



Original articles

Clinical characteristics and outcome of Wilms tumors with a favorable histology in Japan: a report from the Study Group for Pediatric Solid Malignant Tumors in the Kyushu Area, Japan

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Abstract

Background/Purpose: Since 1996, the standard treatment of Wilms tumors in Japan has been based on the regimen of the Japanese Wilms Tumor Study. However, in Japan, there have been no reports about Wilms tumors that analyzed the clinical features and patient outcome in a large series until now. This study aims to assess the clinical characteristics of patients with Wilms tumor with a favorable histology from a retrospective standpoint in the Kyushu area in Japan and, furthermore, to analyze the historical changes of clinical features and outcome from the 1980s to the 1990s.

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Methods: Between 1982 and 1996, 90 cases of Wilms tumors with a favorable histology were registered in the Kyushu area. Regarding the clinical feature and outcome, they were divided into 2 groups (group A, 1982-1989, n = 50; group B, 1990-1996, n = 40). The outcome was analyzed based on the 5-year overall survival rate.

Results: The clinical features (age, sex, initial symptom, location, stage) demonstrated no definite differences between group A and group B. Regarding the operation, the rate of an initial complete resection in the early stages was significantly higher in group B than in group A. All stage V cases in group B underwent a bilateral tumor biopsy instead of a radical nephrectomy as the initial operation. The 5-year overall survival rate throughout the whole period was 87.8%, whereas the rates were 84.0% for group A and 90.0% for group B ($P = \text{NS}$), respectively. Of particular note, the outcome of patients with stage I and stage V in group B substantially improved in comparison to that in group A. However, in advanced cases, no significant improvement in the outcome was noted.

Conclusions: This is the first report about the clinical features and outcome for Wilms tumors with a favorable histology in Japan from the 1980s to the 1990s. The present study suggested that in the early-stage cases, an initially complete resection followed by standard postoperative chemotherapy substantially improved the outcome of the patients in group B. In the stage V cases, the performance of renal salvage surgery may have positively contributed to the improvement in the outcome in group B. However, in the advanced stage cases, no definite improvement was noted. In the future, an improved efficacy of the treatments for Wilms tumors based on the standard protocol established by the Japanese Wilms Tumor Study in 1996 is expected in Japan.

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Wilms tumor is the most common malignant neoplasm of the kidney observed in children. The multimodality treatment for such tumors, including surgery, chemotherapy, and radiotherapy, has undergone refinement in recent years. The National Wilms Tumor Study (NWTs) was established in 1969 as a central organization for the treatment of Wilms tumor in the United States and Canada. Standard protocols were established according to the clinical stage and histology. Some modifications have been made over the past 35 years, and NWTs-5 is continuing its activities up to the present. On the other hand, in Europe, the International Society of Pediatric Oncology (SIOP) was the central organization for the treatment of Wilms tumor. The NWTs recommends an initial radical operation, followed by adjuvant chemotherapy and radiotherapy, whereas the SIOP emphasizes the efficacy of pre-nephrectomy treatment.

In Japan, about 40 to 50 cases of Wilms tumor occur every year. The treatment regimen in Japan depended on the decision of each institution until the Japanese Wilms Tumor Study (JWiTs), which was based on the NWTs-4 regimen, was established in 1996. In Japan, there have so far been no reports on Wilms tumors that analyzed the clinical features and patients outcomes in such a large series until now. In the current study, we retrospectively assessed the clinical characteristics of patients with Wilms tumor with a favorable histology from the Kyushu area in Japan and, furthermore, analyzed the historical changes in the clinical features and outcome from the 1980s to 1990s.

1. Material and methods

From 1982 to 1996, 98 patients with Wilms tumor from the Kyushu Area of Japan were registered by the Committee

for Pediatric Solid Malignant Tumors. Of the 98 patients, 90 had a favorable histology, whereas 8 had an unfavorable histology. In this study, the patients with a favorable histology were divided into 2 groups (group A, 1982-1989, n = 50; group B, 1990-1996, n = 40) to assess the historical changes of the clinical features and outcome in the patients with Wilms tumor. They were analyzed for the following clinical characteristics: age, sex, initial symptoms, location, stage, treatment, and prognosis. The staging was performed according to the classification system developed by the committee for malignant tumors of the Japanese Society of Pediatric Surgeons. The 5-year overall survival distributions were estimated using the method of Kaplan and Meier. The survival time was defined as the time from the initial diagnosis to death. The log-rank test was used to assess any statistical significance in the prognostic factors.

2. Results

2.1. Clinical features and treatments of the 90 cases of Wilms tumors

The mean age of the patients was 3.0 ± 3.1 years in group A and 2.6 ± 1.9 years in group B. There was a higher percentage of boys in both groups. Regarding the initial symptoms, abdominal tumor was the most common symptom in both group A (74.0%) and group B (64.0%). The tumor location was more frequently observed on the left side both in group A (53.0%) and group B (65.6%). For the bilateral cases, 3 cases (7.5%) were observed in group A, and 6 cases (12.0%) were observed in group B. Regarding the clinical stage, 23 were stage I, 13 were stage II, 5 were stage III, 6 were stage IV, and 3 were stage V in

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