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## Case Report

# Wilms tumor presenting as small bowel obstruction in a neonate: A diagnostic challenge ☆☆☆

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## ABSTRACT

Wilms tumor is the most common primary malignant renal tumor of childhood which usually presents between 2 and 6 years of age. Its presentation in the neonatal period is extremely rare and presenting with intestinal obstruction is perhaps unknown. We report a 2-day-old baby girl who manifested features of acute upper gastrointestinal obstruction with frequent post-feeding vomiting and abdominal distension. The initial abdominal radiograph showed abnormally displayed small bowel loops to the right hemiabdomen. Subsequent ultrasound and computed tomography scan of the abdomen detected a massive left renal mass. Left-sided nephrectomy was performed, and histopathology demonstrated left-sided Wilms tumor with favorable histology. Post-treatment yearly follow-up for 5 years recorded a disease-free, normally thriving child.

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## Background

Wilms tumor is the most common primary renal tumor in children [1]. After neuroblastoma, it is the second most common pediatric intra-abdominal tumor. Wilms tumor typically presents in children below age six years and the mean age at diagnosis is 4 years [2]. Only a few of all reported cases of pediatric Wilms tumor presented during neonatal period and, so far we explored, none presented with intestinal obstruction. Here is how our reported case is interesting and challenging as it presented in early neonatal life with intestinal obstruction. We share our experiences of overcoming the radiological dilemmas and diagnostic challenges we faced to detect a case of Wilms tumor presenting in unusual age with unknown features. Additionally, most neonatal intra-abdominal mass/tumor are usually detected during antenatal ultrasound, but our case skipped detection during antenatal visit and put extra diagnostic confusion on us [3,4].

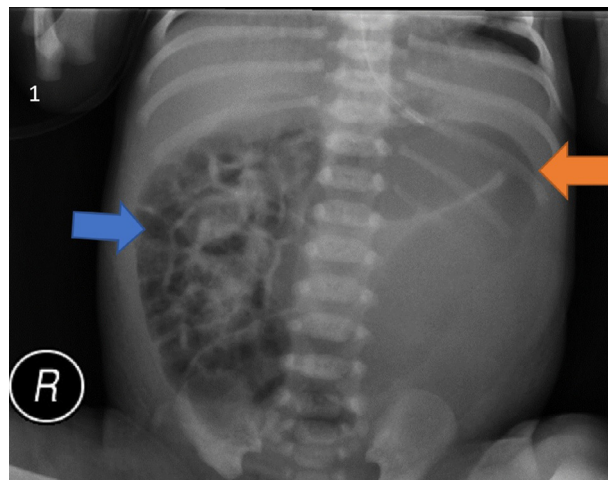
## Case report

A 26-year-old gravida 2 para 1(G2P1) woman with insulin-treated gestational diabetes mellitus vaginally delivered a 39-week-old girl weighing 3.1 kg. Her Apgar score was 9 and 10 at first and fifth minutes, respectively. Prenatal sonographic assessment throughout pregnancy was normal. Her first child, a boy, was in good health with no known medical illnesses. There was no family history of any kind of malignant disease.

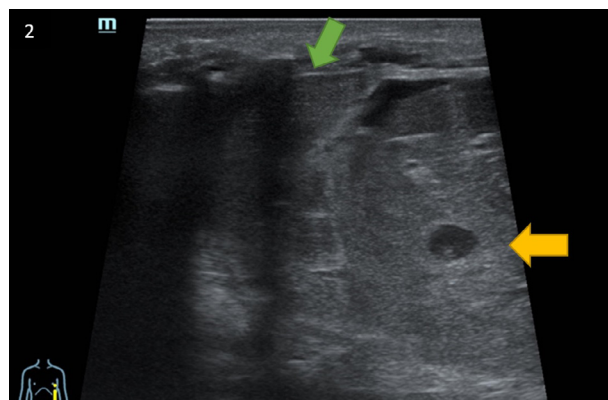
On the second day of life, the new-born girl developed gradual abdominal distension and recurrent vomiting of meconium stain fluid. Although she passed meconium before the development of the symptoms, it was scanty. Physical examination revealed a hemodynamically stable, non-syndromic baby girl with significant abdominal distension. There was no associated ano-genito-urinary abnormality, hemihypertrophy, aniridia, or spinal deformity noted in the patient. Abdominal auscultation detected increased frequency and pitch of bowel sounds. A multidisciplinary team consisting of a neonatologist, a pediatric surgeon, and a pediatric radiologist was formed for diagnosis and subsequent management of the child.

An abdominal plain radiograph showed small bowels displaced to right hemiabdomen with gas-filled transverse and descending colon in the left abdomen. No pneumatosis intestinalis or pneumoperitoneum suggestive of necrotising enterocolitis were detected (Fig. 1).

Ultrasound of the abdomen revealed a heterogenous solid-cystic retroperitoneal mass displacing the spleen superiorly. No suprarenal mass was detected (Fig. 2). Subsequent contrast-enhanced abdominal and pelvic computed tomography (CT) scan detected a large heterogeneously enhancing mass infiltrating the lower pole of the left kidney without any internal calcification. The renal parenchyma in the non-infiltrated upper pole appeared normal, however, was complicated with focal caliectasis (Fig. 3A and 3B). The renal mass pushed the small bowel loops to the right hemiabdomen and compressed them producing features of intestinal obstruction.



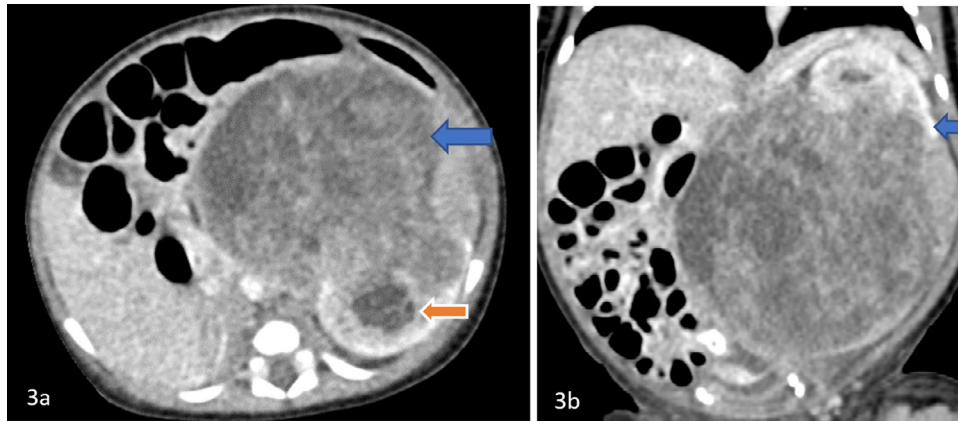
**Fig. 1 – Abdominal radiograph showed abnormal displacement of small bowel loops towards the right (blue arrow) with gas-filled transverse and descending colon seen only at the left side of the abdomen (orange arrow). (Color version of figure is available online.)**



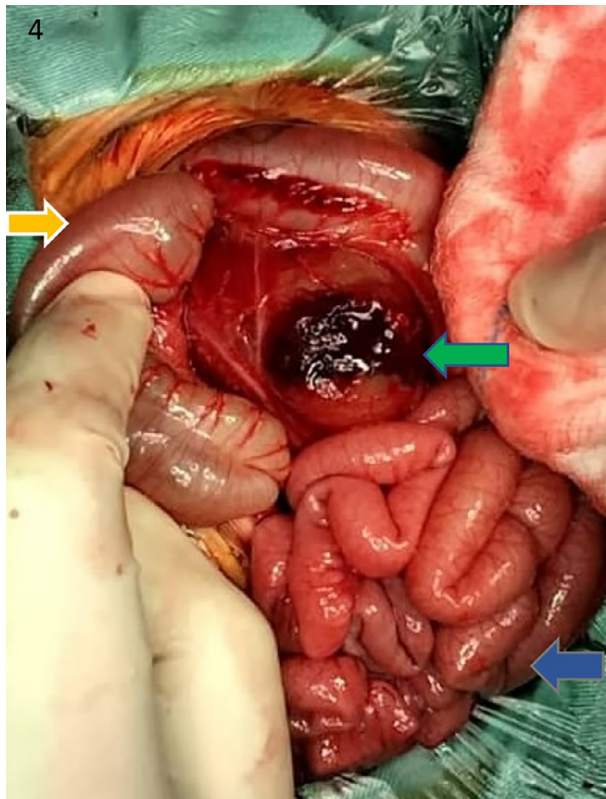
**Fig. 2 – Ultrasound abdomen showed a huge retroperitoneal mass (yellow arrow) displacing the spleen (green arrow) superiorly. (Color version of figure is available online.)**

The small bowel loops were fluid-filled but not significantly dilated. No evidence of bowel malrotation or volvulus was detected (Fig. 4). Owing to the obstructive nature of the left renal mass, the team decided to go for exploratory laparotomy and left nephrectomy after obtaining parents' consent. Intra-operative findings demonstrated a left retroperitoneal mass attached to inferior pole of the left kidney.

After surgical removal, the mass was sent for histopathological evaluation. On gross examination, it was a pinkish, fleshy, cystic, solid mass measuring 7cm x 8cm. Histology demonstrated a well-circumscribed tumor with the presence of primitive columnar cells with areas of necrosis. Some areas demonstrated blastemal elements, however, no anaplasia was noted. The left renal capsule was found infiltrated with tumor cells but the resected end of the renal vessels and the adja-



**Fig. 3 – (A)** Post-contrast CT abdomen in axial view showed large heterogeneously enhancing mass arising from the lower pole of the left kidney (blue arrow) with associated focal caliectasis of the left kidney (orange arrow). **(B)** Post-contrast CT abdomen and pelvis in coronal view showing normal upper pole renal parenchyma producing a claw sign (blue arrow) with the underlying mass inferiorly. (Color version of figure is available online.)



**Fig. 4 – Intraoperative image** showing cystic-solid retroperitoneal mass (green arrow) causing displacement of fluid-filled small bowel (blue arrow) and transverse colon (yellow arrow) intra-abdominally. (Color version of figure is available online.)

cent lymph nodes were free. Immunocytochemistry showed strong WT1 positivity in both blastemal and epithelial components suggesting a Wilms tumor with favorable histology. Meanwhile, the parents and brother of this patient were ge-

netically tested for Wilms tumor gene and were found negative. Postoperatively, the patient was followed up yearly for five years. In every follow-up visit, we detected complete resolution of the tumor with no evidence of disease recurrence.

### Discussion and literature review

Wilms tumor, also called nephroblastoma, is the commonest primary malignant renal tumor in children accounting for 6% of overall childhood malignancies and more than 90% of pediatric renal tumors [5-8]. Most (95%) cases of Wilms tumors are sporadic and few cases may occur in association with other congenital malformations such as aniridia, hemihypertrophy, cryptorchidism, hypospadias, gonadal dysgenesis, pseudohermaphroditism, and horseshoe kidney, or genetic syndromes such as Beckwith-Wiedemann syndrome, Denys-Drash syndrome, and WAGR syndrome (Wilms tumor, aniridia, ambiguous genitalia, mental retardation) [9-12]. In terms of genetic basis, certain genes, notably WT1 or WT2 genes on chromosome 11, WTX gene on X chromosome, and CTNNB1 gene on chromosome 3 have been identified by DNA sequencing of the tumor genomes in familial cases of Wilms tumor. Even sporadic cases of Wilms tumor are thought to have emerged from genetic mutations [13-16]. Approximately 2% cases of Wilms tumor occur in families. The familial cases are generally bilateral and develop at an earlier age [17]. Nephrogenic rests, which represent abnormal persistence of residual embryonic metanephric tissue in the kidney beyond 36 weeks' gestational age, are best known as precursors of Wilms tumor. Nephrogenic rests may be observed in approximately 1% of normal children that eventually regress in most of the cases. If not, it can undergo malignant transformation and give rise to Wilms tumor. Nephrogenic rests are encountered in up to 40% of unilateral and over 90% cases of bilateral Wilms tumor [18-22].

The mean age at diagnosis of Wilms tumor is 4 years with most cases appearing between ages 2 and 5 years. Silent ab-



dominal mass, typically brought into medical attention by the caregiver while bathing the child or changing the clothes, is the commonest mode of presentation [23–26]. Overall, neonatal tumors are most often benign and malignant neonatal tumors are rare representing only 2% of all malignancies in childhood [27]. In one report on neonatal tumors recorded over a period of five years, 43 out of 51 cases (84.3%) (15.6%) had benign and 8 (15.6%) had malignant tumors, of which only one patient had Wilms tumor [28]. A report on neonatal abdominal mass requiring surgical intervention also revealed majority (87%) cases were benign in nature [29]. Wilms tumor presenting in neonatal period is extremely rare. A report on 3,340 cases of Wilms tumor registered over a period of 15 years (from 1969 through April 1984) identified only 27 (0.8%) cases presenting in neonatal period [30].

Neonatal Wilms tumor presenting with intestinal obstruction is perhaps the rarest event since, as far as our efforts went, we failed to find it documented in the published literature. A report on 15 cases of neonatal Wilms tumor showed 12 patients (80%) presented with asymptomatic abdominal mass detected during routine neonatal examination and three (20%) were identified during antenatal ultrasound [31]. Published literature are, however, available on the development of post-nephrectomy intestinal obstruction/intussusception among the Wilms tumor patients [32–34]. Wilms tumor presenting with features of urinary tract obstruction is also reported in the literature [35].

Common differential diagnoses of neonatal retroperitoneal mass include polycystic kidney disease, Wilms tumor, neuroblastoma and mesoblastic nephroma, and rhabdoid tumor of which neuroblastoma should get priority consideration because of its near-similar location and age group of presentation like that of Wilms tumor [36–37].

Clinically, Wilms tumor does not cross the midline, but neuroblastoma does. However, from the radiological perspective, the most classic sign to differentiate Wilms tumor from neuroblastoma is claw sign which refers to sharp demarcation between the tumor and the normal renal parenchyma producing a sharp angle on both sides of the mass, mimicking the shape of a crab or lobster's claw.

Claw sign develops when a lesion arises in a solid organ and expands outwards thinning the parenchyma between it and the surface. The claw sign helps to determine the origin of a mass from a solid organ, such as kidney [38–40]. Other distinguishing features include the presence of calcification which is common in neuroblastoma but uncommon in Wilms tumor [41].

## Conclusion and practice point

Presentation of Wilms tumor in neonatal period is rare and its presentation with small bowel obstruction on second day of life is perhaps unknown in the published literatures. Our case was unique to be the first of this kind. It helped to give a reminder for the neonatologists, pediatricians, pediatric surgeons, and pediatric radiologists that Wilms tumor should be considered in the differential diagnosis of neonatal intestinal obstruction where other common causes are not evident.

## Patient consent

Written informed consent was obtained from the patient's parents for the publication of this case report.

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