

Case Report

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A rare presentation of antenatally diagnosed Wilms tumor: A case report

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KEYWORDS

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ABSTRACT

Background: Wilms' tumor is the most common renal malignancy in children, with a peak incidence between 1 and 4 years of age. The prevalence of antenatal renal tumors is scarce, around 7 in 100,000 live births. We report a case of an antenatally diagnosed Wilms tumor.

Case Presentation: A 2-month-old infant presented with a right renal lump since birth. The antenatal USG done at 35 weeks of gestation, showed an ill-defined heteroechoic area of size 57.3X29.9 mm at the upper pole of the right kidney, with few cystic areas with increased echogenicity. A right nephroureterectomy was done. It was WT1 and Vimentin positive and focally positive for Cyclin D1, whereas negative for Desmin, SMA, and PAX8, thus confirming the histopathological diagnosis of Wilms' tumor.

Conclusion: Antenatal Wilms' tumor is rarely detected. A high degree of suspicion and active investigations should be conducted in antenatal and immediate neonatal periods for prompt decision-making and better outcomes.

INTRODUCTION

Wilms' tumor (WT) is the most common renal tumor in the pediatric population, with a peak incidence between 1 and 4 years of age [1]. The prevalence of antenatally diagnosed renal tumors is extremely rare, recorded to be around 0.16% of all cases of Wilms' tumor [2]. Here, we report a case of antenatally detected WT managed successfully.

CASE REPORT

A 2-month-old male infant presented to the outpatient department of Paediatric Surgery, with the complaint of a right-sided lump in the abdomen since birth. There was no history of hematuria, fever, or any difficulty in feeding. The general physical examination was unremarkable. On per abdomen examination, there was a smooth-surfaced, hard, non-tender lump with distinct margins, measuring about 5X5 cm palpable in the right lumbar region, slightly mobile in supero-inferior and mediolateral directions. The patient was diagnosed as having a tumor in the right renal area, on antenatal ultrasonography (USG) done at 35 weeks of gestation, showing an ill-defined heteroechoic area of size 57.3 X 29.9 mm at the upper pole of the right kidney with few cystic areas with increased echogenicity (Fig. 1A). Postnatal USG abdo-

men showed a bulky right kidney with a heteroechoic area of size 61X 36.2 mm at the upper pole with few cystic areas, with vascularity on color Doppler (Fig. 1B).

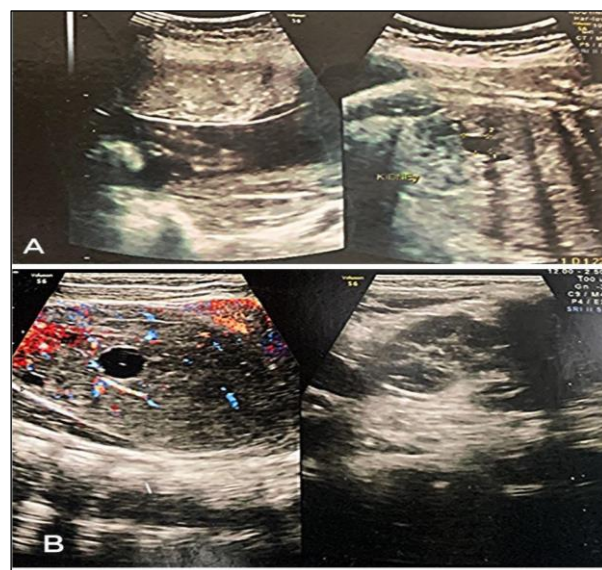


Fig. 1: A) Antenatal USG showing bulky right fetal kidney with multiple small cystic areas, with increased echogenicity. B) Postnatal USG of the neonate showing bulky right kidney with a heteroechoic area at the upper half of the right kidney with vascularity on color Doppler.

MRI abdomen also corroborated the findings of USG and showed the upper half of the right kidney replaced by an ill-defined heterogeneously T2 hyperintense mass lesion, measuring 40x43X45mm, with multiple small cystic areas within the lesion. The lesion was abutting the superior surface of the liver, without any obvious infiltration. Anteriorly, it was abutting the large bowel (Fig. 2). All hematological investigations were within normal limits, viz. Hemoglobin 9.3gm/dl, Total Leukocyte count 11,200/cu mm, Platelet counts 2 lacs/cu mm, Prothrombin time 11.3sec, INR 0.9, and Urine examination was also normal. A detailed checkup to rule out other congenital anomalies like aniridia, macroglossia, mental retardation, hamartomas, and neurofibromatosis were done in a multidisciplinary manner. After a detailed discussion with the tumor board, the treatment protocol was formulated as per the COG/NWTSG guidelines. Hence, the patient was taken up for a right nephroureterectomy. Per-operatively, the right kidney was grossly enlarged (10cmX4cmX5cm) with a firm, fleshy tumor present in and replacing the superior three-fourths of the kidney. The tumor was abutting the inferior surface of the liver superiorly, inferior vena cava medially, and right lateral abdominal wall, laterally. The right adrenal gland could be delineated separately. The right renal vein was engorged, but without any evidence of any thrombus (Fig. 3A). On the cut section, the tumor looked fleshy, with one large and two small cystic areas (Fig. 3B)

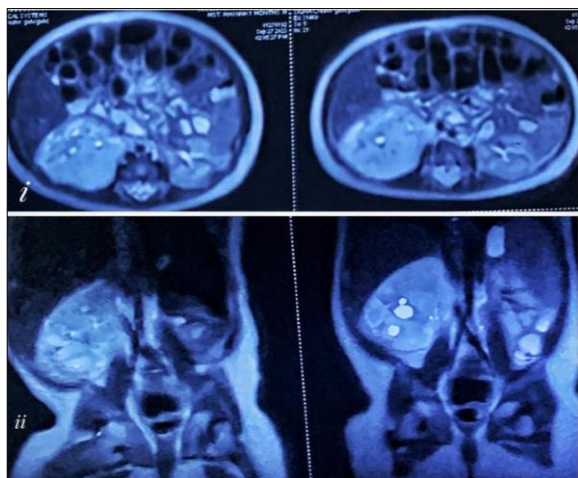


Fig. 2: MRI images showing the upper half of the right kidney were replaced by an ill-defined heterogeneously T2 hyperintense mass lesion in (i)coronal and (ii) cross-sectional sections.

Histopathology confirmed it to be a non-anaplastic tumor with a blastemal component of more than 60%, with no positive lymph nodes. The tumor came out to be WT1 and Vimentin positive and focally positive for Cyclin D1, whereas negative for Desmin, SMA, and PAX8, thus confirming the histopathological diagnosis of WT, stage 1, low risk (with intact Gerota's fascia) (Fig. 4A & 4B).

The postoperative period was uneventful, with the patient discharged on the fourth postoperative day, in fair health.

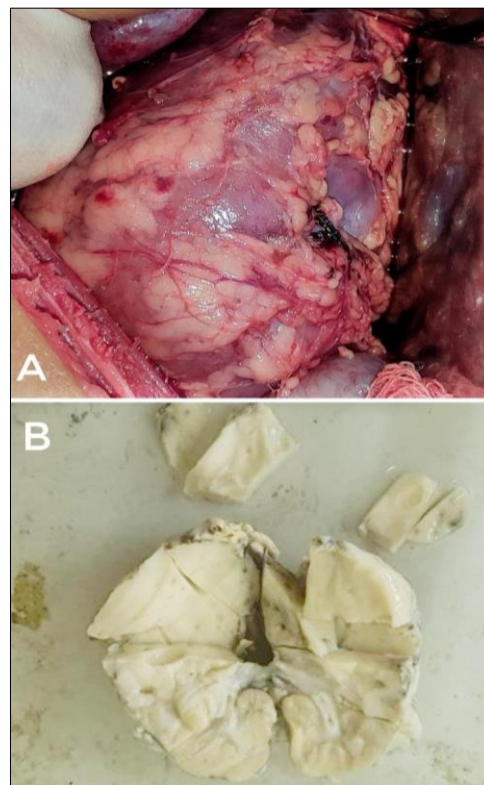


Fig. 3: A) Per operative image showing a grossly enlarged right kidney with tumor and enlarged right renal vein. B) Cut-section of the right kidney showing a fleshy tumor with few cystic areas.

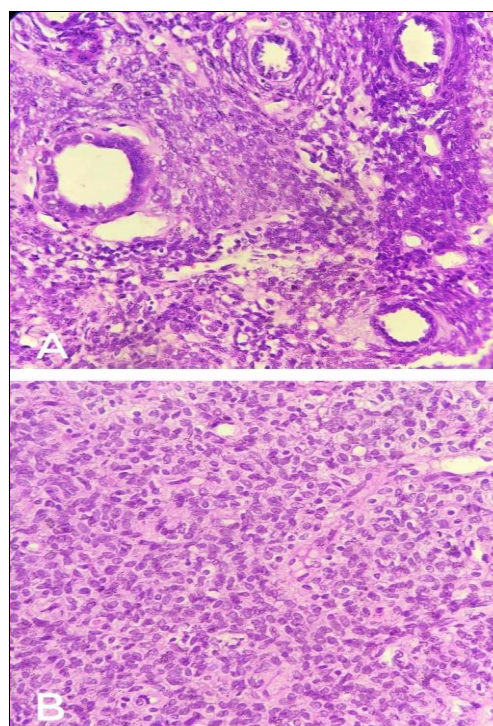


Fig. 4: A) Microsections examined reveal classic triphasic Wilms tumor composed of blastemal, epithelial, and stromal components. low power (H&E 100x). B) On H and E, a predominance of stromal cells. There is no evidence of anaplasia. (400x)

The patient is in regular 3 monthly follow-ups, without any new complaints and no obvious sign of tumor recurrence clinically or radiologically (USG abdomen, Chest X-ray, and hematological investigations).

DISCUSSION

Though WT is the most common renal malignancy in children, accounting for 85% of cases [3], antenatal diagnosis and even presentation in infancy are extremely rare. The most commonly detected tumor antenatally has been Congenital mesoblastic nephroma (CMN) while others like WT, neuroblastoma, and teratomas are rare. The detection of these tumors with antenatal MRI and ultrasound has been tried previously in various studies, but differentiation between CMN and WT has been difficult [3]. Till now, only ten cases of antenatally diagnosed WT have been reported in the literature [1-9].

This tumor is mostly seen in children between the ages of 1 to 5 years, with a peak at 3 years of age, and extremely rare in people over 15 years of age [10]. Gender disparity as such is not seen, but the female-to-male ratio is higher in Asian countries (4:1) [11]. It is mostly unilateral, but bilateral in 4-8% of cases [9]. WT is most commonly associated with WT1, WT2, CTNNB1 (Beta-Catenin), GPC3, IGF2/H19 genes [12-15].

Patients present most commonly with an abdominal mass, followed by hematuria and hypertension, and also, malaise, fever, weight loss, anorexia, left-sided varicocele, and hypercalcemia [16,17]. Ultrasonography (USG) is the first line of investigation, but all pediatric patients with renal masses should undergo computed tomography (CT) or magnetic resonance imaging (MRI) for additional information, like metastasis, resectability and contemplating the line of treatment [18].

An antenatally diagnosed renal tumor can be associated with polyhydramnios, hydrops fetalis, and acute fetal distress [2]; polyhydramnios being the commonest and hydrops rare; though our patient had none of these associations.

Treatment consists of surgery, chemotherapy, and radiotherapy, depending on the stage of the tumor.

We followed COG/NWTSG guidelines and performed the right nephroureterectomy. Chemotherapy and radiotherapy were not contemplated as the tumor was unifocal with intact Gerota's fascia and adrenal gland and renal vessels were free of tumor infiltration.

If a tumor is detected antenatally, we keep the patient on our institution's monitoring list, taking a multi-departmental approach, keeping the patient under the direct supervision of the obstetrics and gynecology department with serial ultrasonography for diagnosis confirmation and advice for institutional delivery, and revert to us as soon as the baby is delivered for further investigations and treatment planning.

The prognosis of these tumors is still a matter of debate, although poor prognosis has been mostly seen in antenatally detected cases. Shamberger RC et al reported that neonates with WT, who have tumor weight less than 550g are considered to have a good prognosis and may not require chemotherapy [19]. However, in a study by Jain V et al, the prognosis was not that satisfactory, with a mortality of 60%, causes described to be hydrops and recurrence, and also delay in presentation and lack of radiotherapy services in the hospital [20], while in our case, the patient was operated successfully and discharged after an uneventful postoperative period, with reasonably good health at the fourth month of follow-up.

In conclusion, Wilms tumor has a favorable outcome, but it is often missed in the antenatal period. A high degree of suspicion and prompt investigative workup should be carried out both antenatally and in the immediate neonatal period, for optimal management and favorable outcome.

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