IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 16, Issue 6 Ver. V (June. 2017), PP 86-88
www.iosrjournals.org

Outcome of Wilm`s Tumour in A Tertiary Care Centre: Our Experience.

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Abstract: *WT is the most common renal tumor of infancy and childhood. Its incidence is one per 10,000 children under the age of 15 years worldwide. Multimodality treatment has resulted in a significant improvement in the four-year survival, from approximately 30% in the 1930s to more than 85% in the modern era. This was also the first solid tumour where the role of adjuvant chemotherapy was established. we report in the present study our experiences of the outcome of multimodality treatment of Wilms' tumor at a tertiary care centre. Our results demonstrated that factors such as late presentation, poverty, poor compliance to treatment, and lack of multidisciplinary collaboration remain major challenges in managing Wilms Tumour patients in a developing country.*

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I. Introduction

Wilms' tumor is the fourth most common childhood cancer. Wilms' tumor may occur as a part of a multiple malformation syndromes including WAGR, Denys-Drash, and Beckwith-Wiedemann syndrome. Wilms' tumor is associated with mutations of a number of tumor suppressor genes. These include WT1, p53 and mutations at the 11p15.5 loci [1]. The majority of patients present with an abdominal lump and CT scan is the usual imaging modality for determining the extent of disease.The treatment of Wilms' tumor (WT) has evolved from surgical excision as the prime method of treatment to combined multimodal treatment. With surgery alone the survival was 30 % in the early 1930 [1], improved to 47 % with use of surgery and radiotherapy in early 1950 [2] and further introduction of adjuvant chemotherapy has led to survival of about 90 % in the present era [3]. This was also the first solid tumour where the role of adjuvant chemotherapy was established. Large randomized controlled trials by various collaborative groups, including the National Wilms Tumor Study Group (NWTSG), The International Society of Pediatric Oncology (SIOP) and the United Kingdom Children's Cancer Study Group (UKCCSG) have facilitated WT treatment to be customized to minimize morbidity for low-risk disease and to maximize the oncologic outcome for high-stage, high-risk patients. Consequently, the outcome for patients with Wilm`s tumour has improved remarkably in the past few decades. There is paucity of data about the epidemiology, pathology, treatment and prognosis of this disease from India. Therefore, we report in the present study our experiences of the outcome of multimodality treatment of Wilms' tumor at a tertiary care centre.

II. Methodology

All the children were diagnosed with Wilm`s tumour at our centre by imaging {Abdominal ultrasonography/Doppler ultrasonography/Contrast-enhanced computed tomography (CECT), histopathology were included in the study. Doppler ultrasonography was performed to detect tumor infiltration of the renal vein and inferior vena cava, and to assess patency of blood flow. Contrast-enhanced CT was used to further evaluate the nature and extent of the mass. The line of treatment for this tumor at our institution was based on NWTS IV protocol. This regime included upfront surgery; postoperative histopathological confirmation of diagnosis, followed by radiotherapy and chemotherapy based on stage of the disease. After completion of treatment, the follow-up was 3-monthly for the initial 3 years and annually thereafter. Every follow-up visit consisted of clinical examination, abdominal ultrasound and chest radiograph.Overall survival (OS) is defined as time from diagnosis to mortality for all-cause of deaths. Patients still alive were evaluated at the date of last follow-up. Event-free survival (EFS) was calculated from diagnosis to relapse or death; patients who experienced no event were censored. OS and EFS were calculated using the Kaplan–Meier method. Hazard ratios (HR) and 95% confidence intervals (CIs) were estimated using Cox proportional hazards model. Wilcoxon rank sum test was used to compare medians.

III. Results

We retrospectively reviewed all the patients treated at our centre for Wilms tumour from 2005 to 2014. Patients lost to follow up for a minimum of 3 years were excluded from the study. A total of 63 patients were treated, of these only 48 patients were followed until the end point. The age of the patients ranged from 7 months to 7 years (mean, 3.1 years). Forty patients (59%) were males. Majority of patients (66 %) had an abdominal mass noted by parents; other symptoms and signs included abdominal pain in 17 patients (36 %), haematuria in 3 patients (7.3 %). One patients had bilateral disease.Only four patients had undergone surgery prior referral to our centre. Thirty three patients had Wilm`s tumour of favorable histology, 6 had anaplastic tumours, 2 had clear-cell sarcoma (CSS), and 2 had rhabdoid tumours. In 5 children with WT, the histology was unclassified with regard to the degree of anaplasia. of the 47 patients with unilateral disease, 44 children underwent total nephrectomy and 3 children underwent partial nephrectomy (3 horse shoe kidneys underwent partial nephrectomy). In one child of bilateral affection, bilateral limited renal resection was done at our institute. Postoperative tumour-bed radiotherapy was given to 18 children with stage III±V disease and 11 with stage I±II disease. The indications in the latter were anaplastic tumours, surgical spillage of tumour, or tumour adherence to surrounding structures noted at nephrectomy.

The median follow-up was 42 months (range 36–84 months). The estimated 5 years event-free survival was 83.3 % and overall survival was 85.2 % (Fig 1). A total of 10 events occurred in follow up (seven had progressive disease, two relapsed) with majority of them presenting with pulmonary metastases. Of these ten patients, seven patients abandoned treatment (lost to follow-up) and the remaining three were given treatment in the form of chemotherapy (NWTS-R protocol).

Stage at Presentation	No of Cases
I	13 (27%)
II	8 (16%)
III	18 (37%)
IV	8 (15%)
V	2 (5%)

Table 1: Stage at Presentation :

Histology	No of cases
Favourable histology	33
Anaplasia	6
clear cell sarcoma	2
Rhabdoid	2
Unclassified	5

Table 2 Histology of the tumour :

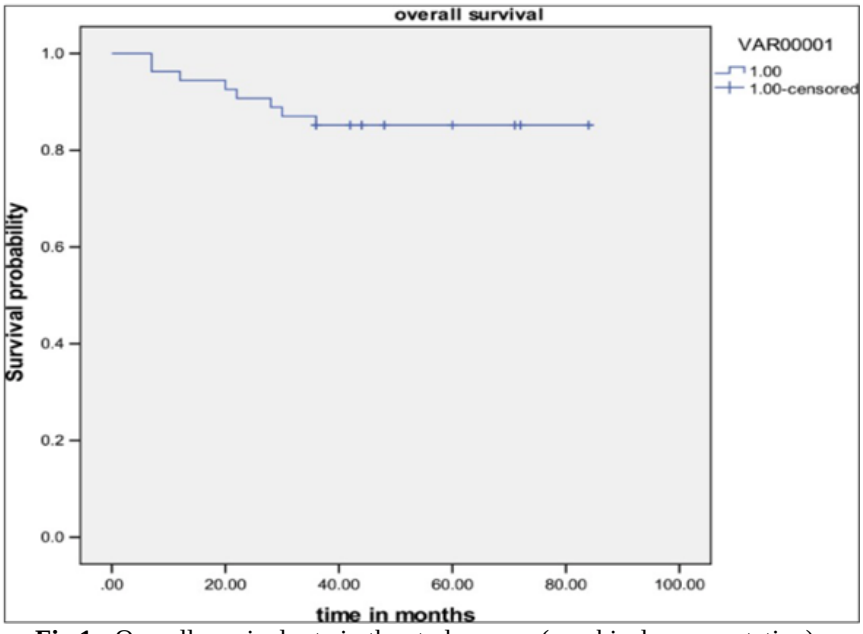

Fig 1 : Overall survival rate in the study group (graphical representation).

IV. Discussion

WT is the most common renal tumor of infancy and childhood. Its incidence is one per 10,000 children under the age of 15 years worldwide [5]. Multimodality treatment has resulted in a significant improvement in the four-year survival, from approximately 30% in the 1930s to more than 85% in the modern era [4].The overall prognosis in Wilm`s tumour remains poor in developing countries. The distribution of the stages in the present series, however, differs from reports from the developed countries [5,6].While 9 of our patients are currently on treatment for primary or recurrent disease, only 39 of 48 are disease-free. Socio-economic factors, reach and availability of tertiary centres, contributed to late presentation and non- compliance with therapy, which are the main reasons for the poor outcome. While total nephrectomy was usually performed for unilateral tumours and on the side with the larger tumour in bilateral cases, kidney-sparing surgery gave very good results in fused kidneys and bilateral disease [7]. Results with renal- sparing surgery for 2 other unilateral tumours gave moderate success, and total nephrectomy might have been more appropriate [4, 8, 9]. The risk factors related to tumor relapse that have been identiedare intraoperative spillage of tumor cells and high-risk stage.Despite advances in adjuvant therapy, surgical extirpation remains an important modality of management for recurrent Wilm`s tumour.

V. Conclusion

In conclusion, factors such as late presentation, poverty, poor compliance to treatment, and lack of multidisciplinary collaboration remain major challenges in managing WT patients in a developing country [10,11,12]. Slightly lower survival rates than those of the NWTS- 3, especially for patients in stages III and IV, were ob- served in our study. Higher clinical stage, an increased number of those with unfavorable histology, tumor spillage or rupture during surgery, nonstandardized therapy of the primary or recurrent tumor, and premature stopping of treatment were the important factors that influenced the outcome of WT at our medical centre. Tackling these issues would further improve outcomes of Wilm`s tumour patients.

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