

WILMS' TUMOR: PRESENTING AS HUGE ABDOMINAL MASS

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ABSTRACT

Wilms' tumor or nephroblastoma accounts for more than 95% of all kidney tumors in children. It is usually discovered during the first few years of life. It is usually unilateral but can occur bilaterally. It is among the intermediate tumors of the kidney with clear metastatic potential, especially to the lungs. Chemotherapy followed by surgery is the standard treatment. We present a case of Wilms' tumor in 18 months old female child.

KEY WORDS: Wilms' tumor; Kidney; Nephrectomy.

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INTRODUCTION

Wilms' tumor (nephroblastoma), an embryonal type of renal cancer accounts for nearly 6% of all pediatric cancers and more than 95% of all kidney tumors in children.¹ The tumor usually arises in a single kidney while synchronous bilateral or multifocal tumors can occur in approximately 10% of patients and tend to present at an earlier age.¹⁻⁴ Wilms' tumor can also be diagnosed in adolescents or adults, but this is extremely rare, representing less than 1% of all renal tumors.⁵ The usual treatment approach in most patients is a combination of surgery and chemotherapy. Substantial progress in the treatment of Wilms' tumor over the past few decades has been made by refining risk stratification and by the use of existing chemotherapy schedules. This has improved overall survival for patients with Wilms' tumor in developed countries to greater than 90% for localized disease and 75% for metastatic disease.^{6,7} This excellent outcome results from collaborative efforts among pediatric surgeons, pathologists, radiologists and oncologists.² The two largest collaborative groups that have studied the optimal management of Wilms' tumor are the Children's Oncology Group (COG) and the International Society of Pediatric Oncology (SIOP).² The COG recommends primary surgery before an adjuvant treatment except in specific circumstances such as synchronous bilateral disease. By contrast, the SIOP approach favours pre-operative chemotherapy for all cases except very young infants (<6 months of age).⁸⁻¹⁰ Clinical outcomes are

excellent in both groups, and there is an ongoing debate on the merits of each approach.⁶

CASE REPORT

An 18 month old female baby presented to the surgical unit of DHQ Teaching Hospital, D.I.Khan with gradually increasing abdominal distension for the last eight months and was associated with pain, fever and haematuria for the last two months. Patient was admitted and on examination she was having asymmetrical mass in the right side of the abdomen which was hard, immobile and not moving with respiration but was ballotable. Ultrasound abdomen revealed right renal tumor most likely Wilms' tumor which was confirmed by CT scan abdomen, which showed huge mass compressing completely on right kidney. X-ray chest was clear. A patient was malnourished. After doing all base line investigations and IVU to assess the other kidney function and arranging blood the patient was operated. Per-operatively she was having huge tumor completely compressing the right kidney and radical nephrectomy was done with complete removal of the tumor and maximum possible lymph nodes. Photographs of the patient/tumor were taken preoperatively and postoperatively. Patient recovered uneventfully. Her biopsy report confirmed Wilms' tumor. Patient was sent to DINAR Radio-nuclear Hospital for further chemotherapy. (Fig. 1-3)

DISCUSSION

Wilms' tumor is the second most common intra-abdominal malignancy of childhood and overall fifth most common pediatric malignancy.^{1,2,11} Survival for the patients was once less than 30% but is currently greater than 90%.¹ This improvement is due to continuous trials, systemic and multidisciplinary approach toward the tumor and still trials are

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Figure 1: Patient of Wilms' tumor before surgery.

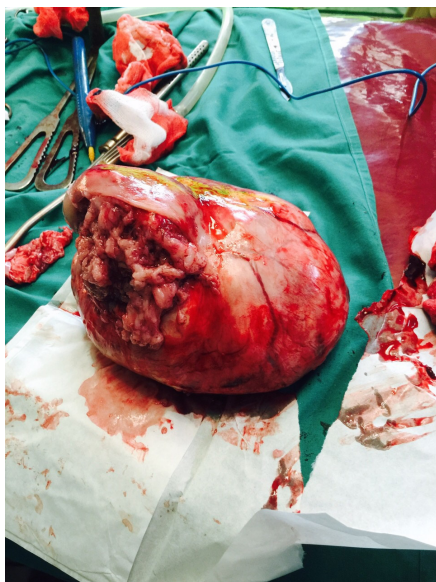


Figure 2: Photograph of excised Wilms' tumor.



Figure 3: Post-operative photograph of the patient with Wilms' tumor.

under way in different countries aiming at excellent survival, while minimizing therapy related toxicity.^{2,3,12}

Tumor stage, histological type, age, tumor size and genetic type are all prognostic factors.¹ Appropriate therapy as well as prognosis depends upon accurate staging at the time of diagnosis and histological assessment of lymph node as well.¹³ International Society of Pediatric Oncology (SIOP) strongly recommends classic open nephrectomy with chemotherapy. The appropriate treatment option for the low risk tumors, i.e. child age less than 2 year and tumor weight less than 550 g, is surgery alone, while tumors with stage 1 should be treated with pre-operative chemotherapy.¹³⁻¹⁵ The main controversies about the timing of surgery is whether or not to administer preoperative chemotherapy, as suggested by the International Society of Pediatric Oncology (SIOP). Some studies suggest dangers of under treatment with an increased incidence of infra-diaphragmatic relapses in patients who did not receive postoperative radiation therapy.^{6,7} Likely, patients with lymph node involvement receiving preoperative chemotherapy have less chances of recurrence.⁸

The role of surgery in the therapy of Wilms' tumor is paramount since a meticulous and well performed procedure will accurately determine the stage of the patient and their future therapy.^{1,2,6,11} Several trials are also underway to assess the role of minimal invasive surgery and nephron sparing nephrectomy but however not recommended because after nephron sparing nephrectomy, these patients need radiotherapy to prevent relapse that will turn kidney fibrotic and in minimal invasive surgery, complete resection and lymph node clearance is not achieved.^{5,6,10}

This patient present in advance stage of the disease. Surgery was done immediately because patient was in distress due to huge abdominal mass. During surgery tumor was completely resected with maximum lymph node clearance. Dramatic recovery and improvement in the condition of patient was observed. Such type of patients if properly operated and treated with chemotherapy, result in improved survival.¹¹

CONCLUSION

Wilms' tumor should be considered in the differential diagnosis of asymmetrical abdominal mass in a child. They should be managed with the collaboration of Pediatric Surgeon, Pathologist, Radiologist and Oncologist.

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