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UROLOGICAL ONCOLOGY

A role for pediatric oncologists/urologists?!

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Like testicular cancer in adulthood, the Wilms' tumor or nephroblastoma in childhood is an example of a curable malignant disease. The review by Huszno et al. in this Journal gives an overview concerning diagnosis and treatment of adult Wilms' tumors [1].

This tumor entity was first described in 1814 by Rance and in 1899 Max Wilms, a German surgeon, described this tumor in detail [2]. Beckwith credited John Hunter with preparing perhaps the earliest specimen between 1763 and 1793 which is now in the collection of the Hunterian Museum of the Royal College of surgeons in London [3, 4].

At the beginning of the last century, the prognosis was poor and the mortality was over 90%. With the introduction of radiation [5] – and later chemotherapy [6] the prognosis improved to almost 90% for all children today [7]. Wilms' tumor in childhood is a paradigm for a successful multimodal cancer treatment. Today molecular marker may be additional prognostic factors for a risk–based therapy [8] e.g. the loss of heterozygosity at chromosome 1p and 16q as suggested by the NWTS–4 study [9].

Up to 2% of these tumors were found in adults [10, 11] with more than 300 cases reported in the literature [11, 12]. In the SEER – Database almost 6.5% (152/2342 patients) were adults [11]. The presentation of adult Wilms' tumor differs from those in childhood with flank pain and loss of weight [12, 13].

As in childhood the prognosis of Wilms' tumors in adults improved over the last 50 years. In 1982 the 3–year survival rate in 31 patients was 24% [14]. In 1990 the 3–year survival rate was 67% in 27 adults, excluding those with anaplastic tumors it was 79% [15]. In 2004 the 4–year median survival in 30 patients was 83% in the SIOP 93–01 study [13]. As these survival rates are somewhat lower compared to children treated during the same period of time, the SEER database demonstrated even a higher difference between children and adults. The 5–year overall survival rate was 88% for the pediatric group and 69% for adults [16]. One reason these differences may be, that adult urologists / oncologists are unfamiliar with the specifics of Wilms' tumors pediatric treatment protocols as well as the difficulties to diagnose a Wilms' tumor (there is rarely a central pathology review performed in adult patients). This delays diagnosis and treatment or the tumor is even misdiagnosed. Izawa et al. presumed that lymph node dissection / sampling may also be a factor. In adults, lymph node dissection in radical nephrectomies is rarely performed [17], in the SEER–Database, however, lymph node sampling was performed in 42.1% [16]. In children, lymph node sampling is also not always adequately performed, leading to an under–staging and therefore under–treatment with the risk of a relapse [18].

In conclusion, adult urologists / oncologists have no standard treatment as do pediatric urologists and pediatric oncologists. In most adult patients the diagnosis is unexpectedly following nephrectomy for presumed renal cell carcinoma. If there is suspicion of a Wilms' tumor, a central pathology review and staging according to the pediatric Wilms' tumor protocol should be performed. In those adults, with a confirmed Wilms' tumor, pediatric oncologist/urologist should immediately be involved in the further treatment of the patient. Only a standardized multidisciplinary and multimodal treatment can improve the survival rate in this group of patients with a potential curable malignant disease.

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