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Successful Surgical Management of a Wilm's Tumor with Vascular Extension into the Inferior Vena Cava: A Case Report

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Abstract

Nephroblastoma is one of the most common renal pediatric solid tumors. It has a strong tendency of extension of the tumor thrombus along to the renal vein, the inferior vena cava, and the right atrium. We report the case of a 02 years and 08 months-year-old child, who received 8 chemotherapy doses for a left kidney nephroblastoma invading the left renal vein and inferior vena cava. Two months later, the tumor size decreased and measured 13 X 10 cm but the vascular thrombus persisted. So, he underwent left nephrectomy and the removal of the intravascular tumoral thrombus under median laparotomy and cavotomy. The postoperative CT scan showed a small hematoma and a thrombosis of the infrarenal vena cava. So, we decided to continue curative anticoagulant therapy for 3 months. Surgical management of nephroblastoma with vascular extension remains a significant challenge. Intravascular extension must be well characterized in all patients in order to indicate preoperative chemotherapy and guide resection approach.

Keywords: Nephroblastoma; Renal vein; Inferior vena cava; Tumoral thrombus; Surgery; Cavotomy

Introduction

Nephroblastoma, also known as Wilms tumor, is one of the most common renal pediatric solid tumors. It has an incidence estimated at 6% of pediatric tumors, it accounts for more than 90% of all childhood kidney tumors (1). It has a strong tendency to invade nearby vessels in the form of a thrombus. In fact, extension of tumor thrombus along to the renal vein into the inferior vena cava occurs in 4-10% of all patients. Tumor thrombus extending to the right atrium is less reported as 0.7-1 Intravascular thrombus occurs in 20–35% of patients, while extension to the IVC ranges from 4 to10% [1,2]. A thrombus reaching the atrium is a more rare event and occurs in less than 1% of all patients [3].

Treatment consists especially on preoperative chemotherapy and surgical excision, and needs multidisciplinary care involving oncologists, pediatric surgeons, and vascular surgeons [4]. We report the case of a 02 years and 08 month-year-old child, who was treated for a left kidney nephroblastoma invading the left renal vein and inferior vena cava, who underwent surgery to remove the left kidney and the intravascular tumoral thrombus.

Case Description

A 02 years and 08 months year-old child, was followed by pediatrics and oncologists for left kidney nephroblastome. The first CT scan showed a tumoral mass measuring 16 X 9.5 cm that was invading the left renal vein and the inferior vena cava (Figures 1 and 2).

The patient has then received 8 chemotherapy doses in order to reduce the tumor size and the extent of the vascular thrombus, but the initial CT scan showed an increase in tumor size and a persistence of left renal vein and IVC thrombus.

Two months later, the tumor size decreased and measured 13 X 10 cm but the vascular thrombus persisted. So, we decided to operate the patient. Surgery was performed under general anesthesia. Via a median laparotomy, we accessed the retroperitoneum. The renal tumor was large, and adherent to the diaphragm and the left colic flexure. After liberating the adhesions and controlling the left kidney, the renal artery and vein and then the ureter, we proceeded to a total nephrectomy (Figure 3).

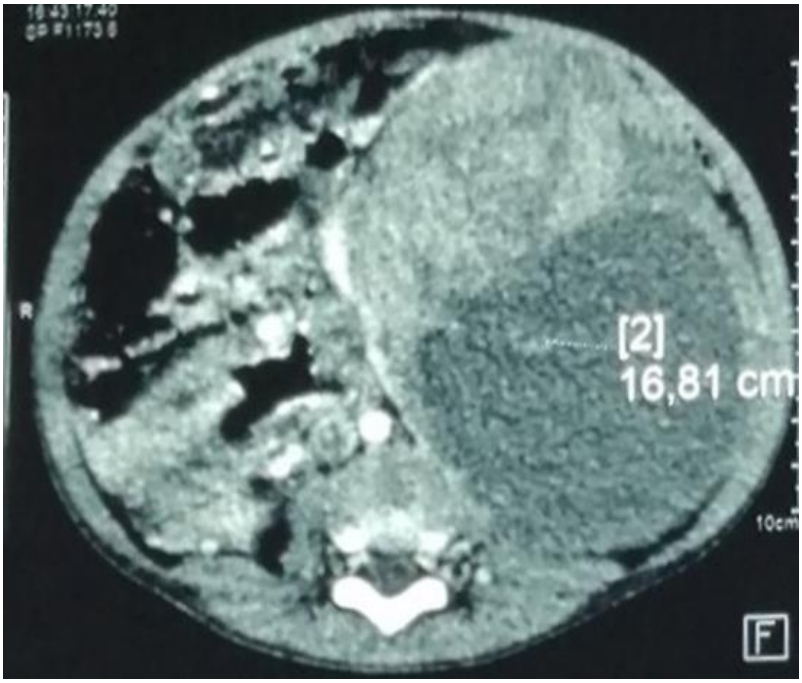

Figure 1: *Preoperative CT scan showing the left renal nephroblastoma (arrow).*

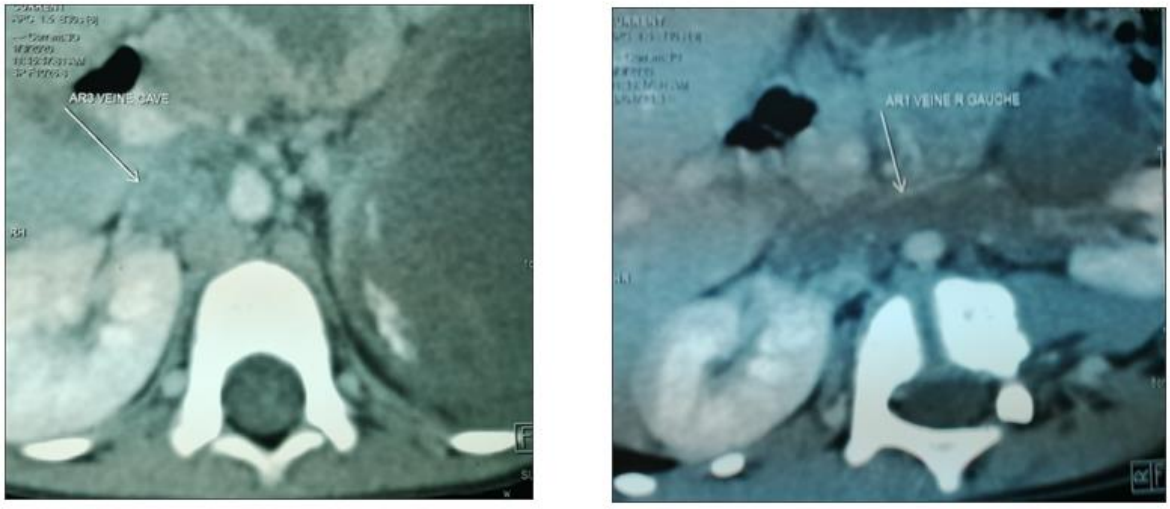

Figure 2: *vascular extension in the left renal vein and inferior vena cava.*

The second step of the procedure consisted on controlling the infrarenal and suprarenal segments of the inferior vena cava. After heparinization, the vena cava was clamped and via a covotomy we extracted the tumoral thrombus (Figure 4).

The last step of the surgery was to perform lymph nodes clearance and then closing of the median laparotomy. The patient was extubated after one day, and was put under curative heparin dose. The control CT scan showed a small hematoma and a thrombosis of the infrarenal vena cava (Figure 5), without clinical symptoms of deep venous thrombosis. So, we decided to continue curative anticoagulant drugs for 3 months.

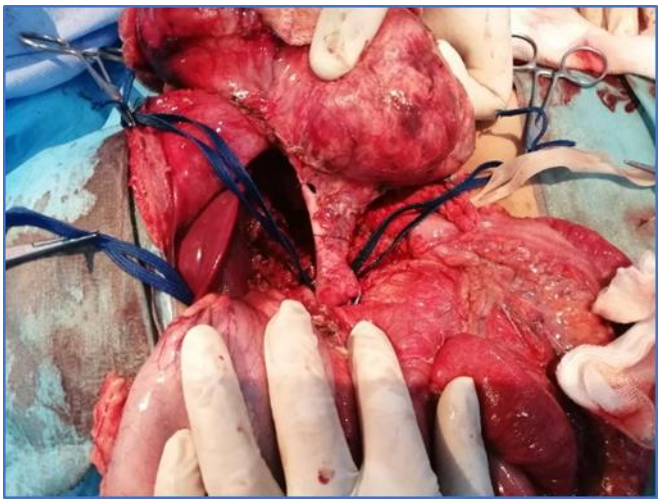

Figure 3: *Surgical control of the tumoral left kidney.*

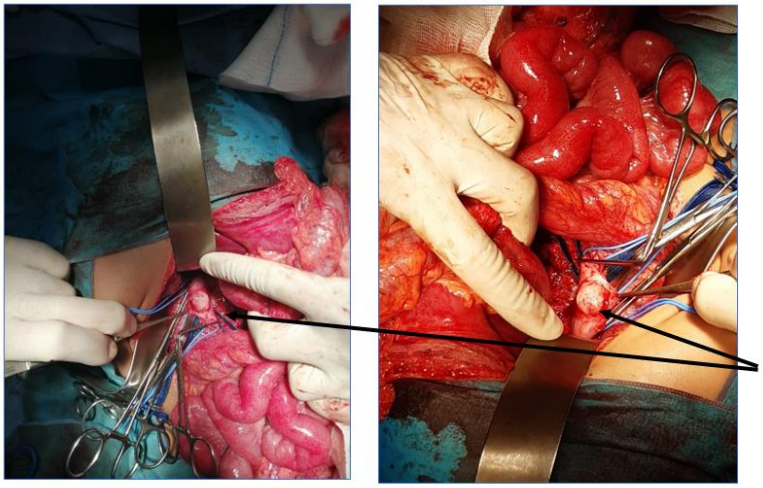

Figure 4: *Extraction of the thrombus from the inferior vena cava.*

Discussion

Nephroblastoma is the most common abdominal pediatric tumor. It is a malignant embryonic tumor that develops from the mesoderm of the intermediate cell mass before differentiation. It typically presents between ages 3 to 5 years but can also be seen in new born infants [5]. Clinical presentation is usually made of an asymptomatic abdominal mass in the majority of children. However, abdominal pain, hematuria, urinary tract infections, varicocele and hypertension or hypotension can be seen. The most common initial presenting symptom is abdominal pain (30% to 40%) followed by high blood pressure (25%), and hematuria (12% to 25%) [6].

Treatment of Wilms tumor without vascular involvement is nephrectomy in majority of cases followed by chemotherapy. There are other protocols that start with chemotherapy first and perform the nephrectomy later. Lymph nodes clearance is very important for staging and to improve general survival [6]. Intravascular extension into renal vein then the vena cava occurs in only 4-10% of patients. It is reported that intravascular extension when Wilms' tumor is located in the right kidney is more common due to the shorter renal vein. It is also reported that vascular involvement is more frequent in older children with a mean age of 3.75 years than younger children [7].

Several classifications for vascular extension were established in the past. Daum's classification that was established in 1994 [8] was the most used classification until 2013, when Abdullah et al [9] proposed a modification of Daum's classification and suggested the addition of a fifth stage for intraventricular extension of the vascular thrombus. The importance of this stage is mainly in anesthetic management. Positive pressure ventilation may cause tricuspid valve immediate cardiac arrest [7].

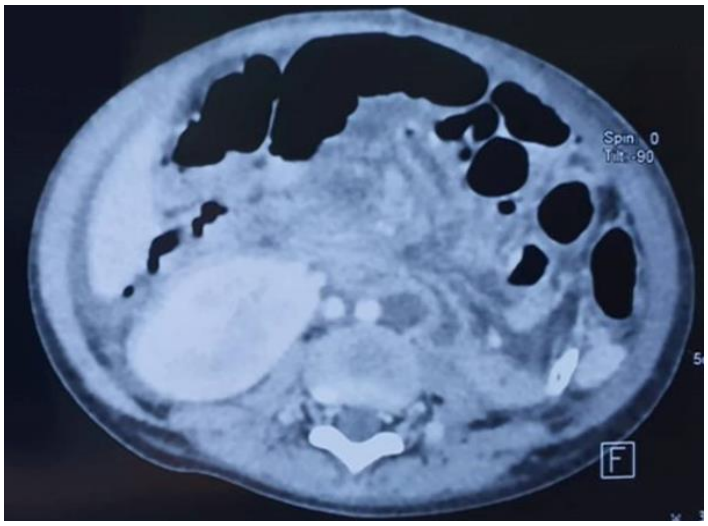

Figure 5: *control CT scan showing the presence of a small hematoma and a thrombosis of the inferior vena cava.*

The operative approach of Wilms' tumor with intravascular extension represents a constant challenge, and the surgeons should have an appropriate experience level. Extension below the hepatic veins can be approached via a laparotomy. However, proximal extension above the hepatic veins requires the same incision extended to median sternotomy [7]. The post-operative outcomes are influenced by intravascular extension [10-12], with an odds ratio of 2.2 for complications when intravascular thrombus is present. There was also an association between decrease in surgical complication's rate and the administration of neoadjuvant chemotherapy [13]. It's interesting to know that survival rates do not differ from those with uncomplicated Wilms' tumor [14].

The prognosis depends on the tumor stage and histology. Favourable histology has a better survival rates, it's estimated at 86% to 99%, while unfavorable histology survival varies from 38% to 84% depending on the stage [15-18]. Recurrence risk is estimated at 15%, it's mainly found within the first 2 years after surgery [6].

Conclusion

Surgical management of Wilms' tumor with vascular extension remains a significant challenge. Intravascular extension must be well characterized in all patients in order to indicate preoperative chemotherapy and guide resection approach. The surgical team should be multidisciplinary involving pediatric and vascular surgeons and should have an appropriate experience level.

References

1. Aspiazu D, Fernandez-Pineda I, Cabello R, Ramirez G, Alvarez-Madrid A, De Agustin JC. Surgical management of wilms tumor with intravascular extension: a single-institution experience. Pediatr Hematol Oncol. 2012; 29: 50-54.
2. Hadley GP, Sheik-Gafoor MH, Buckels NJ. The management of nephroblastoma with cavo-atrial disease at presentation: experience from a developing country. Pediatr Surg Int. 2010; 26: 1169-1172.
3. Al Diab A, Hirmas N, Almousa A, Abu-hijlih R, Aljlouni F, Sultan I, et al. Inferior vena cava involvement in children with wilms tumor. Pediatr Surg Int. 2017; 33: 569-573.
4. Emir S. Wilms tumor with intravascular tumor thrombus. Transl Pediatr. 2014; 3: 5.
5. Akyön MG, Arslan G. Pulmonary embolism during surgery for a wilms' tumour (Nephroblastoma). Br J Anaesth. 1981; 53: 903-905.
6. Leslie SW, Sajjad H, Murphy PB. Wilms Tumor (Nephroblastoma). In: StatPearls. Treasure Island (FL): Stat Pearls Publishing; 2020.
7. McMahon S, Carachi R. Wilms′ tumor with intravascular extension: A review article. J Indian Assoc Pediatr Surg. 2014; 19: 195.
8. Daum R, Roth H, Zachariou Z. Tumor infiltration of the vena cava in nephroblastoma. Eur J Pediatr Surg. 1994; 4: 16-20.
9. Abdullah Y, Karpelowsky J, Davidson A, Thomas J, Brooks A, Hewitson J, et al. Management of nine cases of Wilms' tumour with intracardiac extension - a single centre experience. J Pediatr Surg. 2013; 48: 394-399.
10. Nakayama DK, Delorimier AA, O'neill JA, Norkool P. A Report of the National Wilms' Tumor Study: Ann Surg. 1986; 204: 693-697.
11. Ritchey ML, Pringle KC, Breslow NE, Takashima J, Moksness J, Zuppan CW, et al. Management and outcome of inoperable wilms tumor a report of national wilms tumor study-3: Ann Surg. 1994; 220: 683-690.
12. Murthi GV, Kocyildirim E, Sellathury S, Cuckow PM, Wilcox DT, Michalski A, et al. Wilms' tumour with persistent intravascular extension: A review of the surgical aspects of management. J Pediatr Urol. 2006; 2: 439-445.
13. Shamberger RC, Ritchey ML, Haase GM, Bergemann TL, Loechelt-Yoshioka T, Breslow NE, et al. Intravascular extension of wilms tumor: Ann Surg. 2001; 234: 116-121.
14. Szavay P, Luithle T, Semler O, Graf N, Fuchs J. Surgery of cavoatrial tumor thrombus in nephroblastoma: A report of the SIOP/GPOH study. Pediatr Blood Cancer. 2004; 43: 40-45.
15. Rossoff J, Tse WT, Duerst RE, Schneiderman J, Morgan E, Kletzel M, et al. High-dose chemotherapy and autologous hematopoietic stem-cell rescue for treatment of relapsed and refractory Wilms tumor: Re-evaluating outcomes. Pediatr Hematol Oncol. 2018; 35: 316-321.
16. Wang X, Song P, Huang C, Yuan N, Zhao X, Xu C. Weighted gene co-expression network analysis for identifying hub genes in association with prognosis in Wilms tumor. Mol Med Rep. 2019.
17. Zhang W, Lu X, Cui P, Piao C, Xiao M, Liu X, et al. Phase I/II clinical trial of a Wilms' tumor 1-targeted dendritic cell vaccination-based immunotherapy in patients with advanced cancer. Cancer Immunol Immunother. 2019; 68: 121-130.
18. Han Q, Li K, Dong K, Xiao X, Yao W, Liu G. Clinical features, treatment, and outcomes of bilateral Wilms' tumor: A systematic review and meta-analysis. J Pediatr Surg. 2018; 53: 2465-2469.