FOETAL ABDOMINAL TUMOUR – MULTIDISCIPLINARY CHALLENGE BEGINNING FROM THE PERINATAL PERIOD

Maciej Zabielski[1], Agata Golik[1], Joanna Kacperczyk-Bartnik[2], Pawel Bartnik[2#], Agnieszka Dobrowolska-Redo[2], Ewa Romejko-Wolniewicz[2]

1. Students' Scientific Group affiliated to 2nd Department of Obstetrics and Gynecology, Medical University of Warsaw, Poland
2. 2nd Department of Obstetrics and Gynecology, Medical University of Warsaw, Poland

#Corresponding author: Pawel Bartnik, 2nd Department of Obstetrics and Gynecology, Medical University of Warsaw, Karowa 2 St, 00-315 Warsaw, Poland, phone number: +48225966421, e-mail: bartnik.pawel@gmail.com

RUNNING TITLE	Foetal abdominal tumour
KEYWORDS	foetal abdominal mass; foetal tumour; ultrasound examination
WORD COUNT	1452
CONFLICT OF INTERESTS	no conflicts of interest

ABSTRACT

With the development of ultrasound techniques and introduction of routine antenatal examinations, detectability of foetal abdominal masses has increased. It occurs during routine anatomical examination at 18-20 weeks of gestation. A 29-year-old patient at 37th gestational week was admitted to the tertiary referral hospital due to weaker foetal movements. She had an eventful perinatal history – her first pregnancy was lost at 35th gestational week because of true umbilical cord knot. During current pregnancy, a foetal abdominal mass was detected in the ultrasound examination. MRI confirmed its cystic nature. The condition of the foetus was monitored with CTG non-stress test. Foetal movements were normal, with no uterine contractile activity. Because of positive oxytocin test result and the poor perinatal history, the patient was qualified to Caesarean section. At 38th week of gestation, a boy – 3200 g, 56 cm – was born, with 8-8-9-10 pts in the Apgar scale. The neonatal ultrasound examination visualized a 21.5x24x29 mm tumour between the left adrenal gland, spleen and pancreatic tail. On the 4th day, the newborn was transferred to the Paediatric Surgery Clinic. There, after the CT scan, an operation was planned for the next month. Unfortunately, the patient's further history remains unknown. Regular ultrasound diagnostic procedures are essential when foetal tumours are suspected in order to monitor their size and character, and thereby diagnosis and prognosis can be dynamic. To work on an appropriate treatment plan, a cooperation between various centres and physicians of various specialties is vital.

BACKGROUND

About 2% of all paediatric tumours are detected in the newborns [1]. Their prevalence is said to be about 1.7-13.5 per 100000 live births, yet due to the fact that many of these tumours result in stillbirth, or are the cause of pregnancy termination, it may be significantly higher [1, 2]. They may have different features and clinical presentation different from other paediatric tumours [2]. The foetal abdominal cavity is one of the most common locations of foetal tumours, accompanied by tumours located in heart, face and neck [3].

The incidence has been increasing due to the fact that the antenatal evaluation was introduced and generalized, as well as because of the improvement of imaging techniques [4]. Speaking about the latter ones, an ultrasound examination is essential in the diagnostic process, followed by magnetic resonance imaging as a complementary study [2].

These masses are often discovered during routine anatomical assessment at 18-20 gestational weeks, being often cystic in nature [4]. However, they may not reveal their presence until very late and are incidental findings even in the third trimester [3]. They may also influence the management of pregnancy or the mode of delivery [2].

One of the most common among the abdominal masses is neuroblastoma, found in about 1:20000 births, which originates from undifferentiated neural tissue of adrenal medulla (in 90% of cases), or sympathetic ganglia [2, 5] . They derive from the urinary system quite often. In this group, mesoblastic nephroma, also known as renal hamartoma is the most frequent [2, 5]. It is often associated with polyhydramnios and is cured with nephrectomy in the majority of cases [2, 5]. Hepatic tumours, including hepatic mesenchymal haematomas (HMH) are extremely rare and may result in both oligo- and polyhydramnios, hydrops and heart failure [5, 6].

The aim of this article is to provide the readers with the complexity and variability of foetal tumours and the importance of proper management as well as to present a case report related to the subject.

CASE REPORT

We present a case of a 29-year-old patient, gravida 2, para 2. She presented at the tertiary referral hospital in Warsaw at 37th gestational week due to the fact that the foetal movements were significantly weaker. She had poor obstetrical history – her first pregnancy was lost at 35th gestational week due to the true umbilical cord knot. The patient suffered from anaemia during the 2nd and 3rd trimester as well as experienced vaginal bleeding in the 1st trimester. The ultrasound examination performed at 12th week of gestation showed no abnormality in nuchal translucency. The following ones - ultrasound and foetal echocardiography - performed respectively at 22nd and 26th gestational week revealed the presence of a tumour in the abdominal cavity. They were complemented with an MRI scan, during which a 25x16x16 mm cystic mass was detected. A control ultrasound examination before the admission, however, showed that the mass was more of a solid nature than cystic one. It was 21x15x16 mm in diameter, hyperechogenic, weakly vascularized, situated between stomach and descending aorta, and set for further observation. No other defects were detected.

On admission, there was no uterine contractile activity, the amniotic membranes remained intact. She was transferred to the Pathology Ward. During the hospital stay, the condition of the foetus was assessed and monitored with cardiotocography, and the non-stress tests were reactive. Its movements were also normal and there were still no uterine contractions. The patient did not give her consent to labour preinduction with Foley's catheter, as well as to oxytocin test. However, a few days later she gave her consent to the latter one and its result was positive. Because of that, as well as the abovementioned eventful perinatal history, she was qualified for the Caesarean section.

It was performed with Pfannenstiel incision and went on without complications. A boy, measuring 56 cm and weighing 3200 cm, was born. He had 8-8-9-10 pts in the Apgar scale in 1st, 2nd, 3rd, and 5th minute respectively. At first, he had muscular hypotension as well as there was distal cyanosis. The newborn's external examination later on showed no abnormalities.

He had an ultrasound performed soon after birth and the presence of a solid tumour was confirmed. It measured 21.5x23.8x29 mm, was peripherally vascularized and located between the pancreatic tail, the spleen and the left adrenal gland [Figure 1].

After a telephone consultation with the main paediatric hospital in Warsaw, a decision about his planned transfer to the surgical ward was undertaken.

The mother felt well after the operation and on the second day she was transferred to the general obstetric ward with the child. On the 5th day, the patient was discharged in good general condition without the child, as he was transferred to surgical ward before.

At the paediatric hospital a CT scan was performed and having confirmed the diagnosis, the patient was scheduled for surgery in the next month. Unfortunately, the mother and the baby did not present after the operation and their further history remains unknown.

DISCUSSION

Foetal abdominal tumours are undoubtedly a challenge not only because they are a rare condition, but also due to the fact that they are discovered late in pregnancy, as well as they are often accompanied by other defects [3]. However, they may also regress spontaneously [3]. According to one large German study, they regressed in approximately 1/5 cases [3]. The regression ratio was the largest - up to 36% - in gastrointestinal lesions (resolution of dilated intestines) [3].

As for additional defects, it is the case of mostly urinary tract and gastrointestinal tract tumours [3]. For instance, duodenal atresia is often associated with Down and Edwards syndromes, in Down syndrome – 20 to even 40 percent of infants [4]. Chromosomal anomalies are often associated with renal dysplasia and megacystis, less often with renal cysts and anal atresia [3]. Another example is Wilms tumour, or nephroblastoma [7]. It is rare in the neonate, yet more frequent in later age and is

associated with various other syndromes, like WAGR, Perlman (gigantism, dysmorphia and renal abnormalities) and Beckwith-Wiedermann (organ hyperplasia) [7]. Another example of co-existence of defects is the megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS), believed to be caused by de novo mutations in ACTG2 gene, which combine with megacystis and other urinary tract malformations associated with microcolon, gut malrotation and decreased or stopped intestinal peristalsis which may require total parenteral nutrition [8, 9].

One of the most common foetal abdominal tumour is neuroblastoma [5]. It accounts for at least 1/5 or such masses, reaching up to over 50% according to some sources [2, 7]. They are typically located suprarenally and on the right side [5]. Besides the abdomen, it may also be located in the olfactory bulb, chest, pelvis and may cause pressure to the spinal cord, leading to limb and bladder paralysis [7]. It may metastasize as well, for example to bones, especially to lower extremities, skull and the bone marrow, causing pancytopenia [7]. It may coexist with neurofibromatosis type I and with Hirschsprung's disease [7]. It may differ, from pure cystic to solid and of different sizes [5, 7]. Approximately half of cases are recognised before a child reaches 2 years of age [7]. The postnatal survival ranges from 75-100%, however it is 50% or lower if it is diagnosed in stage IV or later in life [1, 7]. The treatment includes surgery and chemotherapy [7].

The most common, however, is echogenic bowel – and by echogenic, as bright as an adjacent bone – present in 1% of foetuses [10]. It may be a sign of meconium ileus or colonic atresia [4].

Some of the tumours are associated with far worse prognosis, for instance the very rare hepatoblastoma, which is also the most common cause of liver malignancy in childhood [1]. It is associated with a survival rate of 22% [1]. It is often associated with extremely low birth weight and can cause a large spectrum of complications, including compression of inferior vena cava, heart failure, foetal hydrops, coagulopathies, anaemia, and metastases [1]. Treatment includes postnatal surgery, sometimes with neoadjuvant chemotherapy [1]. Another aggressive one is hepatic rhabdoid tumour, extremely rare, with younger patients having lower survival rate [11].

CONCLUSIONS

Foetal tumours can have various clinical picture and can present with a wide spectrum of symptoms, therefore they may cause various consequences. It is, therefore, essential that thorough ultrasound examination is performed every time, since they can change their size and character, and may result in different diagnosis and prognosis. New methods are developed – including transplacental treatment or high-intensity ultrasound – to minimalize the invasiveness of treatment [4]. Without any doubt, the most important fact is that these patients should be admitted to tertiary referral hospital and a multidisciplinary approach and cooperation, including neurologists, paediatricians and paediatric surgeons, is required.

CITE THIS AS

MEDtube Science Jun, 2019, Vol. VII (2), 16 – 19

ABBREVIATIONS

HMH – hepatic mesenchymal haematomas
MMIHS – megacystis microcolon intestinal hypoperistalsis syndrome

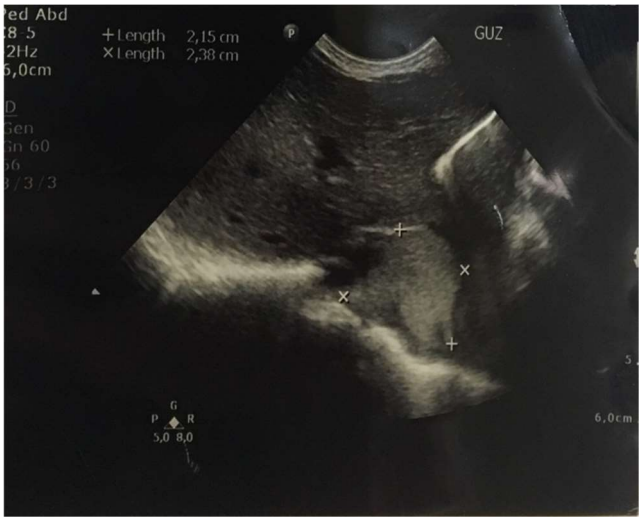
REFERENCES

1. Masmejan S, Baud D, Ryan G, Van Mieghem T. Management of fetal tumors. Best Pract Res Clin Obstet Gynaecol. 2019;pii: S1521-6934(18)30201-3.
2. Cho JY, Lee YH. Fetal tumors: prenatal ultrasonographic findings and clinical characteristics. Ultrasonography 2014;33(4):240–251.
3. Amari F, Beyer DA, Diedrich K, Weichert J. Fetal intra-abdominal tumors: assessment of spectrum, accuracy of prenatal diagnosis, perinatal outcome and therapy at a tertiary referral center. Eur J Obstet Gynecol Reprod Biol 2013;167(2):160-166.
4. Dulay AT MD, Copel JA MD. What you need to know about fetal abdominal masses. Cont OB/GYN 2010;7:24-35.
5. Pilu G, Nicolaides K, Ximenes R, Jeanty P. Fetal tumors. Available from: https://sonoworld.com/Client/Fetus/html/chapter-11/tumors.html. Accessed: 13th April 2019.
6. Abramsky R, Haim AB, Marks K et al. Prenatal diagnosis of a large abdominal cyst – Recommendations and management. J Ped Surg Case Rep 2018;29:22-25.
7. Kawalec W, Grenda R, Kulus M. Pediatria. PZWL Wydawnictwo Lekarskie. Warszawa, 2015: 624-625, 631.
8. Puri P, Shinkai M. Megacystis microcolon intestinal hypoperistalsis syndrome. Semin Pediatr Surg 2005;14(1):58-63.
9. Genetic and Rare Diseases Information Center. Megacystis microcolon intestinal hypoperistalsis syndrome. Available from: https://rarediseases.info.nih.gov/diseases/3442/megacystis-microcolon-intestinal-hypoperistalsis-syndrome. Accessed: 13th April 2019.
10. McNamara A, Levine D. Intraabdominal fetal echogenic masses: a practical guide to diagnosis and management. Radiographics 2005;25(3):633-645.
11. Kapral N, Melmer P, Druzgal CH, Lancaster L. Pediatric hepatic rhabdoid tumor: A rare cause of abdominal mass in children. Radiol Case Rep 2018;13(3):724-727.

LIST OF FIGURES

Fig. 1. Ultrasound image of a hyperechogenic, solid mass in newborn's abdominal cavity.

FIG. 1. ULTRASOUND IMAGE OF A HYPERECHOGENIC, SOLID MASS IN NEWBORN'S ABDOMINAL CAVITY.




MEDtube
sharing
medical
knowledge™