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Case Report

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Prenatal diagnosis of congenital mesoblastic nephroma

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Abstract

Objective: Although congenital mesoblastic nephroma is a quite rare tumor, it is the most common renal tumor during fetal and neonatal period. It is usually benign and curable by just a surgical approach consisting of nephroureterectomy. Hence, the prenatal diagnosis of congenital mesoblastic nephroma and distinguishing it from other tumors are essential. In this study, we have aimed to present a case of congenital mesoblastic nephroma which was diagnosed prenatally only by ultrasound in our clinic.

Case: A twenty-five-year-old, 35 weeks pregnant woman was referred to our clinic because of fetal intraabdominal mass. Ultrasonographically, a homogeneous solid mass was detected at the right renal fossa of the fetus. The mass was thought to be congenital mesoblastic nephroma and weekly follow-up was planned.

Conclusion: Prenatal diagnosis of congenital mesoblastic nephroma is possible by a careful ultrasonographic evaluation.

Key words: Congenital mesoblastic nephroma, prenatal diagnosis, ultrasonography.

Konjenital mezoblastik nefromun prenatal tanısı

Amaç: Konjenital mezoblastik nefrom oldukça nadir rastlanan bir tümör olmasına rağmen, fetal ve neonatal dönemin en sık görülen renal tümörüdür. Genelde benign karakterlidir ve sadece nefroüreterektomiden oluşan bir cerrahi yaklaşım ile kür sağlanabilir. Bu durum, konjenital mezoblastik nefromun prenatal tanısını koymayı ve diğer tümörlerden ayırmayı önemli kılmaktadır. Bu makalede, sadece ultrasonografi ile konjenital mezoblastik nefromun prenatal tanısını koyduğumuz bir olguyu sunmayı hedefledik.

Olgu: Yirmi beş yaşında, 35 haftalık gebe, dış merkezden fetal intraabdominal kitle nedeniyle sevk edildi. Ultrasonda, fetüsün sağ böbrek lojunda homojen ekojeniteye sahip solid kitle saptandı. Kitlenin konjenital mezoblastik nefrom olabileceği düşünüldü ve haftalık takibe alındı.

Sonuç: Dikkatli bir ultrasonografik inceleme ile konjenital mezoblastik nefromun prenatal tanısını koymak mümkündür.

Anahtar sözcükler: Konjenital mezoblastik nefrom, prenatal tanı, ultrasonografi.

Introduction

Approximately 2/3 of abdominal masses during fetal and infantile periods are related with renal problems, and most of them are hydronephrosis and multicystic dysplastic kidneys.[1] Although congenital mesoblastic nephroma (CMN) is less than 5% of infantile renal tumors, it is the most frequently seen renal tumor in the first three months of life.[2] Also, it is 80% of renal tumors reported during neonatal period.[3] Congenital mesoblastic nephroma consists of mesenchymal cells and they are generally benign.[4] In this study, we aimed to present and discuss a case that was diagnosed with prenatal CMN in the 35-week-old fetus in our clinic.

Case Report

Twenty-five years old, G2P1 patient who was on her 35 weeks of gestation according to her last menstrual period (LMP) was referred to our clinic due to fetal

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intra-abdominal mass. Her ultrasound imaging was consistent with her LMP, her placenta and amniotic fluid amount was normal, and a single living fetus was observed. A solid mass (47x51x54 mm) which was generally homogeneous, displaying heterogeneity due to necrosis on the center, including some cystic areas, and could not be distinguished from renal parenchyma clearly was observed on the right renal canal of the fetus. In the color Doppler imaging, it was conspicuous that the mass had intense vascularization and these vessels were in circle forms on the periphery (**Figs. 1a** and **1b**). Left kidney, both surrenal glands and other organs were normal. Based on these findings, it was considered that the mass could be congenital mesoblastic nephroma and weekly follow-up was planned.

The size of the mass did not change during follow-ups. The labor began at 38th week and she vaginally delivered a female baby (Apgar score: 8-9; birth weight: 2770 g). In the tomography imaging when infant was 8-day-old, a solid massive lesion originating from the center of right kidney, reaching 5x.5 cm axial size and cannot be distinguished from kidney clearly, coherent with congenital mesoblastic nephroma was found. Right nephroureterectomy was applied to baby when she was two weeks old.

During the macroscopic pathologic examination, a solid, gray-whitish lesion which was partly including thin fibrous septums and was 6x5 cm on renal tissue compressed in a narrow area and material was observed on the right nephrectomy surgery material. When lesion was examined microscopically, fusiform cells creating fascicles beside some normal renal elements, and cellular mesoblastic nephroma focuses with increased cellularity near classical nephroblastoma area were found (**Figs. 2a** and **2b**). In the immunohistochemical analysis of the material, proliferation index was found to be high in SMA (smooth muscle actin) focal positive, desmin-negative and Ki67, and cellular fields. Therefore, it was considered that the mass was a mixed type congenital mesoblastic nephroma. Surgical borders of ureter, renal capsule, perinephritic adipose tissues and vessels were intact. It was only decided to do follow-up and baby was discharged healthily.

Discussion

Perinatal renal tumors are rare and seen about 5% of the cases.[3] Although CMN is less than 5% of infantile renal tumors, it is the most frequently seen renal tumor during fetal and neonatal periods.[2,3,5] CMN can be suspected when solid renal mass found in prenatal sonography is unilateral and in an echogenicity reminding myoma.[4,6,7] Polyhydramniosis accompanies to 70% of cases.[7] Even though the formation mechanism of polyhydramniosis is not known well, mass pressuring gastrointestinal system, polyuria created by renal perfusion increased due to hyperdynamic circulation of tumor, and fetal polyuria triggered by hypercalcemia

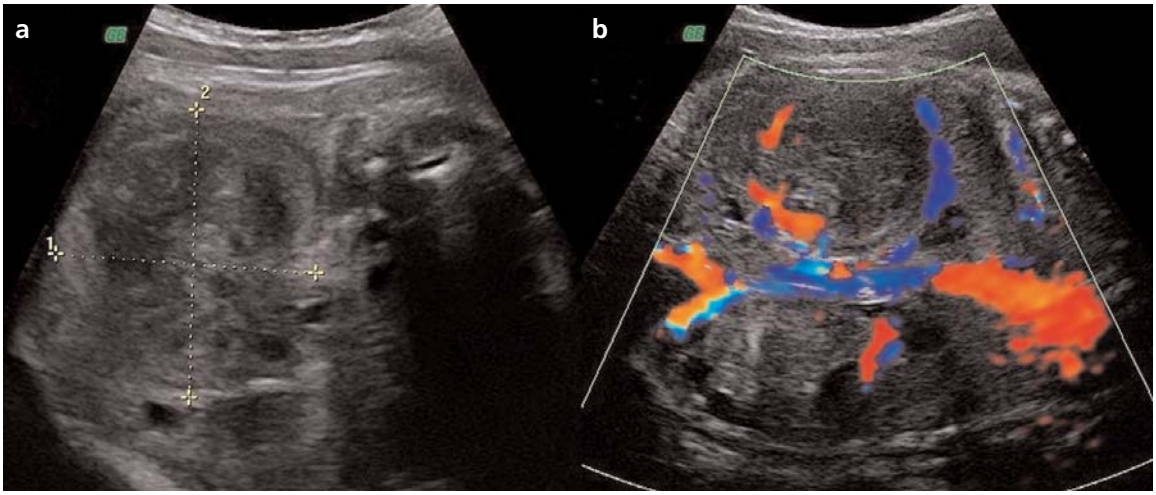

Fig. 1. Axial image of the tumoral mass appearing at the right renal fossa of the fetus (**a**) and the ring view of the highly vascular mass at the color Doppler sonography (**b**).

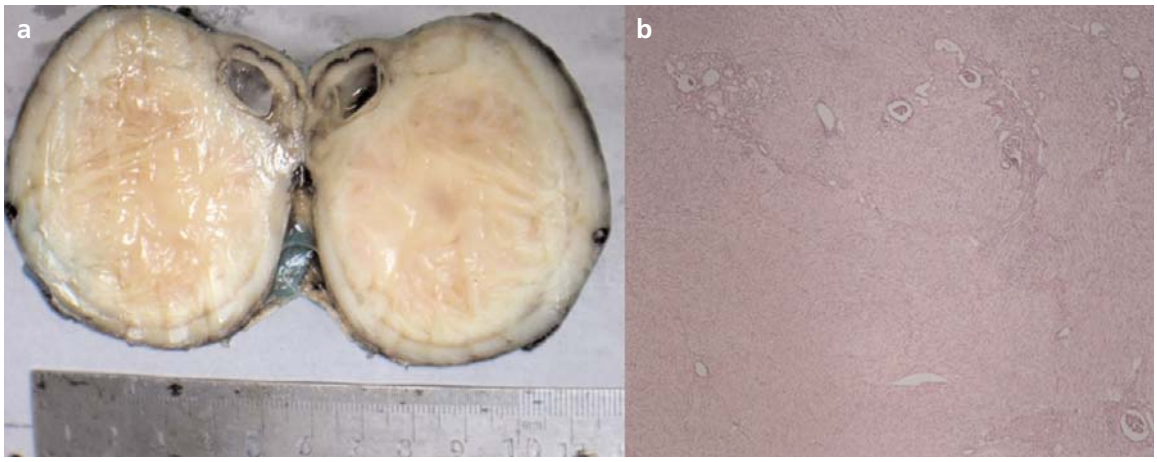

Fig. 2. Macroscopic (**a**) and microscopic (**b**) images of the right nephrectomy material. Microscopy reveals fascicles of spindle-shaped cells and small number of regular renal elements (HE x100).

associated with prostaglandins released from tumor are among the suggested mechanisms.[3,8,9]

Hyperdynamic circulation may cause cardiac failure and hydrops fetalis except polyurea.[4] Four CMN cases which developed hydrops fetalis, took place in the literature where two were diagnosed prenatally were all resulted in death. Therefore, in cases developing hydrops fetalis, it is recommended to deliver baby immediately.[3] In our case, despite the intense vascularization, no polyhydramniosis or hydrops fetalis was developed.

The most important step of the prenatal diagnosis of congenital mesoblastic nephroma is to carry out differential diagnosis of Wilms tumor from adrenal neuroblastoma. Neuroblastoma has a mixed echogenity appearance originating from adrenal gland and seemed to have solid and cystic components in ultrasound imaging. On the other hand, its margins are apparent and it is a mass separate from kidney, direct renal parenchyma invasion is not observed, and it moves asynchronous with kidney during fetal respiration; all these helps to do differential diagnosis of CMN from neuroblastoma.[3,5,6] Wilms tumor is generally surrounded with a capsule having clear margins and invades renal parenchyma completely. In congenital mesoblastic nephroma, as in our case, the margins of the mass may not be distinguished from renal parenchyma clearly.[3,6] Also CMN is an angiomatosis tumor characterized by arteriovenous shunts that may be reflected to color Doppler with intense vascularization and ring appearances.[4,10] Differential diagnosis of the mass can be performed considering that Wilms tumor is rarely seen at prenatal period and during early months of life. Yet, histopathology is inevitable for final diagnosis.[3,6]

Some authors recommend delivery by cesarean section with the concern that rupture of mass may occur during vaginal deliveries.[9] In our case, no traumatic complication occurred during vaginal delivery at term.

Conclusion

With a careful ultrasonographic examination, it is possible to establish prenatal diagnosis of CMN. When CMN is detected during perinatal period, as long as hydrops does not occur, it should be followed up considering that it is a tumor in benign nature. At least, anticipatory approach allowing fetal lung maturity should be preferred. The delivery should be carried out at a tertiary institution which has obstetrician, neonatologist, pediatric surgeon and pediatric oncologist.

Conflicts of Interest: No conflicts declared.

References

1. Glick RD, Hicks MJ, Nuchtern JG, Wesson DE, Olutoye OO, Cass DL. Renal tumors in infants less than 6 months of age. J Pediatr Surg 2004;39:522-5.
2. England RJ, Haider N, Vujanic GM, Kelsey A, Stiller CA, Pritchard-Jones K, et al. Mesoblastic nephroma: a report of

the United Kingdom Children's Cancer and Leukaemia Group (CCLG). Pediatr Blood Cancer 2011;56:744-8.

3. Chen WY, Lin CN, Chao CS, Yan-Sheng Lin M, Mak CW, et al. Prenatal diagnosis of congenital mesoblastic nephroma in mid-second trimester by sonography and magnetic resonance imaging. Prenat Diagn 2003;23:927-31.
4. Won HS, Jung E, Lee PR, Lee IS, Kim A, Kim JK, Cho KS, Nam JH. Prenatal detection of mesoblastic nephroma by sonography and magnetic resonance imaging. Ultrasound Obstet Gynecol 2002;19:197-9.
5. Goldstein I, Shoshani G, Ben-Harus E, Sujov P. Prenatal diagnosis of congenital mesoblastic nephroma. Ultrasound Obstet Gynecol 2002;19:209-11.
6. Fuchs IB, Henrich W, Brauer M, Stöver B, Guschmann M, Degenhardt P, et al. Prenatal diagnosis of congenital mesoblastic nephroma in 2 siblings. J Ultrasound Med 2003;22:823-7.
7. Yamamoto N, Yoshizako T, Uchida N, Kitagaki H, Horie A, Kumori K, et al. Mesoblastic nephroma: a case report of prenatal detection by MR imaging. Magn Reson Med Sci 2006;5:47-50.
8. De Paepe ME, Shapiro S, Young L, Carr SR, Gundogan F. Intrauterine demise due to congenital mesoblastic nephroma in a fetus conceived by assisted reproductive technology. Fertil Steril 2011;95:1119.e15-7.
9. Siemer S, Lehmann J, Reinhard H, Graf N, Löffler G, Hendrik H, et al. Prenatal diagnosis of congenital mesoblastic nephroma associated with renal hypertension in a premature child. Int J Urol 2004;11:50-2.
10. Montaruli E, Fouquet V. Prenatal diagnosis of congenital mesoblastic nephroma. Fetal Diagn Ther 2013;33:79-80.