Short term surgical outcomes of Wilms tumour from a single institute

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Abstract

Wilms tumour is the commonest solid tumour of childhood in Pakistan. In developed world the long-term outcome of Wilms tumour have significantly improved reaching up to 98% long term survival. We have no National data of prevalence, treatment and survival of this condition in Pakistan. Surgical treatment also varies in various parts of the country. We have studied our patients presenting with a diagnosis of Wilms tumour to our institute from Jan 2014 to April 2016. A total of 42 patients were operated for Wilms tumour. The most common symptoms were abdominal mass (75%), pain (28%) and fever (19%). A total of 48 tumour resections were performed, 45 total nephrectomies and 3 had nephron sparing surgery (NSS). Mean operative time was 225±78.7 minutes. Mean blood loss was 165±223.5ml. Mean size of the tumour was 102±48.4mm and mean weight of the tumour was 433±400.7gm. Ninety percent patients had a favourable histology. Mean high dependency unit (HDU) stay was 1.16±1.2 day and mean hospital stay was 6.89±3.47 days. Complications were observed in 8 patients. Surgery remains a major part of treatment for Wilms tumour.

Keywords: Wilms tumour, Nephroblastoma, Wilms nephrectomy, nephron sparing surgery

Introduction

Malignant solid tumours constitute a major cause of morbidity and mortality in children and comprise about half of all childhood malignancies.[1] Wilms tumour (WT), the fifth most common paediatric malignancy, is diagnosed in approximately 500 children each year in the United States.[2,3] There is no National tumour registry in Pakistan. Pakistan Medical Research Council Cancer Study group revealed that malignant tumours in under 15 years constituted 4.38% of total malignancies diagnosed.[4] The most common solid tumours in children are Wilms tumours, neuroblastoma, lymphoma, retinoblastoma and bone tumours most occurring in patients under 5 years of age.[5] In one study of 338 paediatric patients under 15 years of age, Wilms tumours constituted 7.4% and lymphomas 20.5% of all tumours.[6]

In developed countries the outcome of children suffering from solid tumours has dramatically improved over the past 30 years.[7,8] Several factors are responsible for this fact. One major factor is the treatment of patients according to well-established guidelines and protocols established by various groups through prospective clinical trials and integrated therapy combining chemotherapy, surgery and radiotherapy. The major examples are International Society of Pediatric Oncology (SIOP) Renal Tumour Study Group (SIOP-RTSG) in Europe & overseas and Renal Tumour committee of Children Oncology Group (COG), previously known as NWTS North America.

SKMCH has a specialized paediatric oncology service to deal with paediatric malignancies including Wilms tumours. All patients are appropriately investigated and are discussed in multidisciplinary meetings prior to commencement of any treatment. We share our experience of the management of patients with Wilms tumour with early surgical outcomes.

Case Series

All patients presenting to SKMH with a diagnosis of Wilms tumour from Jan 2014 to April 2016 were included. Patients who presented after nephrectomy from elsewhere were excluded from this review. Core needle biopsy at presentation was performed in all patients for tissue diagnosis. All patients followed UKCCLG (United Kingdom Children Cancer and Leukemia Group) protocol for therapy.Two-drug preoperative chemotherapy with Vincristine and Actinomycin was used in non-metastatic tumour for 4 cycles and Doxorubicin added for metastatic tumours and total duration of preoperative chemotherapy was increased to 6 cycles. Mean duration of time was 5.08±2.58 weeks.

Total of 42 out of 44 patients were operated. The most common symptoms were abdominal mass (75%), pain (28%), fever (19%), hematuria (12.5%) and weight loss (6.2%). 21 had right sided tumours, 15 had left and 6 had bilateral tumours. Clinical stage at presentation was Stage

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I in 34%, II in 22%, III in 16%and IV in 27% patients.One patient with WAGR syndrome had bilateral Wilms and two patients had associated Denys Drash syndrome.

Forty eight resections were performed, 45 total nephrectomies and 3 nephron sparing surgery (NSS). Two laparoscopic and 5 lap assisted procedures were performed. Rest of the procedures were conventional open technique. Laparoscopic procedure was performed in relatively smaller sized tumours. Inferior vena cava (IVC) was explored in 6 patients. Two patients had a splenectomy and one required a colonic resection and anastomosis for enbloc tumour excision. Seven patients (16%) had a per-operative tumour rupture, diagnoses either at CT scan or at operation.

Mean operative time was 216±78.7 minutes. Mean blood loss was 165.4±223.5 ml. Mean size of the tumour was 102.7±48.45mm and mean weight of the tumour was 433±400.7gm. Pathological stage according to National Wilm's tumour studies (NWTS)[9] was I in 62% patients, II in 20%, III in 14% and IV in 4%. 90% patients had a favorable histology. Five patients (12%) had a complete histological response, 25 (60%) had partial response, 1 had a disease progression and rest had no response to chemotherapy. Renal vein was involved in 3(7%) of the patients. Adrenal was involved in 2 (5%) patients. Margins were positive in only 1 (2%) patient who had unresectable advanced intraperitoneal and reteroperitoneal disease. Mean number of lymph nodes harvested was 3.96±3.7.

Post-operative Cefuroxime and Metronidazole were given for a median duration of 2 days (1-6 days). Mean time to initiate oral sips were 27.7±12.83 hours and initiation of soft diet was 70.5±58.1 hours. Mean high dependency unit (HDU) stay was 1.16±1.2 day and mean hospital stay was 6.89±3.45 days. Complications were observed in 8 patients. 4 patients had persistent vomiting post-operatively prolonging admission. One patient underwent laparotomy for bowel obstruction. One patient developed pneumonia and was intubated. Two patients had wound infections.

Preoperative chemotherapy was given to 42(94%) patients, 2 (4%) had inconclusive biopsies and underwent upfront nephrectomy. One patient (2%) had a tumour bleed and had to undergo emergent resection. Post-operative chemotherapy was given to all patients. Vincristine, Actinomycin and Doxorubicin (VAD) was the most commonly prescribed regimen for a mean duration of 21.41±11.05 weeks. We had one mortality in our series due to inoperable tumour, who received supportive care with not for cure intent, however no surgical mortality was reported.

Thirteen patients had pulmonary metastases, these resolved after neo adjuvant chemotherapy in 9 patients. 3 had to undergo wedge resections using Video assisted thoracoscopic surgery (VATS). One patient had bilateral staged metastatectomy. Another patient had extensive metastatic disease with involvement of liver lungs and mediastinal nodes. All these metastases resolved with chemotherapy and did not require surgical resection.

Discussion

Outcomes of patients with Wilms Tumour have significantly improved with combined therapy approaches integrating chemotherapy, surgery and radiotherapy.[10,11] Multimodality treatment strategies now result in overall survival rates approaching 90%. In tumours with favorable histology, the NWTS reported 96% four year survival rates for Stage I tumours, 92% for stage II and 87% for stage III for stage IV or unfavorable histology, the five year survival rates are now 73%.[10] Although our follow-up is mainly to describe early surgical outcomes, however the stage of disease and current status of our patients, minimal morbidity and no mortality indicate possible good outcomes.

For Wilms tumour, two most commonly followed guidelines are the SIOP and COG guidelines. SIOP guidelines recommend that all patients should undergo pre-operative chemotherapy before surgery. The advantage of SIOP protocol is reduction in the incidence of tumour rupture by reducing the vascularity of the tumour, reduced risk of tumour spillage, a more favorable stage distribution and reduced treatment burden. However no histopathological diagnosis is achieved before initiating chemotherapy which might result in giving chemotherapy for a benign condition or giving wrong chemotherapy for some other diagnosis.[9]

The COG recommends upfront surgery followed by chemotherapy. This provides accurate staging and treatment strategy but with higher incidence of tumour rupture and intraoperative spillage.[11]

At SKMCH we follow UKCCLG (United Kingdom Children Cancer and Leukemia Group) guidelines and always perform a core needle biopsy before initiation of neo adjuvant chemotherapy. This is followed by surgery; total nephrectomy with lymph node dissection. This is again followed by a recommended course of chemotherapy based on the final histology at nephrectomy specimen and nodal disease status. There is a lot of concern regarding a pre-operative biopsy of the tumour but it has been shown through studies that biopsy of the tumour does not result in any added risk to patient's prognosis[11] as the open biopsy would do. Open biopsy is therefore

contraindicated and upstages the tumour.

SKMCH is a referral center and receives patients from all over the country. Our patients have a delayed presentation and usually present with very large tumours. Our median tumour size was 120mm and median tumour weight was 351g. Twenty seven (27%) of our patients had metastatic disease at presentation. Despite this all our patients had very good post-operative outcomes with results comparable to that of other studies.

Conclusion

Surgery remains a major part of treatment for Wilms tumour. By strictly adhering to the recommended guidelines, multidisciplinary team approach and organized care of individual patients, the outcomes of surgery can be greatly improved.

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