CASE REPORT

Antenatally detected solid tumour of kidney

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SUMMARY

Congenital renal tumours are rare and usually benign. Polyhydramnios is the most common mode of presentation. Although most cases have been diagnosed postnatally, with advances in imaging technology, an increasing number of cases are being detected on antenatal scans. We describe a case of solid tumour of kidney detected in the second trimester of pregnancy and managed by surgery in the postnatal period.

BACKGROUND

Nearly two-thirds of infantile or fetal abdominal masses are of renal origin. Most are benign and related to hydronephrosis and multicystic dysplasia.¹ Congenital renal tumours are rare, comprising 2.5–7% of all perinatal tumours.² Although imaging plays an important role in the detection of renal tumours and also in the process of differential diagnosis, histology is the most important diagnostic and prognostic factor. Nephrectomy alone is the treatment of choice with a reported survival of over 98%.³

CASE PRESENTATION

A 24-year-old primigravida woman was referred to our department at 20±4 weeks of gestational period due to the presence of polyhydramnios and a fetal renal mass identified on ultrasonography (USG). A detailed USG was carried out in our institute, which showed polyhydramnios and an echogenic mass involving the entire right kidney with hyperechoic rim of the tumour surrounding its relatively hypoechoic centre (figure 1A). The left kidney and bilateral adrenal glands were normal. As symptoms were not severe enough to warrant intervention at that stage, we followed the mass till

term. The woman underwent caesarean delivery (indication obstructed labour) in our institute at term, that is, 37 weeks and gave birth to a 2900 g male neonate. Postnatal examination showed 8×4 cm reniform-shaped mass in the right lumbar region with nodular surface and firm consistency.

INVESTIGATIONS

Further workup was carried out for right renal mass by postnatal USG and contrast-enhanced CT (CECT) scan and α -fetoprotein (>35 000 ng/mL). Postnatal USG showed similar findings as prenatal USG. Inferior vena cava (IVC) and right renal vein flow were normal in USG Doppler. CECT scan of the abdomen confirmed the presence of a low attenuation mass involving the entire right kidney (figure 1B). Opposite kidney was normal. The IVC and the right renal vein were compressed. CT scan of the chest was normal. The complete blood count, serum electrolytes, coagulation profile, renal and liver function tests were normal.

DIFFERENTIAL DIAGNOSIS

A common differential diagnosis in decreasing order of frequency are:

- Congenital mesoblastic nephroma (CMN)
- Wilms' tumour
- Rhabdoid tumour
- Clear cell sarcoma
- Hamartomas (eg, angiomyolipoma)
- Ossifying tumour of infancy
- Metanephric stromal tumour

TREATMENT

Intraoperatively, a solid tumour 8×4×4 cm replacing the entire right kidney (figure 2A) with involvement of renal hilum and minimal adhesion in to the

Figure 1 (A) Transverse axial ultrasound of the right kidney showing an echogenic mass involving the entire right kidney with hyperechoic rim (black asterisk) of the tumour surrounding its relatively hypoechoic centre (white arrow). (B) Contrast-enhanced CT scan of the abdomen showing a low attenuation mass involving the entire right kidney (white asterisk) and normal left kidney.

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Figure 2 (A) Intraoperative photograph showing a solid tumour (white asterisk) replacing the entire right kidney. (B) Cut section of the tumour showing solid homogeneous fleshy mass without haemorrhage, necrosis and cystic degeneration.

adjacent tissue planes was found. The neonate underwent right radical nephroureterectomy. A cut section of the tumour showed rubbery solid homogeneous fleshy mass without haemorrhage, necrosis and cystic degeneration (figure 2B).

OUTCOME AND FOLLOW-UP

The specimen was sent for histopathological examination, which showed monomorphous sheets of closely packed spindle cells imparting a small blue cell appearance and prominent irregular vascular spaces (figure 3A). A magnified view of the histopathological slide showed sheet-like proliferation of plump, atypical spindle cells with abundant cytoplasm, vesicular nuclei and nucleoli; frequent mitotic figures, that is, 25–30/10 on high power field (figure 3B). Histopathological findings were suggestive of CMN, cellular type. At the final follow-up 2 months ago, the child was 2 years of age and doing well. The child was being followed up for last 2 years with 6 monthly USG abdomen and chest X-ray. There was no local recurrence or metastasis.

DISCUSSION

Approximately two thirds of infantile abdominal masses are of renal origin, and most are benign.⁴ Hydronephrosis and cystic renal disease will account for most of these masses. The

differential diagnosis of solid tumours includes CMN, Wilms' tumour and much less common lesions such as rhabdoid tumour, clear cell sarcoma of the kidney, renal cell carcinoma, angiomyolipoma, ossifying renal tumour of infancy and metanephric stromal tumour.^{5–8} If in 1967 Bolande *et al*⁹ was credited for reporting the first case of CMN in infants younger than 3 months of age, then Ehman *et al*¹⁰ should be praised for diagnosing it antenatally, that is, 35 weeks. Over the last two decades, various authors reported similar cases with a prenatal diagnosis.^{11–12} Intrauterine demise due to CMN in a fetus conceived by assisted reproductive technology has been reported.¹³ CMN comprises 2–4% of solid renal tumours.¹⁴ Usually, cases with CMN present with an abdominal mass at less than 6 months of age; however, these lesions may be detected prenatally on routine obstetric ultrasound examination. Polyhydramnios, owing to excessive urine production, may be found in 71% of pregnancies associated with CMN.¹⁵ Pathologically, CMN is divided into two groups (classic and cellular) based on histopathological appearance. Cellular CMN is also distinguished from classic CMN by the presence of a t (12;15) (p13; q25) translocation associated with the fusion of the *ETV6* and *NTRK3* genes.¹⁶ Grossly classic CMN has a firm, whorled appearance and the border with surrounding normal

Figure 3 (A) Histopathological examination of the tumour showing monomorphous sheets of closely packed spindle cells imparting a small blue cell appearance and prominent irregular vascular spaces (H&E stain, ×10 zoom). (B) Magnified view showing sheet-like proliferation of plump, atypical spindle cells with abundant cytoplasm, vesicular nuclei and nucleoli; frequent mitotic figures (H&E stain, ×20 zoom).

kidney is not sharply defined. Cellular CMN usually appears softer with cystic and haemorrhagic areas, and the boundary with non-neoplastic kidney is often clearly demarcated.

Our case presented as antenatally diagnosed right renal mass with polyhydramnios. Diagnosis was made by USG in the second trimester only. Postnatally on day 3 of life, the neonate underwent radical nephroureterectomy. Complete excision without any tumour spillage was achieved. In spite of the cellular variety, we did not encounter any local recurrence or metastasis after a follow-up period of 2 years, as reported by other authors.^{17–20} Prenatal diagnosis is helpful because it improves the pregnancy outcome by institutional delivery with the added participation of a paediatric surgeon if required. As described by Bolande *et al*,²¹ complete surgical resection is the treatment of choice in cases of CMN, especially in cases of classical variety. Children with CMN has excellent outcome at all ages and indications for adjuvant chemotherapy are very few. Primary nephrectomy should be the treatment in children <3 months of age, and in those presenting aged >3 months, alternative diagnoses should be considered.²² For cellular variety, strict follow-up should be carried out and additional chemotherapy can be considered for cases in which local recurrence or metastasis occurs, and those occurring after 3 months of age may behave more aggressively and may need chemotherapy.

Learning points

- Congenital mesoblastic nephroma (CMN) is the most common antenatally diagnosed renal tumour and can be detected prenatally on routine obstetric ultrasound examination.
- In absence of associated symptoms (hydrops fetalis, severe hydramnios), antenatal cases of CMN can be safely followed up until full gestational period.
- Neonate can be safely rescued with radical nephroureterectomy in early postnatal period.
- Strict follow-up is necessary especially in cellular type of CMN.
- CMN cases presenting after 3 months of age may behave more aggressively and may need chemotherapy.

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Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Glick RD, Hicks MJ, Nuchtern JG, *et al*. Renal tumors in infants less than 6 months of age. *J Pediatr Surg* 2004;39:522–5.
- 2 Isaac HJ. Tumors. In: Gilbert Barnes E, ed. *Potter's pathology of the fetus and newborn*. St Louis, MO: Mosby, 1997;2:1242.
- 3 Howell CG, Othersen HB, Kiviat NE, *et al*. Therapy and outcome in 51 children with mesoblastic nephroma: a report of the National Wilms' Tumor Study. *J Pediatr Surg* 1982;17:826–31.
- 4 Pinto E, Guignard JP. Renal masses in the neonate. *Biol Neonate* 1995;68:175–84.
- 5 Davidoff AM, Shochat SJ. Nephroblastoma (Wilms' tumor). In: Ziegler MM, Azizkhan RG, Weber TR, eds. *Operative pediatric surgery*. New York, NY: McGraw-Hill Professional, 2003:1169–80.
- 6 Marsden HB, Lawler W. Primary renal tumors in the first year of life. A population based review. *Virchows Arch A Pathol Anat Histopathol* 1983;399:1–9.
- 7 Lowe LH, Isuani BH, Heller RM, *et al*. Pediatric renal masses: Wilms' tumor and beyond. *Radiographics* 2000;20:1585–603.
- 8 Kacar A, Azili MN, Cihan BS, *et al*. Metanephric stromal tumor: a challenging diagnostic entity in children. *J Pediatr Surg* 2011;46:e7–10.
- 9 Bolande RP, Brough AJ, Izant RJ Jr. Congenital mesoblastic nephroma of infancy. A report of eight cases and the relationship to Wilms' tumor. *Pediatrics* 1967;40:272–8.
- 10 Ehman RL, Nicholson SF, Machin GA. Prenatal sonographic detection of congenital mesoblastic nephroma in a monozygotic twin pregnancy. *J Ultrasound Med* 1983;2:555–7.
- 11 Goldstein I, Shoshani G, Ben-Harus E, *et al*. Prenatal diagnosis of congenital mesoblastic nephroma. *Ultrasound Obstet Gynecol* 2002;19:209–11.
- 12 Guschmann M, Tonnies H, Buhner C, *et al*. Myoid differentiation in mesoblastic nephroma: clinicopathologic and cytogenetic findings of a rare case. *J Pediatr Surg* 2002;37:E22.
- 13 De Paape ME, Shapiro S, Young L, *et al*. Intrauterine demise due to congenital mesoblastic nephroma in a fetus conceived by assisted reproductive technology. *Fertil Steril* 2011;95:1119.e15–17.
- 14 MacLennan GT, Cheng L. Neoplasms of the kidney. In: Bostwick DG, Cheng L, eds. *Urologic surgical pathology*, 2nd edn. Edinburgh: Mosby Elsevier, 2008:77–172.
- 15 Haddad B, Haziza J, Touboul C, *et al*. The congenital mesoblastic nephroma: a case report of prenatal diagnosis. *Fetal Diagn Ther* 1996;11:61–6.
- 16 Rubin BP, Chen CJ, Morgan TW, *et al*. Congenital mesoblastic nephroma t(12;15) is associated with ETV6-NTRK3 gene fusion: cytogenetic and molecular relationship to congenital (infantile) fibrosarcoma. *Am J Pathol* 1998;153:1451–8.
- 17 Bisceglia M, Carosi I, Vairo M, *et al*. Congenital mesoblastic nephroma: report of a case with review of the most significant literature. *Pathol Res Pract* 2000;196:199–204.
- 18 Herman TE, Siegel MJ. Special imaging casebook. Congenital cystic mesoblastic nephroma. *J Perinatol* 2000;20:198–200.
- 19 Ko SM, Kim MJ, Im YJ, *et al*. Cellular mesoblastic nephroma with liver metastasis in a neonate: prenatal and postnatal diffusion-weighted MR imaging. *Korean J Radiol* 2013;14:361–5.
- 20 Santos LG, Carvalho Jde S, Reis MA, *et al*. Cellular congenital mesoblastic nephroma: case report. *J Bras Nefrol* 2011;33:109–12.
- 21 Bolande RP. Congenital mesoblastic nephroma. *Arch Pathol* 1974;98:357.
- 22 England RJ, Haider N, Vujanac GM, *et al*. Mesoblastic nephroma: a report of the United Kingdom Children's Cancer and Leukaemia Group (CCLG). *Pediatr Blood Cancer* 2011;56:744–8.

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