

## Case Report

# Atypical presentation of Wilms tumor, an 11-year-old girl with abdominal pain and anemia: a case report

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**Abstract:** Wilms tumor is the most common renal tumor in children. The tumors often develop quite large before being noticed and most of them are unilateral. Ultrasound (US) scan, computed tomography (CT), and magnetic resonance imaging (MRI) are typically applied for identifying tumor location and size. Surgery is the first choice for treatment, and biopsy is also taken to confirm the diagnosis. Here, a case of Wilms tumor is presented in which the initial diagnosis was for abdominal pain with anemia but the patient was then treated with radical nephrectomy and chemotherapy. As a pediatric physician, more attention should be paid to abdominal pain, especially in children with anemia since it might be the sign of Wilms tumor.

**Keywords:** Abdominal mass, Wilms tumor, children

### Introduction

Wilms tumor is the most common renal tumor in children, affecting approximately 650 children each year in the United States [1]. In Taiwan, approximately 500 cases of children under 15 years had Wilms tumor each year [2]. Approximately 4~7% of Wilms tumor patients have bilateral kidney involvement and earlier ages of onset than unilateral Wilms tumors [3]. The mean age at diagnosis is 3 years old in which 80% of individuals are diagnosed before the age of 5 years [4, 5]. The tumors often develop quite large before being noticed. The symptoms of Wilms tumor include hematuria (20%), coagulopathy (10%), and hypertension owing to the activation of the renin-angiotensin system (20~25%). About 10% present as fever, anorexia, and weight loss. In rare cases, tumor rupture and bleeding can cause acute abdomen [6]. Ultrasound (US) scan, computed tomography (CT) and magnetic resonance imaging (MRI) are routinely used at diagnosis to identify tumor locations and sizes [7, 8].

Treatment of Wilms tumor is determined both by stage and histological classification (favorable or anaplastic). Surgery is the first choice for treatment, and biopsy is also taken to confirm the diagnosis. Furthermore, treatment is mostly combined with chemotherapy [9]. Herein, a case is presented of an 11-year-old girl initially diagnosed as abdominal pain with anemia but finally diagnosed as Wilms tumor and treated with radical nephrectomy and adjuvant chemotherapy.

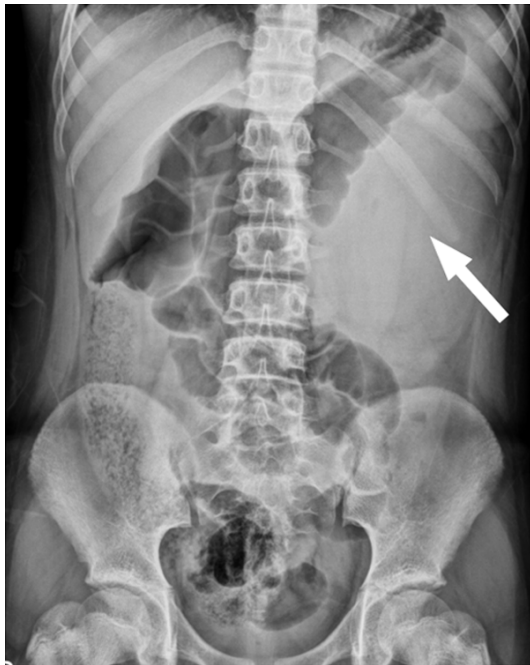
### Case Report

An 11-year-old girl suffered from intermittent abdominal pain and postprandial vomiting from May 5, 2017. Decreased appetite and activity were also noticed. As her abdominal pain persisted, she was brought to our outpatient department on May 7, 2017. There was no fever or diarrhea prior to admission. At outpatient department, the laboratory data revealed leukocytosis (WBC: 24540/ $\mu$ l) and elevated C-Reactive Protein (CRP) level (4.204 mg/dl)

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**Table 1.** Laboratory data of the patient for first four days after hospital admission

Reference range	WBC ( $\mu$ L)	Hemoglobin (g/dl)	Platelet ( $\mu$ L)	CRP (mg/dl)	PT (Secs)	APTT (Secs)	INR
	4500-10,000	12.0-16.0	130,000-40,000	< 0.3	8.0-12.0	24.3-32.7	< 1.5
Examination date							
2017.05.07	24540	11.2	321,000	4.204			
2017.05.08	21270	9.5	264,000	-	12.7	42.6	1.25
2017.05.10	14780	10.1	262,000	19.747			



**Figure 1.** Abdomen X-ray shows a mass lesion with the distribution of bowel loops over left side (arrow).

(Table 1). Tenderness was noticed over left upper quadrant (LUQ) and the periumbilical area. With the additional signs of vomiting and abdominal pain, acute gastritis was suspected and she was admitted to the ward for further evaluation and management.

After admission, physical examination showed mild pale conjunctiva, soft abdomen with diffuse tenderness over the lower left area. Mild tachycardia up to 120/min but relatively stable blood pressure (130/86 mmHg) was also noted. Abdominal X-ray (KUB) showed some gas and stool over the colon and a mass over the left side (Figure 1). US scan revealed a mass on the left kidney of approximately 10.5 × 13.1 cm in size and suspected rupture (Figure 2). Also, abdominal CT scan revealed a big heterogeneous mass (~10 × 12 cm) on the left

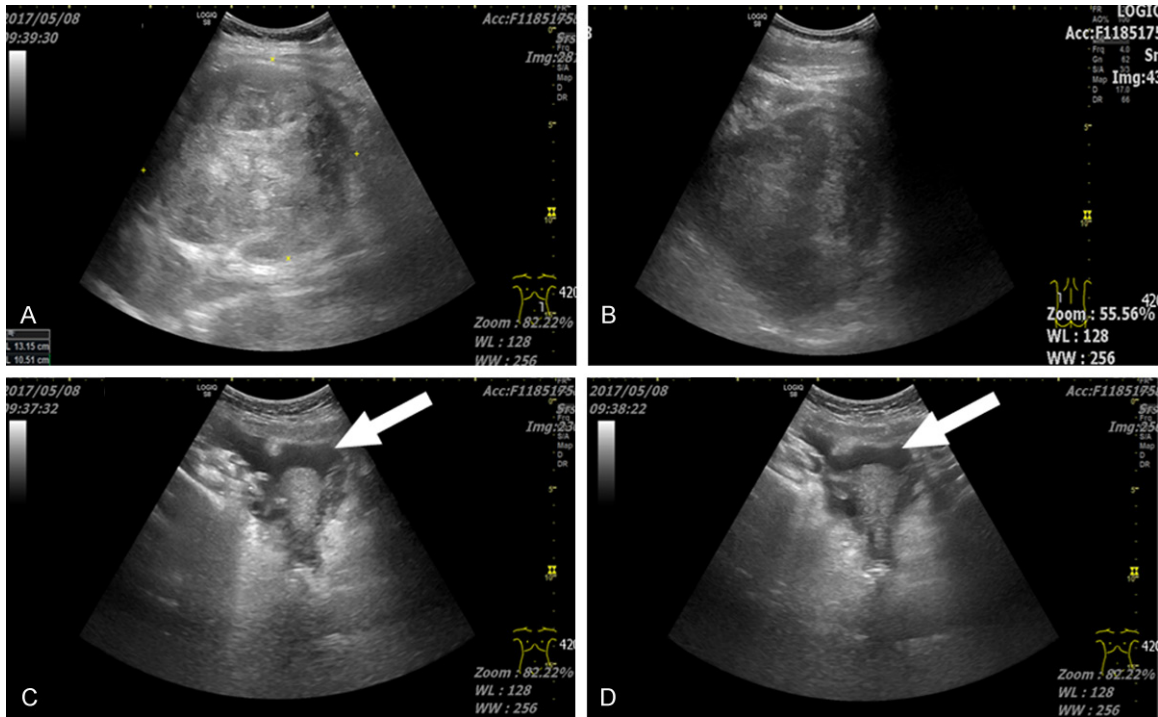
kidney with surrounding retroperitoneal fluid and mild ascites at the lower abdominal cavity. Left renal tumor bleeding with big retroperitoneal hematoma ruptured into lower peritoneal cavity was highly suspected (Figure 3). Due to unstable vital signs and active internal bleeding, the patient was transferred to the pediatric intensive care unit (PICU) for close observation.

After transferring to the PICU on May 8, 2017, packed red blood cell (PRBC) transfusion was provided due to intra-abdominal blood loss (Hemoglobin: 11.2 → 9.5 g/dl). Fresh frozen plasma (FFP) was also provided because of prolonged prothrombin time/partial thromboplastin time (PT/APTT) (Table 1). The patient was then transferred to the medical center-level hospital for surgery intervention on May 11, 2017. At the medical center-level hospital, left side radical nephrectomy was performed, and the pathological report showed it was Wilms tumor with favorable histology. The tumor was confined to the renal parenchyma, whereas the renal pelvis, hilar vessels, adrenal gland, and peri-renal soft tissues were free of tumor. Adjuvant chemotherapy was also arranged after surgical intervention. Currently, the patient is still receiving chemotherapy treatment and has been under the regular follow-up at the medical center-level hospital.

### Discussion

Wilms tumor is the most common renal tumor in children, approximately 3% in all childhood cancer [5]. The pathogenesis of Wilms tumor may be caused by abnormal renal development. It has been reported that congenital anomalies and constitutional chromosomal abnormalities are associated with Wilms tumor. Approximately 9% of individuals with Wilms tumor have a congenital anomaly shown by the data from the British National Registry of

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**Figure 2.** US scan revealed a left kidney mass of  $\sim 10.5 \times 13.1$  cm (A) and suspected kidney or mass rupture (B). (C and D) Some ascites over lower abdomen were also detected (arrows).

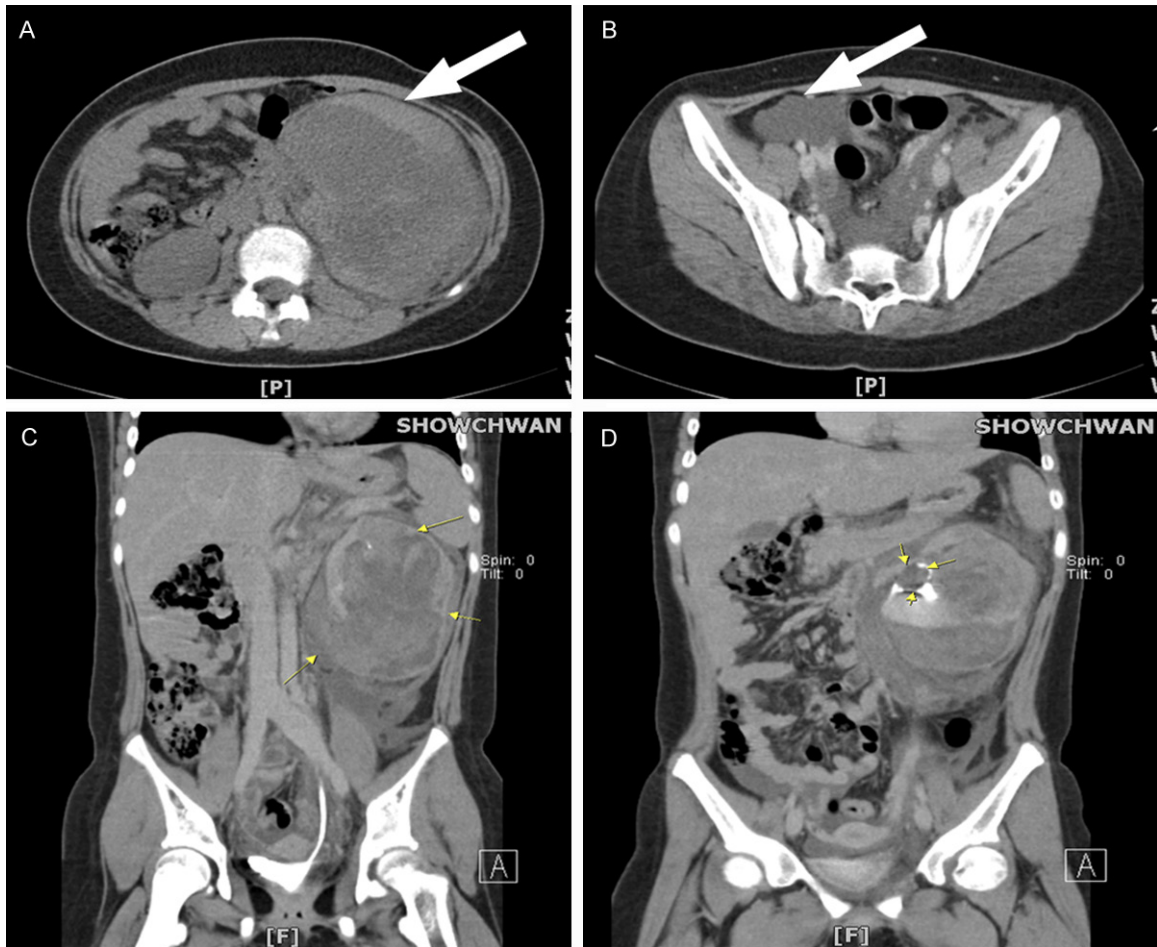
Childhood Tumours [9]. Previous studies showed Wilms tumor was associated with the mutations in the Wilms tumor 1 (*WT1*), *TP53*, *FWT1*, and *FWT2* genes [9, 10]. The *WT1* gene is located at 11p13 and encodes a zinc finger transcription factor playing a crucial role in renal and gonadal development [11]. *WT1* is a classic tumor suppressor gene and the wild type allele is somatically inactivated in tumors with constitutional *WT1* mutations or deletions [9].

Wilms tumor also has the great chance of combining with other syndromes, such as WAGR (Wilms-aniridia-genitourinary-mental retardation) syndrome, Denys-Drash syndrome, both with the *WT1* mutation, Beckwith-Wiedemann syndrome associated with genetic and epigenetic abnormalities at 11p15, and other congenital anomalies [9, 12]. Thus, lung CT scan, genitourinary US scan, and gene analysis should be performed.

The symptoms of Wilms tumor may vary wildly and some cases may not have any obvious signs. However, in some children, abdominal pain and a palpable mass may be observed [13]. Asymptomatic situations are challenging

for correct diagnosis. In this study, the patient was initially diagnosed with abdominal pain and mass and after careful examination by a number of imaging techniques, i.e. KUB, US and CT, she was finally diagnosed as having Wilms tumor. In general, abdominal imaging techniques including US scan, CT and MRI can be applied for the evaluation of abdominal mass, a most common clinical presentation of a pediatric renal malignancy [14]. Histology can differentiate Wilms tumor from other types of cancers, such as clear cell sarcoma, rhabdoid tumor, congenital mesoblastic nephroma, renal cell carcinoma, and renal medullary carcinoma. Additionally, histology can classify different cell types [13].

Spontaneous rupture of Wilms tumor is uncommon [15]. In a recent study, a total of 187 patients with Wilms tumor were examined, but only 2.1% showed preoperative ruptures [16]. In this case, tumor rupture was noted before operation. The International Society of Pediatric Oncology (SIOP) has suggested that any tumor with rupture should be regarded as stage III, because tumor rupture has a significant impact on prognosis due to tumor dissemination. If tumor spillage occurs, it will increase recurrence



**Figure 3.** CT scan images. A. Retroperitoneal hematoma ruptured into lower peritoneal cavity (arrow). B. Retroperitoneal fluid and mild ascites at lower abdominal cavity (arrow). C. A big heterogeneous mass (~10 × 12 cm) at left kidney (arrows). D. ~2.0 cm filling defect was in left ureteropelvic junction, neoplastic invasion should be considered (arrows).

rate up to 20% [16, 17]. However, it is difficult to detect preoperative Wilms tumor rupture by the radiological technique, but CT detection of preoperative Wilms tumor rupture may aid the surgeon in preoperative planning before laparotomy. Furthermore, our patient showed the symptoms of abdominal pain and anemia. It has been reported that clinical symptoms of abdominal pain, anemia, and shock could aid in the diagnosis of preoperative tumor rupture [18].

Treatments for Wilms tumor usually start with surgery to remove tumor and kidney. Adjuvant chemotherapy and radiation therapy will then be performed according to the stage of the cancer [6]. Taiwan Pediatric Oncology Group (TPOG) also developed the protocol for treating Wilms tumor [2]. Treatment for Wilms tumor has been successful in pediatric oncology, with the long-

term survival being > 90%, when disease was localized to the abdomen and > 70% with metastatic disease [19].

In conclusion, the case of a child with vomiting and abdominal pain who was finally diagnosed as Wilms tumor is presented here. When approaching patients, age is considered very important, because older children are with higher risk of misleading diagnosis and developing to malignancy. Asymptomatic situations are also a challenge for determining the correct diagnosis. As a pediatric physician, more attention should be paid to abdominal pain, especially in children with anemia, because it might be the sign of Wilms tumor.

#### Disclosure of conflict of interest

None.



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