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Full Length Article

Wilms tumor in childhood: Single centre retrospective study from the National Institute of Oncology of Rabat and literature review

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ABSTRACT

Background: Wilms tumor is a very common renal malignancy in children. Prognosis has been improved dramatically during the last few decades because of multimodal treatment and successful sequential studies. Through a retrospective study conducted in the National Institute of Oncology of Rabat, concerning children with Wilms tumor treated following the International Society of Pediatric Oncology protocol (93-01) between 2005 and 2010, we report the experience of our institute in treatment of this malignancy. We analyze also the clinicopathologic and therapeutic aspects impacting the outcome results and compared to literature data.

Results: Fifty-two patients with Wilms tumor treated in the department of radiotherapy after receiving chemotherapy and surgery at the department of hemato-oncology in children hospital of Rabat were enrolled. The main characteristic was the high prevalence of locally advanced and metastatic stages (32.6% of stage IV). With a median follow up of 54.8 months [20–79], we observed a complete response in 32 cases (61.5%), local recurrence in only one case (1.9%), metastatic relapse in 3 cases (5.8%), both local and metastatic recurrence in 3 cases (5.8%) and disease progression in 8 cases (15.4%). The mean duration of overall survival was 91.2 months. The estimated 2-year and 5-year overall survival were 78.7% and 70.1% and for metastatic patients 68.8% and 62.5% respectively. At univariate analysis several parameters were tested for survival, but only age, anaplasia, lymph node involvement, type of metastasis and response to treatment were found to significantly impact the overall survival. Outcome was better for localized tumors (stage I, II and III) compared with disseminated tumors (stage IV and V) combined. Also a better survival rate was found in the low and intermediate risk group compared to high risk, but not statistically significant.

Conclusion: The relatively low outcome found in this series compared to literature can be mainly explained by the higher prevalence of metastatic disease compared to other series, but also by diagnosis and therapeutic delay, more likely because of bad socioeconomic conditions and lack of coordination between different operators. However, our results are nevertheless comparable to maghrebian series. Our department has established many procedures for improving the outcome and further studies are necessary to evaluate their efficiency.

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1. Background

Wilms tumor or nephroblastoma is the most common renal malignancy in children and the sixth pediatric cancer. It represents 5–10% of solid tumors and 1/3 of embryonic neoplasia [1–3]. The last few decades have seen a dramatic change in the prognosis of this disease. Indeed, survival rates have improved to a current five-year overall survival rate of 90%, whereas it was only 30% with surgery alone and only 47% with the combination radiotherapy-surgery [4–6]. This improvement in outcome is due mainly to multimodal treatment regimens (chemotherapy, surgery, and radiation) based on sequential clinical trials.

The target of this survey is to report the epidemiological, clinical and therapeutic features of patients treated for nephroblastoma in the department of radiotherapy in the National Institute of Oncology (NIO) of Rabat, Morocco with a summary review of literature data.

2. Methods

2.1. Data collection

This is a retrospective study of 52 cases of Wilms tumor (WT) treated in the department of radiotherapy during a period of 6 years (2005–2010), after receiving chemotherapy and surgery at the department of hemato-oncology in children hospital of Rabat.

Analysis of clinical features, pathological findings, imaging studies, different therapeutic modalities and outcome was achieved based on data from both hospitals' cancer registry of the NIO and the children hospital of Rabat.

We got permission to access and use data from the department head of cancer registry of Rabat. We also obtained verbal consent from all parents or legal guardians of the patients included in this study.

Patients were treated according to SIOP protocol 93-01 [5]. This regimen included upfront chemotherapy followed by surgery and then postoperative histopathological confirmation of diagnosis. A risk-based therapy (including adjuvant chemotherapy and radiotherapy) is assigned based on results of the initial staging and histological group disease.

Histological subtype determining histoprognosis group and staging (I, II, III, IV or bilateral) were assessed according to the revised SIOP Working Classification of Renal Tumours of Childhood (2001) [7,8].

2.2. Statistical analysis

Several factors were tested for outcome: age, gender, stage at presentation, histological sub-type, risk group, lymph node status, metastasis and response to treatment.

Median follow-up time was defined from the date of diagnosis to date of death or date last follow-up.

Overall survival (OS) is defined as time from diagnosis to mortality (for all-cause death) and patients still alive were censored at the date of last follow-up.

Survival rates were evaluated for all patients using the Kaplan Meier curve (SPSS 13.0) and compared by log-rank test with a statistical significance of 5%.

3. Results

Our series enrolled 52 patients, with a median age of 4 [3–5] years (range from 17 months to 14 years), sex ratio (F/M) was 0.9 (Table 1). A single case of hemihypertrophy was found. However, no other associated congenital syndrome has been noted.

Table 1
Clinico-pathological and evolutionary features.

Clinico-pathological and evolutionary features		Number of cases (%)
Age	<5 years	29 (55.8)
	>5 years	23 (44.2)
Gender	Male	27 (51.9)
	Female	25 (48.1)
Stage at presentation	I	1 (1.9)
	II	4 (7.7)
	III	28 (53.9)
	IV	17 (32.7)
	V	2 (3.8)
Histology	Low risk	2 (3.8)
	Intermediate risk	35 (67.3)
	High risk	15 (28.8)
Anaplasia	Absent	47 (90.4)
	Diffuse	4 (7.7)
	Focale	1 (1.9)
Metastasis at presentation	Lung only	12
	Liver only	2
	Lung + Liver	4
Response to treatment	Complete response	32 (61.5)
	Local relapse	1 (1.9)
	Metastatic relapse	3 (5.8)
	Progression	8 (15.4)
	Local and metastatic relapse	3 (5.8)
	Total	47 (100)

The median time to medical visit was 30 days [15–60], and 84% of children consulted within three months from the beginning of their symptoms. Clinical features were dominated by abdominal mass in 96.2% of cases associated with pain in 30.8% of cases, hematuria in only 19.2% (10 cases) and fever in 13 cases (25%). However no case of hypertension or acute abdomen symptoms was noted.

Diagnosis was based mainly on the clinical and imaging data. Indeed, abdominal ultrasound made in all patients and coupled to CT scan in 82.6% of cases have shown the tumor usually as a large renal mass, greater than 10 cm of diameter in 80.8% of cases and well limited. They have found thrombosis of inferior vena cava (IVC) in 10 cases (19.3%), combined to an intra-cardiac thrombosis in 5 cases which was shown by a chest CT scan.

The two imaging modalities have allowed evoking diagnosis of WT in 88.4% of cases. However, in 6 atypical cases (11.5%), diagnosis has been assessed histologically by fine needle aspiration and/or percutaneous cutting needle biopsy for the strong suspicion of neuroblastoma in 5 cases and of a retroperitoneal abscess in the sixth one. Both CT scan and chest radiography have been used for screening lung metastases. The approach used in our institution is the routine use of chest radiography (performed in 96% of children), supplemented by chest CT scan (performed in 36.5% of cases), only in cases of abnormal chest radiography. This assessment revealed 18 metastatic cases (34.6%) at diagnosis (67% in the lung only, 11% in the liver only and 22% in both lung and liver).

Fifty children (96%) received preoperative chemotherapy according to SIOP protocol 93-01 [5], which allowed the regression of 50% or more of the tumor mass in 43 cases (82.7%).

Surgery was performed in 96% after neoadjuvant chemotherapy, except for 2 patients who have undergone initial surgery (because the diagnosis of WT was missed). The surgical treatment consisted on radical nephrectomy in all cases associated with partial contralateral nephrectomy in one case of bilateral nephroblastoma, and a metastasectomy was achieved in two cases (liver in one case and lung in the other one). Lymph node dissection has been performed in 15 cases (28.8%) and revealed metastatic nodes in 9 of them (17.3%).