A Case Report of Meconium Ileus-Peritonitis with a Prenatal Diagnosis of Sacro-Coccygeal Teratoma

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Abstract

Backround: To discuss the diffuculties in the diagnosis of meconium peritonitis and to determine the role of magnetic resonance in differential diagnosis.

Case: Twentyfour-year-old woman with a 33-gestational-weeks pregnancy was hospitalized due to polyhydramnios and a fetal pelvic semi-solid mass with areas of calcifications, measuring 66x55 mm in diameter, showing no vascularization that was diagnosed by obstetric ultrasonography, magnetic resonance and fetal Doppler imaging. Antenatal diagnosis of a type 4 sacrococcygeal teratoma turned out to be meconium ileus-peritonitis during postpartum laparotomy and cystic fibrosis was final diagnosis.

Conclusion: When an intraabdominal mass is seen in a fetus with ultrasonograghy and magnetic resonance, especially accompanied by ascites, intraabdominal calcifications and bowel dilatations, meconium peritonitis and ileus should be considered in the differential diagnosis.

Keywords: Meconium peritonitis, sacrococcygeal teratoma, magnetic resonance.

Prenatal sakrokoksigeal teratom tanısı almış bir mekonyum ileus-peritonit olgusu

Amaç: Mekonyum ileus tanısındaki zorluklar ve tanıda fetal manyetik rezonans görüntülemenin yerini tartışmak.

Olgu: Yirmidört yaşında, 33 hafta 3 günlük tekil gebeliği mevcut hastada, obstetrik ultrasonografi, manyetik rezonans ve fetal Doppler incelemelerinde, fetal pelvis yerleşimli, 66x56 mm boyutlarında, semisolid, yer yer kalsifikasyonlar içeren, vaskülarite göstermeyen kitle ve polihidramnios saptanması üzerine hospitalize edilmiştir. Bu antenatal incelemeler sonucu tip 4 sakrokoksigeal teratom öntanısı alan fetus, doğumu takiben yapılan laparotomi sırasında mekonyum ileus–peritonit olarak değerlendirilmiş ve ileri tetkiklerde kistik fibrozis saptanmıştır.

Sonuç: Ultrasonografi ve manyetik rezonans görüntüleme fetusta batında kitle izlenmesi halinde, özellikle de asit, intraabdominal kalsifikasyon, barsak dilatasyonu gibi bulgular eşlik ediyorsa mekonyum ileusu ve peritoniti ayırıcı tanıda düşünülmelidir.

Anahtar Sözcükler: Mekonyum periteniti, sakrokoknigeal teratoma, magnetik rezonans.

Backround

In gynaecology there have been many improvements about congenital anomalies during last 20 years. The main cause of this improvement is the common usage of obstetric ultrasonography. Determination of constitutional anomalies by ultrasonography is about 70%. Thus antenatal control becomes more important.[1] Accurate diagnosis of gastrointestinal system anomalies, just like consti-

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tutional ones, is closely associated with postnatal outcomes becuse of better birth planning, early surgical intervention, fewer complications.

Ultrasonography is essential for the diagnosis of gastrointestinal system anomalies. The most often findings of these anomalies are dilation of bowels, polyhydramnios, hyperechogenic bowel and ascites. Most of these findings are not specific, they can arise lately during pregnancy and they may be due to temporary variations.[2] The sensitivity of obstetric ultrasonography to designate these anomalies depend on the specific characteristics of the anomaly itself.[3] During recent years some groups suggested using magnetic resonance (MR) for the determination of gastrointestinal anomalies or imaging the normal bowel adjacent to the intraabdominal cyst.[4]

In our article a case, which had been diagnosed as type 4 sacrococcygeal teratoma by ultrasonography and obstetric MR but which appeared to be meconium ileus-peritonitis peroperatively, has been presented. Difficulties in the diagnosis of meconium ileus and the place of fetal MR imaging has been discussed.

Case

A 24 year old woman, who gave two vaginal births, who had an operation of ectopic pregnancy applied. She had a pregnancy, one fetus, of 33 weeks and 3 days according to her LMD. After second level ultrasonography and Doppler examination a semisolid mass, localized adjacent to fetal pelvis, having 66x56 mm dimensions, containing some calcifications, having no vascularization and polyhydramnios were seen. Sacrococcygeal teratoma was the prediagnosis. The patient did not have a history of birth with a congenital anomaly. But first degree relative marriage was present (cousins). Obstetric examination findings were: collum was dilated (2 cm), no effacement, head first, head was mobile, pouch was intact. According to obstetric ultrasonography intrauterine, alive, one fetus was present. Biparietal diameter was 82 mm, head circumference was 301 mm, abdomen circumference was 330 mm, femur length was 65 mm. Amniotic liquid index was 40 cm and the placental location was the front wall. In the fetus pelvis a septate mass between urine bladder and columna vertebralis with dimensions 55x70 mm, with heterogeneous echogenicity and calcifications was seen, coexistent with ascites in the abdomen (Figure 1).

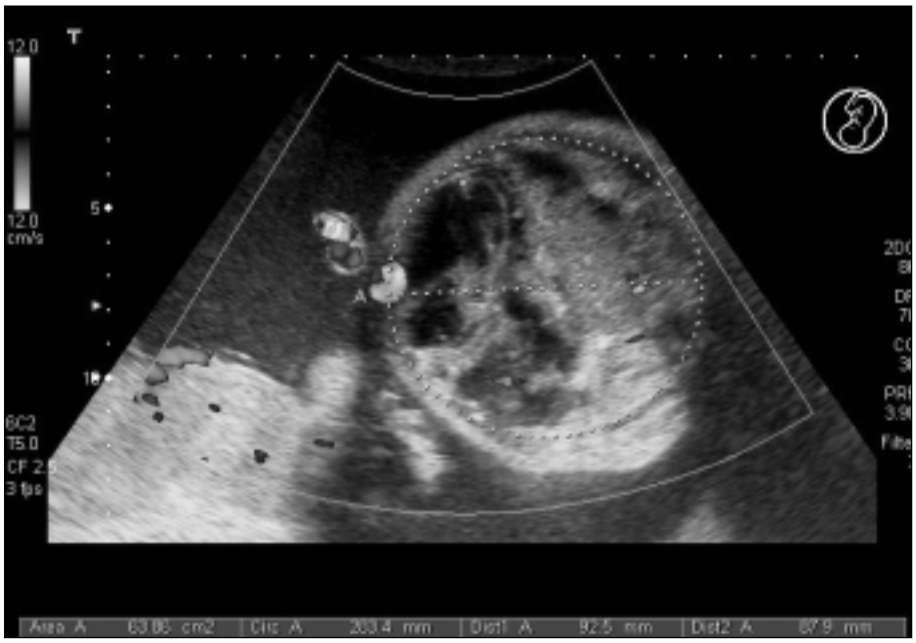
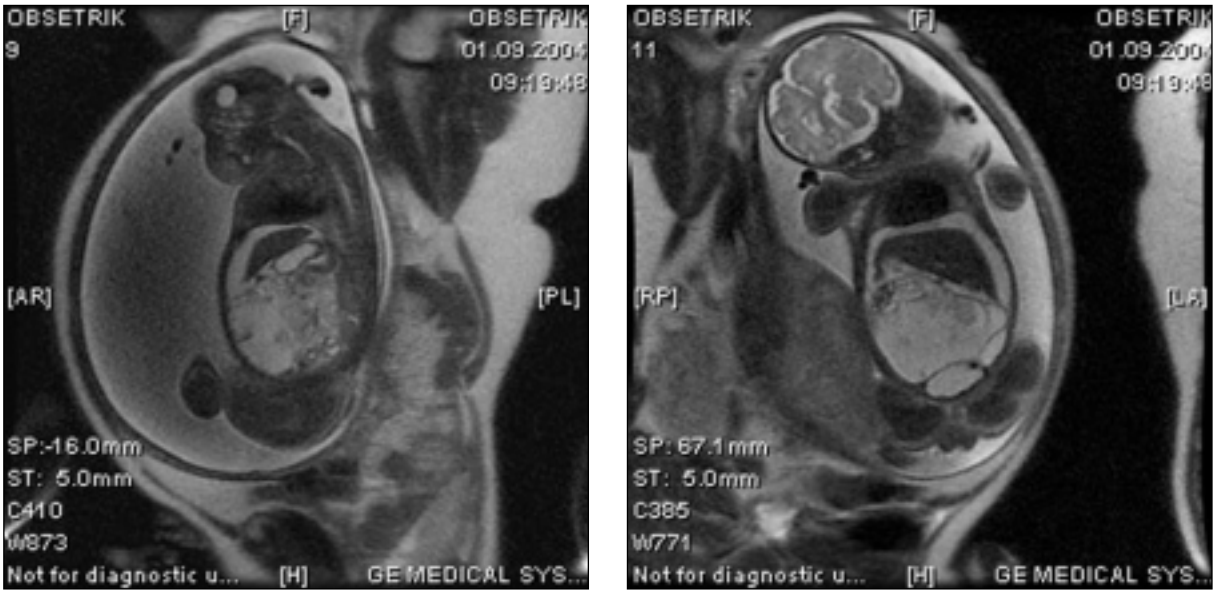


Figure 1. In the fetal pelvis a calcified,septate mass with dimensions of 55x70 mm, with het erogeneous echogenicity. Also ascites in abdomen.

With these findings the patient was sent to Radiology department for obstetric MR imaging. Obstetric MR imaging was made within a 1.5 Tesla (Signa; GE Medical Systems, Milwaukee, Winconsin) MR machine by using body-coil. The fetal position was determined by sonography. Neither maternal premedication nor contrast agent were used for fetal sedation. The images were obtained at axial, coronal and sagittal planes, with Half-Fourier Single Shot Turbo Fast Spin Echo T1 and T2w sequences. Space occupying lesion, filling fetal pelvis and abdomen, consisting of solid components but cystic structure predominantly was measured 60x70 mm. The extension of the lesion lead to presacral area. At the T2w sequences cystic components appeared to be hyperintense, at the T1w sequences they appeared to have hypo-intermediate intensity. Hypointense foci were accepted to be calcifications. Ascites and an increase in the amnios liquid amount in fetal abdomen were coexistent (Figures 2 and 3). Fetal stomach, duodenum and the jejunal loops seemed to have normal diameters (Figure 4). The lumen of rectum seemed to be patent and colon segments could not be visualized. It was thought to be due to the compression of the lesion. Mild dilation in the collecting system of fetal kidneys was owing to the distal compression (Figure 5). It was thought that with the MR findings the space occupying lesion, filling fetal pelvis-presacral area and abdomen, consisting of solid components but cystic structure predominantly, was a sacrococcygeal teratoma with an intraabdominal location (type 4 according to American Academy of Pediatrics).[5] Following examinations were suggested.

At the follow-ups it was seen that the amount of ascites increased. Contractions began at the 20th day of hospitalization. Because the actual approach for the type of birth depends on the dimensions of the tumor (mass > 5cm), the birth took place by cesarean section 1. She gave birth to a girl, 49 cm, 3400 gr, and the first minute APGAR score was 8. The newborn and pediatric surgery groups examined her. Their findings were: heart apex rate 140/min, cardiovascular and respiratory system examinations were normal. There was abdominal distension and mild venous dilation. There were no bowel sounds by oscultation, anus was open and no meconium was seen at rectal touch. There was gas image, filling half of the abdomen, and levels in the plain abdomen radiogram. At first meconium ileus and peritonitis, pouch colon, colonic atresia and aganglionic megacolon was considered. The baby had an operation 36 hours after birth. At laparotomy, all of the bowel walls and the omentum were covered



Figures 2 and 3. Sagittal and coronal images show septate structures, cystic predominant space occupying lesion with a solid content, hypointense calcification on the septa, fetal ascites, polyhydramnios.

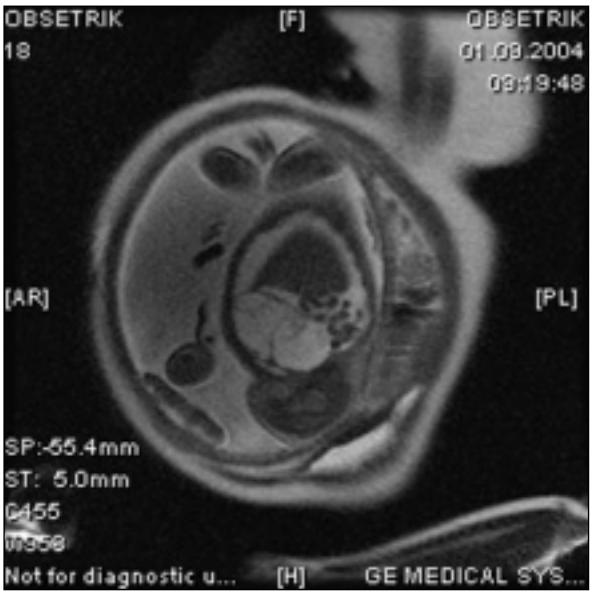


Figure 4. Coronal image shows bowel loops with normal calibration close to the space occupying lesion.

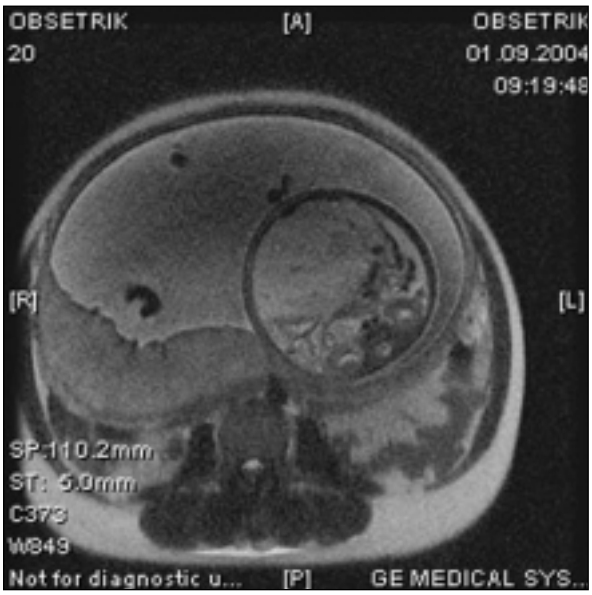


Figure 5. Axial image shows the space occupying lesion and dilation in the renal collecting system bilaterally.

by cystic meconium. Omentum was necrotic and adhered to abdominal wall. There were adhesions. Necrotic omental structures and adhesions were excised. Many loculated cystic structures (meconium cysts) were excised and aspirated. Ascending colon and ileum formed a mass in the form of volvulus. There was ileal perforation 40-50 cm before the ileocecal valve and the distal parts from here were filled with meconium plug untill sigmoid colon. Meconium was decompressed manually to the proximal parts and was cleaned. Loopostomy to the skin was applied at this area in the right lower quadrant. So meconium ileus prediagnosis was confirmed. Biopsy taken from the distal part showed us the presence of ganglionic cells, which eliminated the diagnosis "aganglionic megacolon". İleostomy was closed 2 months later. She was found to have cystic fibrosis, usually accompanied by meconium ileus, after advanced studies had been made. The baby is now 9 months old, weighs 5600 gr and goes on to have therapy.

Discussion

Meconium peritonitis is a rare, fetal and neonatal condition. It especially takes place after antenatal bowel perforation, involving small bowel.[2] Ileum perforated 40-50 cm before the ileochecal valve in our case. Although predispositions are atresia of small bowels, meconium ileus, volvulus and intussusception, mostly it is idiopathic.[6] Ascending colon and ileum formed a mass in the form of volvulus in our case. Meconium peritonitis is classified into three groups; massive generalized peritonitis (type 1), meconium pseudocyst (type 2), prenatal total restoration resulting in residual intraabdominal calcification (type 3). Prognosis is variable and mostly depends on the predisposing pathology and coexistent anomalies.[7] Prognosis is better in the cases of meconium peritonitis without gastrointestinal malformation.[8] In our case meconium ileus was accompanied by volvulus in the ascending colon and ileum.

Although meconium ileus and peritonitis can be diagnosed by ultrasonography, findings are variable. Prenatal sonographic imaging can be supported by findings such as bowel obstruction, meconium pseudocyst, intraperitoneal calcifications, fetal bowel dilation, sometimes by polyhydramnios or fetal hydrops. There may only be the image of fetal ascites.[7] The most typical antenatal ultrasonographic finding is intraperitoneal calcifications. These calcifications form owing to inflammatory reaction stimulated by intraperitoneal meconium and resulting in the calcification of fibrous tissue.[6] When an intraabdominal echogenic

cyst is seen, meconium pseudocyst must be considered for differential diagnosis. After perforation, the bowel content, that spreads into the abdomen, is surrounded by bowels and a fibrous tissue forms around it. This pseudocyst has an irregular and thick wall and may contain debris, septations, calcification or all of them.[7] There was a mass with a heterogeneous echogenicity, septations and calcification and ascites in the abdomen in our case.

Incidence of sacrococcygeal teratoma is 1/40000 and it is the most often seen fetal neoplasia. It may also have similar imaging charasteristics. It originates from presacral area. Depending on the extension to pelvis and abdomen, it is classified into 4 groups. Type 4 is an internal mass with no external component. Its frequency is 10% among all teratomas. 85% of sacrococcygeal teratomas contain solid-cystic mixed forms. Calcification may be seen in 2/3 of the cystic lesions with a thick wall and containing solid components. It is usually accompanied by polyhydramnios. It may be seen with placentomegaly and fetal hydrops. MR is useful in determining the dimensions of the lesion and the abdominal, especially intrapelvic extension of the lesion.[9,10]

Meconium pseudocyst and sacrococcygeal teratoma may have similar imaging charasteristics. Lesion with cystic-solid components, calcified content, polyhydramnios and hydrops may be seen in both conditions. Dilated bowels may also be seen in both meconium ileus-peritonitis and in the condition of being secondary to compression effect of the mass. In both cases proximal bowels may have normal width and normal imaging charasteristics. At MR imaging cystic content appears hyperintense in T2w sequences, in T1w sequences it appears in intermediate intensity, in sacrococcygeal teratoma it may appear hypointense or have intermediate signal intensity compared to cystic content.

In our case of 33 week gestational age the lesion was of mixed structure and contained solid-cystic components. The cystic areas appeared hypoechoic at ultrasonography. MR imaging revealed proximal jejunal loops with normal width and no dilated bowel loops. Sonography revealed no echogenic bowel appearance. In fetal abdomen there was free peritoneal fluid and dilation of renal collector system secondary to compression bilaterally. With these findings, the prenatal diagnosis of our case was type 4 sacrococcygeal teratoma after second level ultrasonography, fetal Doppler and MR imaging modalities were applied. If ultrasonography follow-ups of the pregnant woman had been made properly, dilated-echogenic bowel loops could have been seen earlier and with MR imaging location of obstruction and loops having abnormal signal characteristics could have helped diagnose meconium ileus. Dilated bowels with meconium can be shown at about 27th week; meconium pseudocyst arises about 32nd week.[9]

Veyrac et al studied fetal gastrointestinal system anomalies with MR. They saw that meconium pseudocyst had fluid-like signal in T2w sequences and had intermediate signal intensity in T1w sequences. So they can be differentiated from some other cysts such as duplication cysts.[2] All of intraabdominal cystic masses should be taken into consideration for differential diagnosis. Among these, sacrococcygeal teratoma, which is the most often fetal neoplasia, is the leader.[12] Intraperitoneal calcifications accompanying a complicated cystic mass make the diagnosis easier.[6]

Although Clatworthy defines meconium ileus as a separate condition, he/she and other researchers have reported that this condition could be the first finding of aganglionic megacolon or cystic fibrosis.[2] The diagnosis of aganglionic megacolon was eliminated after peroperative biopsy. Postoperative advanced studies revealed the diagnosis of cystic fibrosis.

Many studies have been made for showing the correlation between the prenatal findings of meconium ileus and peritonitis and newborn outcomes.[7,13,14] Eckoldt et al determined a probable gastrointestinal system anomaly in 96 of 21616 patients retrospectively, and prenatal bowel perforation and/or meconium peritonitis in 11 of these. Prenatal findings of patients, having the diagnosis of postnatal meconium peritonitis or pseudocyst, were intraabdominal cystic echogenic structures accompanied by dilated bowel loops (4 cases), dilated bowel loops (3 cases), free intraabdominal fluid (2 cases), echogenic bowel without dilation (1 case).[7]

The diagnosis of fetal gastrointestinal system anomalies by MR is supplied by abnormal bowel loop dimensions, abnormal bowel loop signal and

abnormal intraabdominal structures accompanied by normal bowel imaging findings. The normal imaging findings of bowel loops, the presence of intestinal dilation and location, and imaging of postatretic bowel loop seem to be reliable in the recognition of meconium ileus and peritonitis by MR.[2] But as in our case, it cannot also be recognized by prenatal obstetric ultrasonography and MR imaging.

MR seems to be more informative for the diagnosis of severe malformations such as megacystitis-microcolon-intestinal hypoperistalsis syndrome, multiple atresia, congenital short bowel.[2] Meconium ileus and peritonitis should be considered in the differential diagnosis, when a mass in the fetal abdomen, accompanied by findings especially ascites, intraabdominal calcification, bowel dilation is seen at ultrasonography and MR imaging. More experience is needed for this subject and the indications of MR imaging should be determined.

References

1. Anteby EY, Yagel S. Route of delivery of fetuses with structural anomalies. *Eur J Obstet Gynecol Reprod Biol* 2003; 106:5-9.
2. Veyrac C, Couture A, Saguintaah M, Baud C. MRI of fetal GI tract abnormalities. *Abdom Imaging* 2004; 29: 411-20.
3. Courteville JE, Gray DL, Langer JC. Bowel abnormalities in the fetus-correlation of prenatal ultrasonographic findings with outcome. *Am J Obstet Gynecol* 1996; 175: 724-9.
4. Saguintaah M, Couture A, Veyrac C, Baud C. MRI of the fetal gastrointestinal tract. *Pediatr Radiol* 2002; 32: 395-404.
5. Elchalal U, Ben-Shachar I, Nadjari M, Gross E, Appleman Z, Caspi B. Prenatal diagnosis of acute bladder distention associated with sacrococcygeal teratoma-a case report. *Prenatal Diagnosis* 1995; 15: 1160-4.
6. Hertzberg BS, Kliewer MA, Bowie JD. Sonography of the fetal gastrointestinal system. In: Sonography in Obstetrics and Gynecology: Principles and Practice. Fleischer AC, Manning FA, Jeanty P, Romero R (Eds). Fifth Edition, Prentice-Hall International Inc, Appleton and Lange, Conneticut, 1996. 411-431.
7. Eckoldt F, Heling KS, Woderich R, Kraft S, Bollmann R, Mau H: Meconium peritonitis and pseudo-cyst formation: prenatal diagnosis and post-natal course. *Prenat Diagn* 2003; 23: 904-8.
8. Shyu MK, Shih JC, Lee CN, et al. Correlation of prenatal ultrasound and postnatal outcome in meconium peritonitis. *Fetal Diagn Ther* 2003; 18: 255-61.
9. Budorick NE. The fetal musculoskeletal system. In: Ultrasonography in Obstetrics and Gynecology. Callen PW (Ed). Forth edition. WB Saunders Company, Philadelphia, Pennisylvannia, 2000. 366-369.
10. Neural tube defects. In: Diagnostic Imaging of Fetal Anomalies. Nyberg DA, McGahan JP, Pretorius PH, Pilu G (Eds). Lippincott Williams@Wilkins, Philadelphia, Pennisylvannia, 2003. 324-329.
11. Siegel MJ, Pediatric and adolescent pelvis. In: Computed Tomography and Magnetic Resonance imaging of the whole body. Haaga JR, Lanzieri CF, Gilkeson RC (Eds). Forth edition. Mosby, St Louis, Missouri, 2003. 2090-2091.
12. Chisholm CA, Heider AL, Kuller JA, Von Allmen D, McMahon MJ, Chesneir NC. Prenatal diagnosis and perinatal management of fetal sacrococcygeal teratoma. *Am J Perinatol* 1999; 16: 47-50.
13. Kamata S, Nose K, Ishikawa S, et al. Meconium peritonitis in utero. *Pediatr Surg Int* 2000; 16: 377-9.
14. Chalibinski K, Deutinger J, Bemaschek G. Meconium peritonitis: extrusion of meconium and different zoographical appearances in relation to the stage of disease. Prenat *Diagn* 1992; 12: 631-6.