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Case Presentation
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Wilms Tumor in an Adult Male

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Abstract

Wilms' tumor (nephroblastoma) is the most common renal tumor in children. Wilms' tumor in adults is extremely rare and is presumed to have a poorer prognosis than in children. It is difficult to differentiate adult Wilms' tumor from renal cell carcinoma based on radiological findings alone. The diagnosis in adults is often made following nephrectomy for a presumed renal cell carcinoma. Because of the paucity of literature concerning Wilms' tumor in adults, there are no standard protocols for the management and therefore managed on the same lines of that of a Wilms' tumor in a child. Herein, we report the case of adult Wilms' tumor in a 22-year-old man, which was diagnosed unexpectedly the following nephrectomy for presumed renal cell carcinoma.

Keywords: Nephroblastoma; Adults; Diagnosis; Treatment

Introduction

The most common kidney tumor in adults is Renal Cell Carcinoma (RCC). Wilms' tumor is rare in subjects who are older than 16 years. Only 3% of Wilms' tumors are reported in adults, which explain the difficulties in diagnosis and treatment of this tumor entity in this age group [1]. Most of the reports in the literature are mainly single case reports of adult patients, and studies on larger patients series are rare [1-4]. Until recently, a standardized treatment for adults with Wilms' tumor has been missing, and exact data on prognosis and late effects are still not available. Primary radical nephrectomy followed by chemotherapy has been the treatment strategy for patients older than 16 years.

Reinhard et al. [2] reported on 30 patients older than 16 years (range 16 to 62 years, median, 25.4 years) that were found to have Wilms' tumor. They were treated according to the pediatric protocol and were analyzed for clinical presentation, stage distribution, and prognosis. Ten patients (33%) had metastatic disease at the time of diagnosis (liver, four patients; lung, three patients; liver and lung, three patients). The local stage distribution showed a predominance of higher stages (stage I, eight patients; stage IIN-, three patients; stage IIN+, four patients; stage III, 15 patients). Histologic studies revealed intermediate-risk in 23 of 30 tumors; two tumors were classified as high-risk, and three tumors were clear-cell sarcomas. Two of 30 patients showed a nephroblastoma and a renal cell carcinoma simultaneously in the same kidney. Complete remission was achieved in 24 patients; four patients relapsed after complete remission, and three of them reached a second remission with further treatment. Event-free survival was 57%, with overall survival of 83% (median observation time, 4 years). The study concluded that adults could be cured by a multimodal treatment in a high percentage of patients, according to pediatric protocols. Toxicity was higher than in children but acceptable in view of the high remission rate. We report a case of Wilm's tumor diagnosed and managed in a 28-year-old male.

Case Presentation

A 22-year-old adult male presented with symptoms of pain in loin, fever and one episode of frank hematuria. A clinical diagnosis of ureteric stone was made and the patient underwent ultrasonography and Computed Tomography (CT) imaging (Figure 1a & 1b). Computed tomography showed a huge space of lesion occupying the middle portion of the kidney as well as the lower pole measuring 7 cm × 8 cm. Based on the CT findings diagnosis of renal cell carcinoma was made and the patient underwent Rt. Sided radical nephrectomy.

Histopathological examination revealed a neoplasm with tubular and papillary structures, the stroma was composed of myofibroblastic cells few vascular emboli were seen and foci of necrosis noted. The tumors showed evidence of blastemal tissue in focal areas (Figure 2). The patient has

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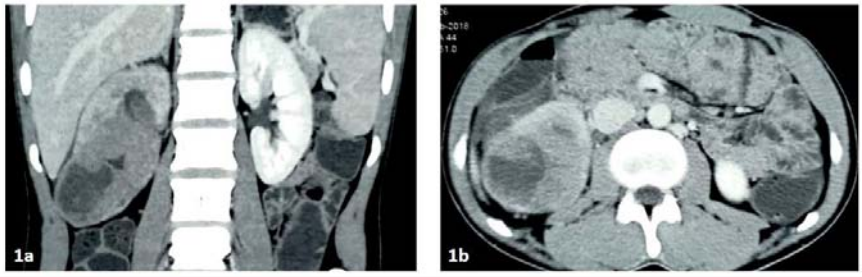

Figure 1a: Computed tomography showing a huge space of lesion occupying the middle portion of the kidney.
Figure 1b: The lower pole measuring 7 cm × 8 cm.

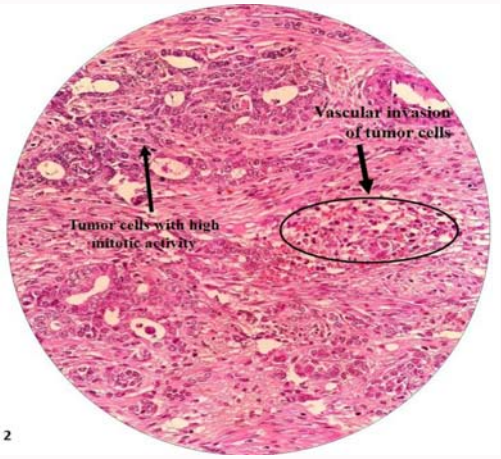

Figure 2: 40X H&E staining shows malignant blastemal cells in a small focal area. Vascular emboli noted and the stroma is composed of myofibroblastic cells.

been put on post-operative chemotherapy similar to the treatment of Wilm's tumor and is on close follow-up for the past 15 months.

Discussion

The clinical presentation of adults with Wilms' tumor differs from that of children. The main symptom of adults is flank pain, and the majority of them have a history of weight loss and of a sudden drop in performance status. This observation is in accordance with other reports on symptoms of Wilms' tumor [2-5]. The tumors are usually asymptomatic in children or present with a painless swollen abdomen. In contrast to the pediatric population, the majority (23 of 30) of the adult patients were operated primarily in the SIOP studies [2]. It is not possible to achieve a safe diagnosis by imaging studies alone in this patient group. Even in the concept of the SIOP Wilms' tumor protocol, patients older than 16 years primarily undergo surgery of the tumor [3].

As Wilms' tumor is a rare entity in adults, its occurrence is hardly suspected by both the Urologists as well as the Pathologists. RCC is the most common tumor in this age group, and in some cases, histological differentiation of nephroblastoma and RCC is also difficult [2]. The difficulties incorrect diagnosis may lead to inappropriate or delayed treatment and may thus contribute to a poorer prognosis in these patients. Histological subtypes do not differ from pediatric tumors [2]. There is no higher rate of high-risk tumors. The biology of the tumors seems to be identical to those in children. The tumors tend to respond to the preoperative treatment protocol and to have a good prognosis if they are treated in the same manner [2].

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