma is not very large and does not restrict deglutition, monitoring is recommended because it can decrease in size in the neonatal period⁽⁹⁾. This decrease in tumor size occurs only in benign mesenchymal tumors and appears to be due to the fact that tumor growth is sustained in utero by maternal estrogens and birth permanently cancels the hormonal stimulus exerted by the estrogens.

Conclusion

1. The prognosis of fetuses with cervical teratoma is reserved, especially if polyhydramnios is present.
2. Cesarean delivery is recommended.
3. EXIT procedure may decrease the incidence of immediate neonatal mortality, but it is difficult and its success depends on the tumor's volume. ■

References

1. Kerner B., Flaum E., Mathews J., Cervical Teratoma: Prenatal Diagnosis and long-term follow up in Prenat Diagn 18,51, 1998.
2. Shipp T.D., Bromley B., Benaceraff B., The ultrasonographic appearance and outcome of fetuses with masses distorting the fetal face J Ultras Med 14, 673, 1995.
3. Chervenak, Tortora, Moya, Antenatal diagnosis of epignathus J Ultras Med 3, 235, 1994.
4. Chervenak F., Isaacson G., Touloukian R., Diagnosis and management of fetal teratomas Obstetr Gynec 66,666,2001.
5. Cringu Ionescu, Anomaliile craniofaciale și ale gîtului în Ghid de diagnostic diferențial ecografic al anomaliilor fetale, Editura Carol Davila 2010.
6. Kang K., Hissong S., Langer A., Prenatal ultrasonic diagnosis of epignathus J Clinic Ultrasound 6, 330, 1988 .
7. Bouchard S., Johnson M., Flake A., The EXIT procedure experience and outcome in 31 cases J Ped Surg 2002, 37, 418-26.
8. Hirose H., Harisson M., The ex-utero intrapartum treatment (EXIT) procedure Semin Neonat 2003, 6, pg 207-14.
9. Liechty K., Cromblestone T., Flake A., Intrapartum airway management for giant fetal neck masses The EXIT procedure Am Journ Obstetr Gynec 177, 870, 1997.