

Characteristics of Clinical Finding in Child Diagnosed with Wilm's Tumor: A Case Series

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Abstract. Wilm's Tumor is the second most common intra-abdominal cancer of childhood, the fifth most common pediatric malignancy overall, and the most common renal tumor in children, representing 95% of all pediatric renal malignancies. However, Most of them present as an asymptomatic abdominal mass. Here we present two cases of Wilm's tumor with different characteristics of clinical symptoms and ages. In Conclusion, Wilms tumor has a variety of clinical characteristics in each patient. The disease is also frequently found asymptotically which leads to a delay in diagnosis. Screening for Wilms' tumor is very important for children who have syndromes or birth defects known to be linked to this disease.

Keywords: epidermoid cyst, penis

Introduction

Wilms' tumor is the most common renal tumor in children which represents 95% of all pediatric renal malignancies [2]. Wilm's Tumor is also the second most common intra abdominal cancer of childhood and the fifth most common pediatric malignancy in the overall [1]. Wilm's tumor is usually presented as an asymptomatic abdominal mass. The characteristics of the clinical finding of Wilm's Tumor in children also varies, including abdominal pain, gross hematuria, fever, Urinary Tract Infection, and hypertension [3]. Therefore, we are presenting a case series of two children diagnosed with Wilm's Tumor with different clinical features.

Case Report (1)

A three-year-old young girl was referred to the Siloam Hospital Makassar with a suspected right abdominal tumor. The patient complained of prolonged malaise, right-sided abdominal pain since one month ago, and intermittent hematuria. At first, there was only a small lump in the right abdomen and then enlarged, followed by other symptoms that have been mentioned, the patient was born full-term following an uncomplicated pregnancy presented.

On physical examination, the patient was found to have a high-grade fever (38.9c) with blood pressure 108/60 mmHg. there is a right abdominal mass measuring 10-12 cm with a freely moveable and bumpy edge, the mass did extend the midline. there is no obvious meaningful examination found.

The hematologic assessment showed that leukocytosis (leukocyte count was 21.4 G/L), differentiation white blood cell count showed 92.2% neutrophil, and The erythrocyte sedimentation rate (ESR) was moderately elevated due to infection. Biochemical assessment based on blood ureum-creatinine & electrolytes seemed to be normal (blood ureum 23 MG/DL, Sodium 140 mmol/L, blood creatinine 0.39 MG/DL). Urinalysis showed microhematuria (Erythrocytes 30/HPF, Leukocytes Negative, Glucose negative).

Abdominal contrast computed tomography (CT) revealed a heterogeneous mass with areas, well defined, +/- 12 cm x 11.4 cm x 13.1 cm originating from the lower middle pole of the right kidney reaches the renal pelvis, crosses the midline, pushes the intestinal loops anteromedially (Fig. 1, Fig. 2).

Case Report (2)

Twelve years old girl referred to the Siloam Hospital Makassar presented with a persistent

painless lump on the right abdomen 2 months ago, the lump was gradually progressive and associated with weight loss and hypertension since last month. There was no history of abdominal pain, vomiting, and hematuria. The patient also complained of feeling tired easily while doing moderate to high-intensity activity. Birth history reveals uncomplicated with full-term vaginal delivery, her development was normal, and her health had been good since the lumps had been noticed.

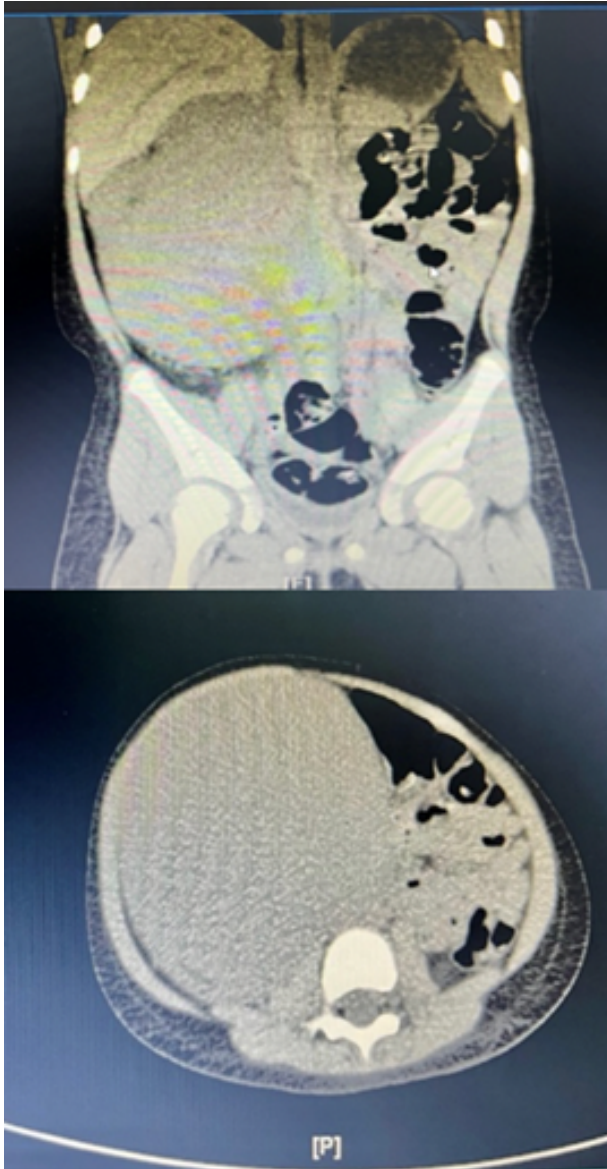


Figure 1. Abdominal CT-Scan; CT-scan showed a bulging of the flank more on the right side and loss of the psoas muscle outline. Bowel loops are displaced anteromedially, causing no surrounding vascular displaced.

On the physical examination, The temperature was 37.3C, and the blood pressure was 135/90, and respiration was 25 breaths per minute. On

abdominal examination, there is a mass of 10 x 7 cm with a regular edge, and no tenderness pain. all other examinations are normal.

Non-Contrast Abdominal CT-Scan Showed an isodense mass with calcification measuring 10,5 cm x 8,6 cm x 8,9 cm on the lower kidney pole that pushes the kidney to posteromedially, which concluded Wilms tumor on the right kidney that causes moderate hydronephrosis (Fig. 3, Fig. 4).

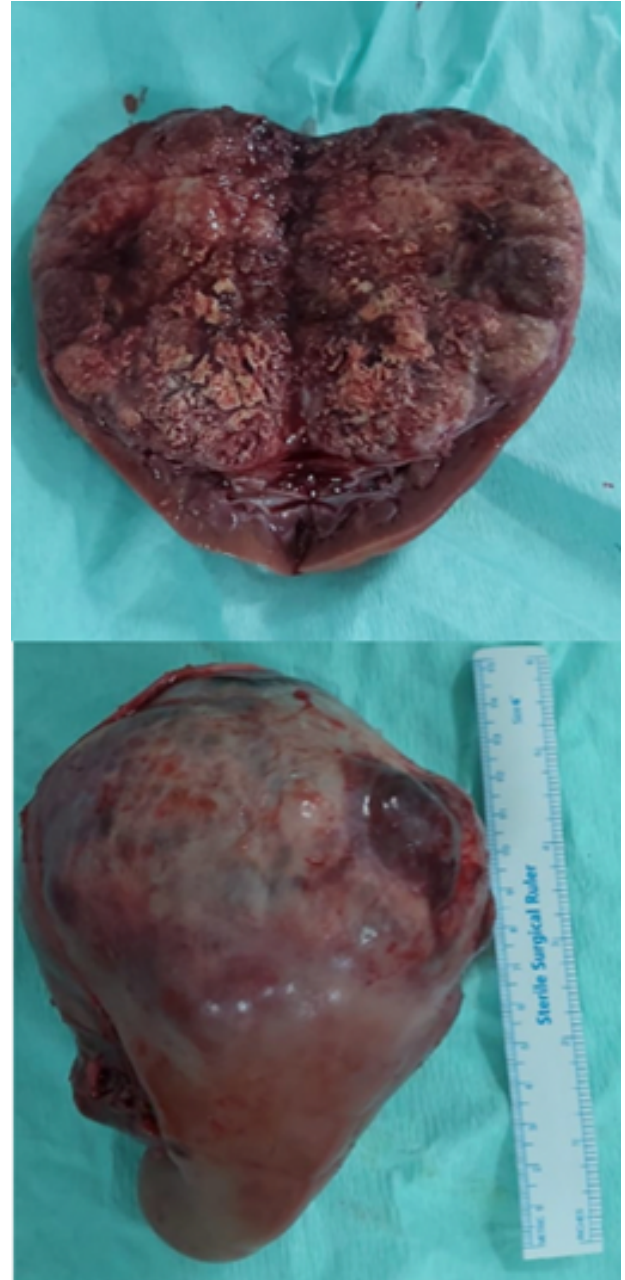


Figure 2. Macroscopic picture of the kidney

Laboratory data revealed leukocytosis (leukocyte count was 11.9 UL), differentiation white blood cell count showed 86.5%, and lymphocyte count was slightly low at 5.3 %. All other serological and biochemical parameters were normal.

Radical nephrectomy was performed on the patient without any course of chemotherapy. There had been no prior chemotherapy biopsy or diagnosis. The operation took about 2 hours and a half. The tumor infiltrated the muscle of the posterior abdominal wall. The specimen that was received in the surgical pathology department was the right kidney, measuring 14x10x7 cm with a soft texture. Microscopic consists of the proliferation of epithelial cells in which the core is rounded-void, atypical, with eosinophilic cytoplasm.



Figure 3. Abdominal CT-Scan non Contrast suggestive of right kidney tumor

There were also hypocellular connective tissue septa consisting of an undifferentiated stromal element and necrosis areas between tumor cells. The tumor consisted only of stromal epithelial components which concluded Nephroblastoma (Wilm's tumor) with low risk

There was no postoperative adjuvant chemotherapy and radiotherapy was given. The patient has been well without recurrence or complication after 6 months of surgery.

Discussion

Wilm's Tumor, known as nephroblastoma, is the most common malignant pediatric renal tumor, responsible for 95% of all malignant kidney tumors in patients under the age of 15 years old diagnosed [4] and Approximately 75% of the cases occur in

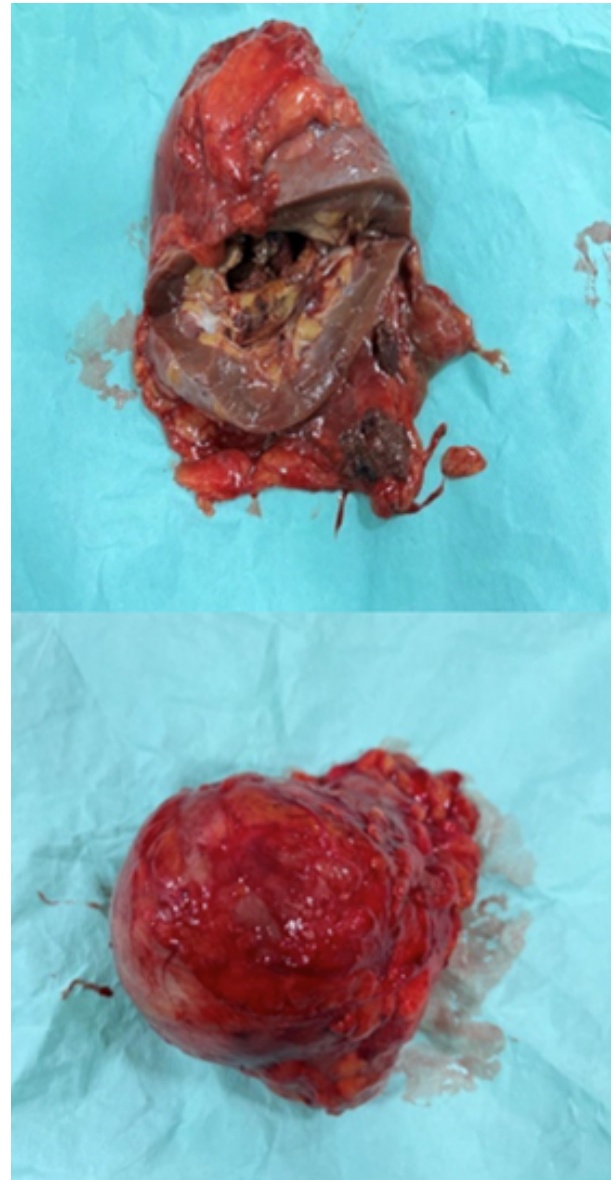


Figure 4. Macroscopic picture of the kidney

children less than 5 years of age with a peak incidence at 2 to 3 years of age. Wilm's Tumor is also the second most common intra abdominal cancer of childhood and the fifth most common pediatric malignancy overall [1]. Both of our patients were under the age of 15 years old. Our second patient was 12 years old, which is only 25 % percent of Wilm's tumor cases that occur over 5 years of age [2]. This concludes that the incidence of Wilm's tumor decreases with increasing age.

Wilms' tumor is highly associated with other congenital anomalies such as aniridia, hemihypertrophy, hypospadias, and kidney malformations such as fused, horseshoe, or polycystic kidneys in as many as 12% of patients [5]. In both of our patients, no other congenital abnormalities were found. Most Wilms tumor patients present asymptotically with an abdominal mass noticed by a parent or pediatrician

on a well-child visit. children with known predisposing clinical syndromes, Clinical findings may include a lump, swelling, abdominal pain (40% of cases); Hematuria (24% of cases), Hypertension (25% of cases), constitutional symptoms such as fever, anorexia, and weight loss (10% of cases) [6]. In our first patient, we found some common symptoms that are usually often found in Wilm's tumor, such as constitutional symptoms, hematuria, right abdominal pain, and a right abdominal mass, which is quite different from the second patient where only a painless abdominal mass associated with weight loss and increased of blood pressure.

Imaging plays a crucial role in the early diagnosis of Wilms' tumor to distinguish it from all other causes of abdominal distention [7]. Conventional ultrasound is the most common method for the initial diagnosis of Wilms' tumor due to its non-invasiveness, affordability, and availability. However, it is less accurate in tumor staging, which is required for the effective treatment of this malignancy [7].

On CT, Wilms' tumor usually appears as a bulky, spherical intrarenal mass, very often with a well-defined rim of compressed renal parenchyma or pseudocapsule surrounding it [7], which was also found in both of our patients (Fig. 3). Some tumors that arise from the periphery of the cortex may grow in an exophytic manner with the bulk of the tumor seen outside the renal cortex. In both of our patients the tumor was growing from the lower

middle pole of the right kidney (Fig. 3). In general, a heterogenous mass replacing the kidney and displacing adjacent organs can be demonstrated, The tumor is hypodense compared to the normal renal parenchyma on contrast-enhanced CT scans with the areas of low attenuation coinciding with tumor necrosis, fat deposition or both [7]. The images in both of our patients were an isodense mass which was not in accordance with the theory studied [7]. However, both of the patient's masses are heterogenous mass that pushes adjacent organs and resulting displacements (Fig. 3) of them. which is corroborated to the theory that has been studied.

To determine the treatment of Wilm's tumor, it depends on the stage of the tumor. There are two classifications used to determine the stage. COG (Children Oncology Group) approach is based on upfront nephrectomy followed by chemotherapy/radiation therapy. SIOP (Society of Pediatric Group) is based on preoperative chemotherapy to decrease tumor bulk [8]. In both of our patients, no preoperative chemotherapy was performed. Therefore, we used the COG approach to determine the staging of the tumor. Both of our patients showed the mass had reached the renal sinus, reached the renal pelvis, crossed the midline, and pushed the intestinal loops anteromedially, but the tumor seems didn't infiltrate the lymph nodes and nearby organs yet, the urologist determined both of the patients were in stages 2 according to the COG criteria (Table 1).

Table 1. Current staging criteria in COG and SIOP trials

Stage	COG (primary nephrectomy)	SIOP (after preoperative chemotherapy)
I	Tumour confined to kidney within intact renal capsule. No previous biopsy (COG). Vessels of sinus clear, hilar lymph nodes examined and negative	Tumour confined to kidney within intact (pseudo) capsule. Vessels of sinus clear. (Non-viable tumour in the sinus/perirenal fat ignored for staging purposes.
II	Tumour fully resected, but tumour extends through the capsule, or into soft tissue or renal sinus or in blood vessels beyond renal parenchyma	Tumour fully resected, but tumour extends through the (pseudo) capsule, or into soft tissue or renal sinus or in blood vessels beyond renal parenchyma
III	Residual disease in abdomen, due to rupture, spillage, tumour biopsy, tumour in abdominal lymph node, or tumour is fragmented and full excision uncertain	As COG, but nephrotic tumour in lymph nodes assessed as positive even if no viable tumour. Also the presence of non-viable tumour at any resection margin
IV	Haematogenous metastases, or tumour in lymph node outside the abdomen	Haematogenous metastases, or tumour in lymph node outside the abdomen
V	Bilateral disease, each side staged as above	Bilateral disease, each side staged as above

[9]

Histologically, Wilm's is divided into favorable and unfavorable histologies [3]. Ninety percent of Wilms tumors will demonstrate "favorable" histology, which generally has a better prognosis. Classical histological features of a "favorable" Wilms tumor include a triphasic pattern of blastema, epithelial, and stromal tissues[3]. Unfavorable demonstrate many high degrees of anaplasia and relatively poorer prognosis and survival. Anaplasia is histologically defined as hyperchromatic, pleomorphic nuclei that are three times larger than adjacent cells and have abnormal mitotic figures[3]. Histopathology on both of our patients tends to be favorable pathology.

Treatment of Wilms tumor is usually nephrectomy followed by systemic chemotherapy, but some protocols initiate chemotherapy first and do the nephrectomy later [3]. COG RTC uses immediate surgery for all unilateral tumors, and the SIOP uses preoperative chemotherapy as the first step in treatment[6]. Current therapy depends on staging and comprises multi-modality chemotherapy and surgery, with or without radiation therapy[10]. In our first patient. Surgery was the first effective treatment for Wilm's Tumor, with a cure possible in selected patients with localized disease [10]. Transperitoneal radical nephrectomy is the mainstay of treatment for most patients with Wilm's Tumor [10]. Both of our patients have been performed radical nephrectomy without pre and post-operative chemotherapy, which was not appropriate with the studies. Based on SIOP protocols, preoperative chemotherapy must be given when the tumor reaches stage 2. Actinomycin D (Act D), and Vincristine (VCR) should be given for 27 weeks [11]. Preoperative chemotherapy is considered to reduce rupture during the surgery and induces a favorable stage distribution. Post-chemo and radiotherapy are considered to reduce the recurrence of the tumor [10].

Both of our patients, Even though they did not receive the neoadjuvant and adjuvant therapy as recommended in theory, There is no significant problem found during the pre and post-surgery. After 6 months, screening was performed. However, there were no recurrence signs found in both of the patients.

Conclusion

In Conclusion, Wilms tumor has a variety of clinical characteristics in each patient. The disease is also frequently found asymptotically which leads to a delay in diagnosis. Screening for Wilm's

tumor is very important for children who have syndromes or birth defects known to be linked to this disease.

Conflict of interest

The authors declare that they have no conflict of interests.

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