

Adult Wilms' Tumor: Review of Literature

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Introduction

The most common renal tumor in adults is renal cell carcinoma, wilms' tumor is extremely rare in adults, representing only 0.5% of all renal neoplasms, 240 cases in adults have been reported [1]. The true incidence of adult wilms' tumor is somewhat uncertain because of confusion in terminology and difficulties in clinical and pathological differential diagnosis [2]. Kilton et al. selected 35 real adult wilms' tumor complying with 6 diagnostic criteria of 192 documented cases [3]. Patients with stage III, IV are reported to account for more than 50% of most adult series [4]. Until recent, exact data on prognosis and late effects are not available [5]. Prognosis for adult patients with unfavorable histology and stage IV disease (hematogenous metastasis) is poor despite aggressive multimodal therapy [6]. There is no standard therapy for treating patients with adult wilms' tumor. Some authors suggest an aggressive treatment regardless of the stage, but a therapeutic procedure based on the NWTs with different treatment depending on tumor stage also has been recommended [7].

Abbreviations

NWTS-G3: National Wilms' Tumor Study-Group 3; SIOP: The International Society of Pediatric Oncology

Discussion

The diagnostic criteria necessary for adult wilms' tumor suggested by Kilton et al [3] are, primary renal neoplasm in age group of > 15 years with histologic features of embryonic glomerulo-tubular structure with immature spindle or round cell stroma and no areas of tumor diagnostic of renal cell carcinoma [3]. Wilms' tumor whether it occurs in childhood or in adult life doesn't contain the mature glandular elements of renal carcinoma, but the findings of abortive glomerular or tubular (epithelium) and embryonic (blastema) structures mixed with immature mesenchymal cells (stroma) are pathognomonic [8]. Classic wilms' tumor is composed of three cell types, blastemal, stromal and epithelial, which can be present in various proportions [9]. The NWTsG classifies wilms' tumor based on the

presence of anaplasia, the revised SIOP histologic classification divides wilms' tumor into three risk groups:

- low risk (cystic partially differentiated nephroblastoma)
- intermediate risk (regressive, epithelial, stromal, mixed, or focal anaplastic nephroblastoma)
- high risk (blastemal or diffuse anaplastic nephroblastoma) [10].

Clear cell sarcoma of the kidney and malignant rhabdoid tumor of the kidney, initially believed to belong to the unfavorable histology family of wilm's tumor, are now considered distinct tumor types [11]. The differential diagnosis of an adult wilms' tumor with mainly epithelial differentiation includemetanephric adenoma. A predominant blastemicwilms' tumor has a strong resemblance to small, blue, round cell tumors which commonly include lymphoma, peripheral neuroectodermal tumor and rhabdomyosarcoma, and rarely metastatic small cell tumor from lung, immature teratoma, and primary renal cell sarcoma. Extensive search for any other components is needed as a poorly differentiated renal carcinoma can have large sarcomatous areas resembling blastema [3]. This disease is difficult to differentiate from renal cell carcinoma based only on imaging technique, but ultrasound observation of rapidly growing abdominal mass, with heterogenous contrast uptake, and surrounded by a pseudo capsule on CT is suggestive of wilms' tumor [1].

The clinical presentation of adults with wilms' tumor differs from that of children, the main symptoms of adults is flank pain, and the majority of them have a history of weight loss and of a sudden drop in performance status [12]. A fine needle or true-cut biopsy may facilitate histologic diagnosis in cases of primarily inoperable tumors or stage IV diseases, this approach was carried in some patients and helped to stratify farther treatment, preoperative chemotherapy could therefore be initiated, and this resulted in a regression of the tumor [13]. In Europe percutaneous needle biopsy generally is used in children to assess the nature of massive renal tumors, if performed before nephrectomy, it has been shown to be effective

in approximately 90% of cases [8]. There are 10 cases of extra renal wilms' tumor in adults documented in the literature. Four of them were in the retroperitoneal region, two cases each in the ovary and endometrium and one case in ovotestis and prostate [3]. The prognosis of adult wilms' tumor is poorer than that of renal adenocarcinoma, in reported cases CT scan showed predominantly hypodense renal mass unlike typical mixed density mass in renal cell carcinoma [14].

The prognosis of unfavorable histology is very poor in adult wilms' tumor, but that of a low stage or the epithelial favorable histology is as, in children (relatively good) [15]. A definitive treatment plan has not been established yet because of the rarity of these tumors in adults. After surgical removal multimodal therapy should begin immediately for long lasting, complete remission in spite of multimodal approach similar to childhood wilms' tumor, the treatment outcome in adults is disappointing [4]. The majority of adult wilms' tumor patients die of metastatic disease [5]. Some authors have suggested that the classic three-drugs regimen used in childhood, may not be effective in adults, indicating the need for new drugs combinations. However, other cases series, have not confirmed this lack of effectiveness, they concluded that adult wilms' tumor has a worse prognosis compared with wilms' tumor in children and should be treated aggressively with 3 drug chemotherapy and radiotherapy to the tumor bed (4500 cGy), regardless of the stage of disease [4].

The NWTSG has recommended preoperative chemotherapy under certain circumstances, including the occurrence of wilms' tumor in a solitary kidney, bilateral wilms' tumor, tumor in a horse shoe kidney, tumor thrombus in the IVC above the level of hepatic vein, and respiratory distress resulting from the presence of extensive metastatic tumor [16]. In 2004, Reinhard et al. [13] reported their experience with 30 cases of adult wilms' tumor. A complete remission was achieved in 24 of their patients. Event free survival was 57% and overall survival was 83%, they concluded that adults can be cured in a high percentage by a multimodal treatment according to pediatric protocols [13]. For patients with recurrent disease, encouraging results recently have been reported using cisplatin or cisplatin compounds, etoposide, and ifosfamide, likewise, in a phase II study of the French society of pediatric oncology, the combination of carboplatin and etoposide was very effective in 26 refractory or recurrent wilms' tumor patients, with 8 patients in continuous complete remission at median follow up of 40 months [17]. Complete responses also were achieved with cisplatin and etoposide in adult patients with progression disease after conventional chemotherapy with dactinomycin, doxorubicin, and vincristine [8].

Ifosfamide is an active drug as well, with 50% rate of objective responses in patients with recurrent wilms' tumor who had failed conventional combination chemotherapy [18]. Conversely, the three classic drugs (vincristine, dactinomycin, doxorubicin) used for the treatment of wilms' tumor in children

have failed to improve long term disease survival in adult patients, for this reason, some authors have suggested treating patients with cisplatin and etoposide, which are currently employed to improve the prognosis of recurrent wilms' tumor in adults [18]. Therefore, also considering the advanced stage of the disease (the presence of lung and liver metastasis), some authors decided to use the most active drugs according to the following regimen : epirubicin and ifosfamide and carboplatin [19].

Conclusion

Although the overall survival rate in children with wilms' tumor about 90% the prognosis for adult patients with nephroblastoma remains less favorable. Adult with wilms' tumor reported in the National Wilms' Study (NWTSG) from 1968 to 1979 and from 1979 to 1987, showed an improvement in the overall 30 years survival rate from 24 to 67% [20]. For patients with recurrent disease, long term remissions have been achieved using high dose chemotherapy, radiotherapy and allogeneic bone marrow transplant or combination chemotherapy with cisplatin and etoposide [21,22]. Because the adult wilms' tumor represents a rare presentation, the retrospective reviews of case reports and limited series can provide some guidelines toward effective rationale therapy [7].

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