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Nephroureterectomy for Pediatric Giant Wilms Tumor

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Abstract

Introduction: Wilms' tumor most often occurs in children, with an average age of about 3 to 4 years. About 75% of cases occur in young children aged less than 5 years. These tumors usually develop in one kidney. In the United States, surgery is the first treatment for most Wilms tumors, followed by chemotherapy. According to the International Society of Pediatric Oncology, neoadjuvant chemotherapy is performed first, followed by surgery. **Case Presentation:** Two Years and six months child was referred to our hospital with giant abdominal mass with difficulty to determine its origin due to its large size. On examination, it was found that the abdomen was distended with a mass palpable in all of quadrant of abdomen. A contrast enhanced abdominal CT showed a solid lumpy mass in the left kidney with a size of 22.36 x 19.66 x 17.12 cm. The renal mass crossed the midline and filled most of the abdominal cavity. The contra lateral kidney shows normal function. **Discussion:** The most patient come with presenting complaint was abdominal mass (50%), and 35% of the patients presented with an abdominal lump. Western studies reported 74% of patients presenting with an abdominal mass. A study by Guruprasad et al reported that 90% of their patients presented with abdominal mass. **Conclusion:** Combination of Neoadjuvant chemotherapy and Surgery is important in the management of Wilms Tumor. Radical Nephrectomy and Ureterectomy can be performed safely if performed by an experienced surgeon

Keywords : Wilms' tumor, Neoadjuvant chemotherapy, Radical Nephrectomy

Abstrak

Pendahuluan: *Tumor Wilms paling sering terjadi pada anak-anak, dengan usia rata-rata sekitar 3 hingga 4 tahun. Sekitar 75% kasus terjadi pada anak kecil berusia kurang dari 5 tahun. Tumor ini biasanya berkembang di satu ginjal. Di Amerika Serikat, pembedahan merupakan pengobatan pertama untuk sebagian besar tumor Wilms, diikuti oleh kemoterapi. Menurut International Society of Pediatric Oncology, kemoterapi neoadjuvan dilakukan terlebih dahulu, diikuti oleh pembedahan.* ***Presentasi Kasus:*** *Seorang anak berusia dua tahun enam bulan dirujuk ke rumah sakit kami dengan massa abdomen yang sangat besar, dan sulit untuk menentukan asal-usulnya karena ukurannya yang besar. Pada pemeriksaan, abdomen tampak membesar dengan massa yang teraba di seluruh kuadran abdomen. CT abdomen dengan kontras menunjukkan massa padat dan berbenjol di ginjal kiri dengan ukuran 22,36 x 19,66 x 17,12 cm. Massa ginjal tersebut melewati garis tengah dan memenuhi sebagian besar rongga abdomen. Ginjal kontralateral menunjukkan fungsi normal.* ***Diskusi:*** *Pasien terbanyak datang dengan keluhan utama berupa massa abdomen (50%), dan 35% pasien datang dengan benjolan abdomen. Studi di Barat melaporkan 74% pasien datang dengan massa abdomen. Sebuah studi oleh Guruprasad dkk. melaporkan bahwa 90% pasien mereka datang dengan massa abdomen.* ***Kesimpulan:*** *Kombinasi kemoterapi neoadjuvan dan pembedahan penting dalam penanganan tumor Wilms. Nefrektomi radikal dan ureterektomi dapat dilakukan dengan aman jika dilakukan oleh ahli bedah berpengalaman.*

Kata kunci : *Tumor Wilms, Kemoterapi neoadjuvan, Nefrektomi radikal*

I. INTRODUCTION

Wilms' tumor most often occurs in children, with an average age of about 3 to 4 years. About 75% of cases occur in young children aged less than 5 years. These tumors usually develop in one kidney. In the United States, surgery is the first treatment for most Wilms tumors[1]. In Europe, they prefer to give a short course of chemotherapy before surgery. Five-year survival is greater than 90% for children with all stages of wilms tumor (WT) favorable histology who receive appropriate treatment. Combination of various therapeutic modalities is important to obtain the best outcome for the patient[2.] Large tumors are difficult to remove because of their proximity to blood vessels and other vital structures. In children, some guidelines recommend the use of chemotherapy, radiation or a combination of both before surgery. An enlarged of mass in abdominal that feels hard is the chief complaint that Wilms tumor patients complain of. Another sign are hypertension, abdominal pain, anemia and sometimes hematuria[3,4.]

Total nephrectomy or partial nephrectomy followed by or without exploratory regional lymphadenectomy is a surgical therapy that can do. Although Wilms tumor can grow to a very large size but rarely invades surrounding structures. There are indications of delayed resection which include very large masses, bilateral disease; pulmonary metastases; extension of tumor thrombus into the IVC. Neoadjuvant therapy may be important in some cases with complications and risk of surgery[1].

II. CASE PRESENTATION

Two Years and six months child was referred to Moewardi Hospital with giant abdominal mass with difficulty to determine its origin due to its large size. On examination, it was found that the abdomen was distended with a mass palpable in all of quadrant of abdomen. The condition of tall patient is 85 cm, weighs 14.9 kg, upper arm circumference 11.5 cm and abdominal circumference 75 cm. patients with malnutrition status and normochromic normocytic anemia. During the examination, hypertension was found but kidney function (normal urea/creatinine) and liver function normal. Patients with no signs of abdominal Patients with no signs of acute abdominal.

Diagnostic was work up also staging using clinical exam, laboratory and radiology. A contrast enhanced abdominal CT showed a solid lumpy mass in the left kidney with a size of 22.36 x 19.66 x 17.12 cm. The renal mass crossed the midline and filled most of the abdominal cavity. The contra lateral kidney shows normal function. There are no signs of metastasis in the liver and major vascular invasion disorders. Lung examination showed no metastasis or infiltrate lesions.

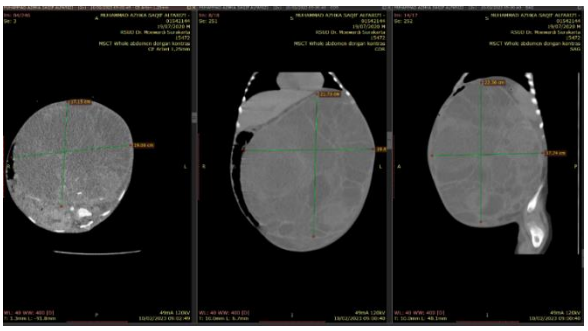


FIGURE 1. CONTRAST ABDOMINAL CT SHOWED A SOLID MASS IN THE LEFT KIDNEY WITH A SIZE OF 22.36 X 19.66 X 17.12 CM.

In our institution, we undertake the *International Society of Pediatric Oncology* suggestions in treatment of Wilms Tumors in which neo adjuvant treatment is the initial treatment for the large nephroblastoma.[5] Patient before performing definitive surgical management was take a biopsy dilanjutkan chemotherapy 3x using a vincristine regimen of 14.8 mg/m2. After neoadjuvant chemotherapy treatment, the size of the tumor remained the same and it was decided to perform surgery. The tumor turned into accessed thru a transperitoneal technique because of its extraordinarily massive length to keep away from rupture of arterial feeding

tumor and intraperitoneal spillage. Initial dissection turned into finished in attention of visibility, extensibility and simplicity of resection. After the transperitoneal incision, the intestine and colon were removed to the medial side to gain retroperitoneal access. A white line incision is performed to access the retroperitoneal, followed by blunt dissection posteriorly to separate the intestine from the colon.

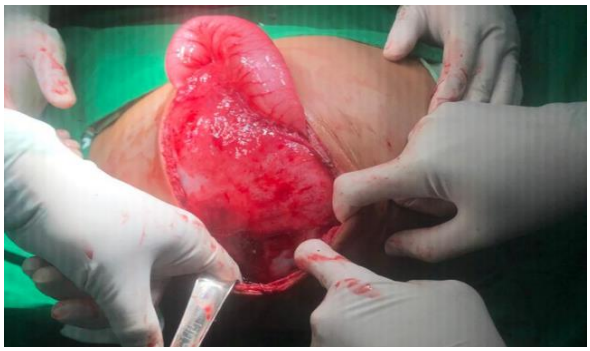


FIGURE 2. TRANSPERITONEAL INCISION IS CONTINUED TO SET ASIDE PART OF THE INTESTINE MEDIALLY AWAY FROM THE ORIGIN OF THE TUMOR

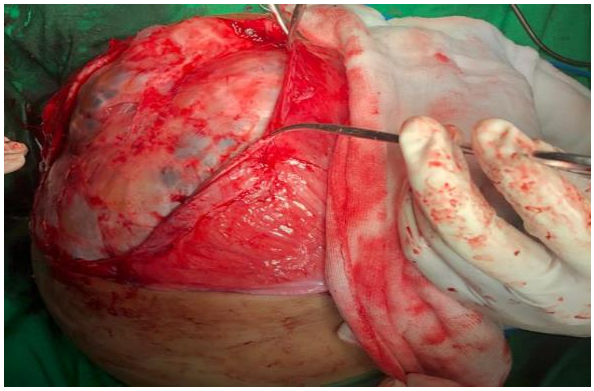


FIGURE 3. WHITE LINE OPENING TO GAIN RETROPERITONEAL ACCESS

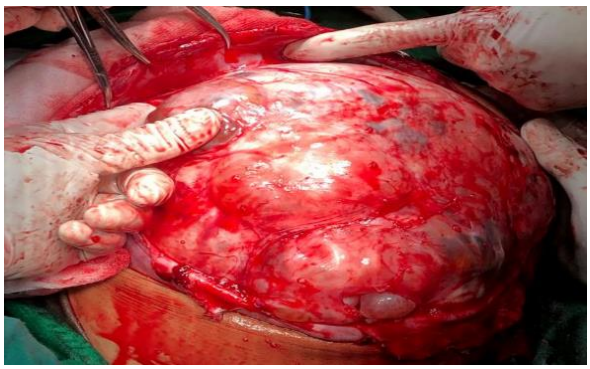
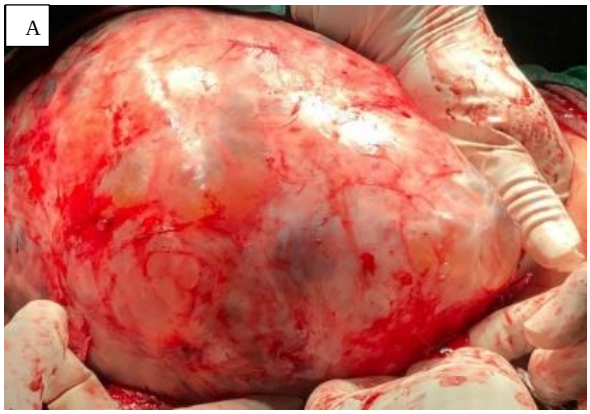


FIGURE 4. BLUNT DISSECTION POSTERIORLY, SEPARATING THE MUSCLES OF THE POSTERIOR ABDOMINAL WALL



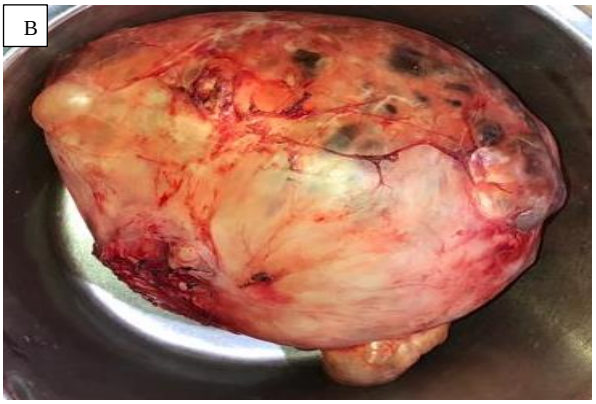


FIGURE 5. (A). CUTTING THE RENAL HILUM AND URETERECTOMY. (B). COMPLETELY RESECTED WITH THE CAPSULE INTACT.

Complete excision was performed resulting in an intact encapsulated tumor. The tumor's dimensions were (26 x 20 x 18 cm) and its weight was 3.8 kg. Histopathological examination of the resected specimen revealed cellular, vascular and degenerative tumor. The cellular stroma of tumor resembles a myxoid, and myofibroblastic component. Cellular/diffuse cell proliferation, embryonic cells, small atypic nuclei and hyperchromaticity. Those cells confirmed the diagnosis of Wilms tumor.

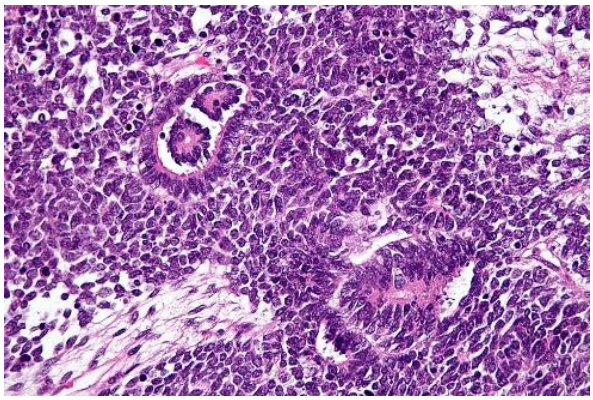


FIGURE 6. PATHOLOGY ANATOMICAL FEATURES SHOW SPINDLE CELL STROMA WITH DIFFUSE/CELLULAR CELL PROLIFERATION, EMBRYONIC CELLS, SMALL ATYPICAL NUCLEI AND HYPERCHROMATICITY.

After the resection process, the patient was treated in the pediatric intensive care unit for hemodynamic monitoring, administration of antibiotics and fluid management. Foley's catheter was removed on the second postoperative day, and enteral feeding was allowed on the second postoperative day. On the fifth postoperative day, the patient was checked with a stable general and good wound healing. A 6-month evaluation is carried out by looking at the patient's clinical and laboratory conditions. Clinically, the patient's complaints became more comfortable, less tight and body weight increased 3 kg compared to 1 months before surgery. There were no complications from the surgical wound and clinical evaluation after 6 months. Renal function tests is increased from laboratory finding.

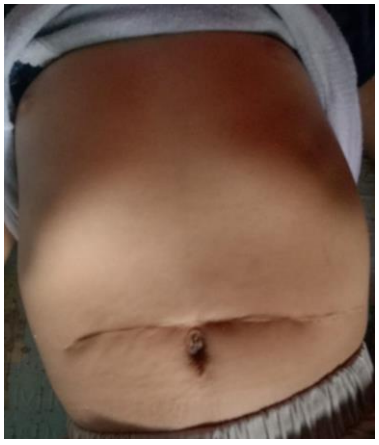


FIGURE 7. NO COMPLICATION OF WOUND POSTOPETION (EVALUATION AFTER 6 MONTHS OF SURGERY)

III. DISCUSSION

The most patient come with presenting complaint was abdominal mass (50%), and 35% of the patients presented with an abdominal lump. Western studies reported 74% of patients presenting with an abdominal mass.[6] A study by Guruprasad et al reported that 90% of their patients presented with abdominal mass. Treatment of Wilms Tumor includes surgery, neoadjuvant or adjuvant chemotherapy, and radiation therapy (RT) if needed. Careful use of to be had remedies is vital to maximum treatment and reduce long-time period toxicities.[7]

Chemotherapy is usually a systemic therapy. This means that the drugs travel through the bloodstream to reach and destroy cancer cells all over the body that may have broken away from the primary tumor in the kidney. Neoadjuvant chemotherapy is recommended to shrink the tumors before surgery in children with bilateral WT, those with initially unresectable unilateral tumors, or those with predisposing conditions and either localized or metastatic unilateral renal tumors. Specific chemotherapy regimens are given for six weeks after which the tumor reaction is assessed.[8]

International Society of Pediatric Oncology (SIOP) guidelines adopted in European countries recommends preoperative chemotherapy as the initial therapeutic modality. The goal of chemotherapy is to reduce the size of the tumor to make surgery easier. The advantages of giving neoadjuvant chemotherapy include 1) reducing tumor mass, 2) preventing spillage, 3) reducing chances of distant spread.[9] Surgery is usually recommended via way of means of week 12 of neoadjuvant chemotherapy primarily based totally on scientific trial information displaying that persevering with chemotherapy past 12 weeks does now no

longer yield persevered tumor shrinkage. The SIOP guideline recommends using 4-week pretreatment with vincristine (weekly) and dactinomycin (biweekly) is given for patients with bilateral tumors, vincristine–dactinomycin for no longer than 9– 12 weeks is usually recommended (doxorubicin is introduced for reinforcement in a few patients). This affected person become given neoadjuvant vincristine for six weeks.[10]

Transverse or Chevron incision, is a design that is often used to carry out radical nephrectomy. The design must be taken to completely expose the kidney and tumor on both sides and to identify mesenteric and renal vessels to prevent inadvertent damage. Response to chemotherapy varies, generally decreasing by 35.7%.[11] The standard surgical procedure for a unilateral WT is a transperitoneal radical nephroureterectomy with ipsilateral lymph node sampling. Although enormous lymphadenectomy isn't always required, perihilar and periaortic or pericaval lymph node samples should be received in all cases, as they're vital for good enough staging and making plans of post-operative management.[12]

Wilms' tumor can grow to a large size when first discovered; however, radical excision of these large lesions is still possible because these tumors are well demarcated and well encapsulated and tend to rarely invade surrounding structures. To make it easier, chemotherapy is done first, but if indeed chemotherapy has resistance or the size is appropriate for surgery, then surgery is performed. The COG report suggested that the surgeon should exercise extreme caution when attempting to resect a Wilms tumor larger than 12 cm in diameter, because there is an increased risk of tumor rupture of the intraperitoneal area which elevates the stage of the disease. In this report, resection of a 20 cm diameter nephroblastoma could be performed safely without intraperitoneal rupture of the tumor.[13]

Radical nephrectomy is performed using a transperitoneal incision. Initial control of the renal hilar blood vessels was recommended before mobilizing the primary tumor. This is not possible with giant tumors which need to be mobilized beforehand to allow safe exposure of the renal hilum[4.] Early attempts to bind the hilar vessels can result in vascular injury to the mesenteric artery, celiac vessels and aorta. In our patient, the surgical approach via a transverse abdominal incision from left to right beyond the midline allowed good exposure. After entering the peritoneum, the colon is moved medially carefully to maintain its blood supply.[14] Opening by opening the white line is still being done. If there is peritoneal attachment to the mass, it is carefully separated. The ureter is identified, tied and cut close to the bladder. The tumor is separated and entered through the posterolateral side separating the tumor from the posterior abdominal wall.[15] Continuous pushing from behind, till the large tumor become absolutely brought into the stomach wall incision being best connected to the renal hilar pedicle which become eventually ligated the usage of non-absorbable suture cloth and divided, beginning with the renal vein first to keep away from any feasible hematogenous tumor spreads. The renal hilum was palpable and the pericaval lymph nodes were sampled and found to be tumor-free by anatomical pathology. Similar observations were reported by others.[16]

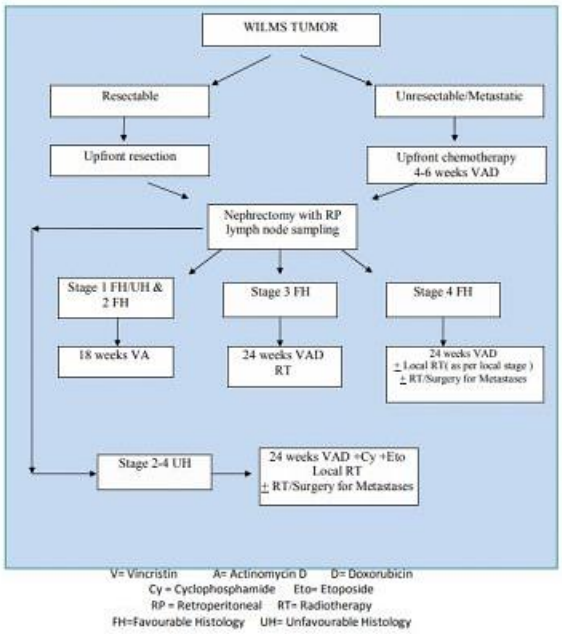


FIGURE 9. GUIDELINES OF WILMS TUMOR BASED ON INTERNATIONAL SOCIETY OF PEDIATRIC ONCOLOGY (SIOP)

IV. CONCLUSION

Combination of Neoadjuvant chemotherapy and Surgery is important in the management of Wilms Tumor. Radical Nephrectomy and Ureterectomy can be performed safely if performed by an experienced surgeon. Surgery with transperitoneal design incision to facilitate access and dissection is carried out carefully to obtain maximum results during resection and avoid vascular injury or damage to surrounding organs.

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