Extrarenal adult Wilms' tumor case report

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Abstract: Wilms' tumor is the most common primary renal malignancy in children. Adult presentations of Wilms' tumor are very rare, and extrarenal adult Wilms' tumors even more so, with only a few reported cases in the literature. Adult patients have traditionally had a worse prognosis than children, due to presentation with advanced stages of disease, diagnostic uncertainty, and limited treatment experience. When treated with multimodal therapy in accordance with the established protocols for pediatric Wilms' tumor, adult patients are able to achieve good outcomes. We report the case of an adult female with an extrarenal Wilms' tumor, and review the literature surrounding this uncommon disease entity.

Keywords: Case report; extrarenal Wilms tumor; adult Wilms tumor; Wilms' tumor; nephroblastoma

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Introduction

Although Wilms' tumors (WTs) is the most common renal malignancy in childhood, presentation in adults is very rare (1-4). Extrarenal presentation of WTs is also an unusual presentation, with studies often limited to case reports (5,6). Due to the rarity of extrarenal adult WTs, treatment guidelines are limited (7). Current guidelines for extrarenal adult WTs suggest similar treatment regimes as childhood and renal WTs; however, various studies question the effectiveness of these guidelines as external adult WTs are an abnormal and rare variant of WTs (1,5). We report a case of adult extrarenal WT, and review the literature on management. We present the following case in accordance with the CARE Guideline.

Case presentation

A 50-year-old female presented with abdominal discomfort and distention. An abdominal/pelvic CT revealed a 30 cm abdominal mass (*Figures 1*,*2*). A percutaneous biopsy was consistent with Wilms' tumor, which is extremely rare in adult patients and in extrarenal location. Neoadjuvant dactinomycin based chemotherapy resulted in a 30% regression in tumor size. Subsequent surgical resection was successful in removing this over 20 cm (7.3 lb) Wilms tumor, with negative surgical margins (*Figure 3*). Gross pathology showed the typical solid and tan appearance, with evidence of cystic changes (*Figure 1B*). On microscopy, a combination of stroma and blastema cells, with epithelial component forming tubules was noted (*Figure 1C*,*D*). Despite proceeding with adjuvant chemotherapy, the patient recurred and succumbed to disease.

Discussion

Wilm's tumor (WT) is the most common primary renal malignancy in children (1-3). Management follows risk stratified protocols set out by the National Wilms' Tumour Study Group (NWTS) in North America and the Society of Paediatric Oncology (SIOP) in Europe. The NWTS prefers a surgery first approach, with subsequent adjuvant

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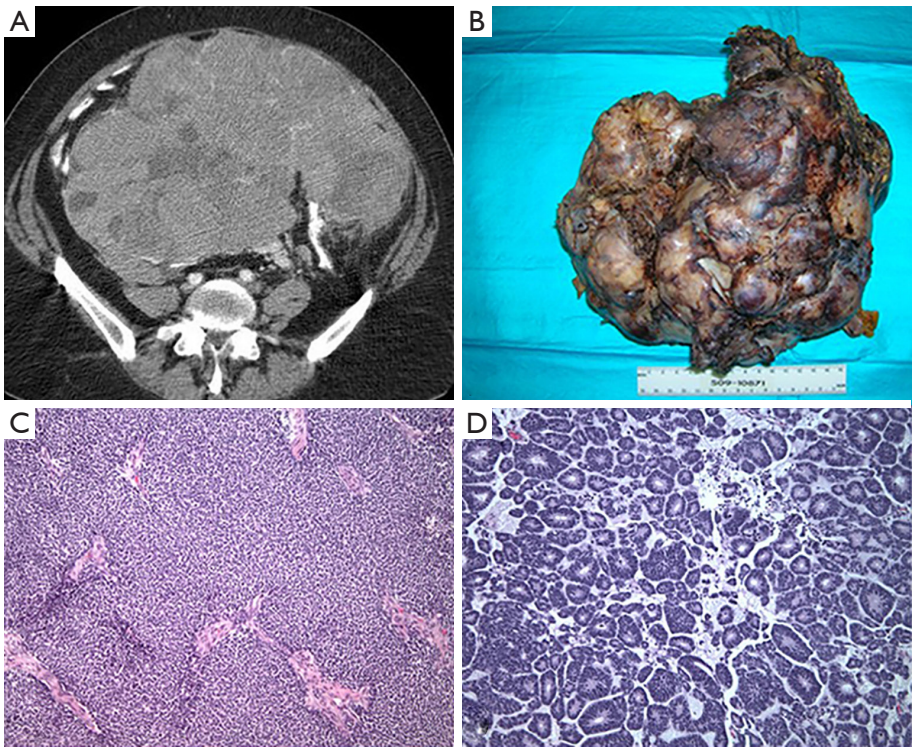

Figure 1 Presentation of the patient's Wilms' displaying (A) abdominal/pelvic CT revealing 30 cm abdominal mass, (B) gross pathology of the tumor, (C) stroma and blastema cells seen on H&E staining at 20× magnification, and (D) epithelial cells forming tubules seen on H&E staining at 40× magnification.

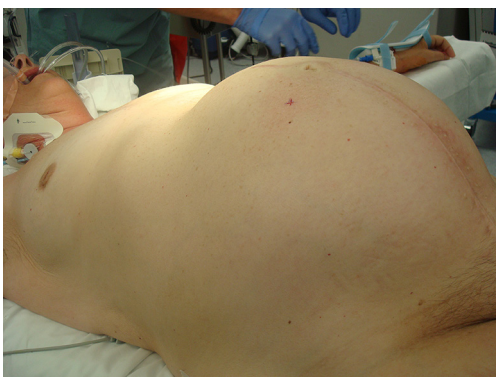

Figure 2 Significant abdominal distension prior to surgical resection.

Figure 3 Fresh surgical specimen of large extrarenal Wilms' tumor.

chemotherapy and radiotherapy based on surgical staging. Conversely the SIOP approach is to give neoadjuvant chemotherapy to all comers, followed by surgical resection, and then adjuvant therapies based on staging. Both groups have demonstrated excellent survival outcomes with protocol driven treatment for pediatric WT.

In adults, WT is extremely rare. Reported incidence is 0.2 per million per year and this comprises 3% of all WT cases (4,8). Extrarenal adult WT is even rarer, with only a small number of reported cases in the literature. These have included tumors arising from the uterus, ovary, testis, and retroperitoneum (6,9-13). Although evidence is limited by

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scarcity of reports, it is felt to behave in a similar fashion to classic WT, and should be treated with the same therapeutic approach (5).

The clinical presentation of WT in adults is distinct from that in children. While children often present asymptomatically, with an enlarging abdominal mass, adults frequently present with flank pain, weight loss, and decreased performance status (2). Histologically, pediatric and adult WT are identical, characterized by the presence of epithelial, stromal, and blastemal elements (14). The diagnosis of adult WT is based on fulfilling the Kilton criteria. These are: (I) primary renal neoplasm, (II) primitive blastematous spindle or round cell component, (III) formation of abortive or embryonal tubular or glomeruloid structures, (IV) no area of tumor diagnostic of renal cell carcinoma, (V) confirmation of histology and (VI) age >15 years (15).

Despite their shared histology, adult WT are traditionally thought to portend a significantly worse prognosis than pediatric WT. The reasons for this are multifactorial. Adult patients tend to present with more advance stages of disease, including higher frequency of metastases (1,2,7,8,14,16). In addition, WT in adults represents a diagnostic dilemma, as the clinical and radiographic features are indistinguishable from more common adult renal neoplasms such as renal cell carcinoma (16). Pretreatment biopsy can assist in making the diagnosis, but is not routinely used in all centers. In our case, it allowed us to diagnose the WT and deliver neoadjuvant chemotherapy, prior to proceeding with surgery.

Further, given its rarity, there is a lack of familiarity with WT amongst adult pathologists and oncologists, which combined with diagnostic dilemma, puts patients are at risk of under treatment from non-compliance with the recommended pediatric protocols (1,17). Finally, even when receiving treatment as per the standard of care protocols, adults tend to experience worse toxicity from the chemotherapy, which can limit treatment (18-20). Notwithstanding these challenges, both the NWTS and the SIOP groups have demonstrated improved outcomes for adult patients with WT through the strict application of the pediatric protocols.

Kalapurakal *et al.* reviewed 23 adults with favorable histology Wilms tumour. Ten patients (43%) had advanced disease (stage III & IV) (16). All tumors underwent centralized pathology review to confirm the diagnosis. All patients were treated with primary nephrectomy followed by adjuvant chemotherapy and in some cases radiotherapy, as per the pediatric NWTS protocols. The 5-year overall survival was 83% and they recommended that therapy in adults with favorable histology WT should be directed according to pediatric NWTS protocols.

Reihard *et al.* reviewed 30 adults with WT treated as per the pediatric SIOP protocols. Of note, 10 patients (33%) had metastases on presentation (2). In this series, the majority of patients started with nephrectomy, although some did first undergo biopsy and neoadjuvant chemotherapy. All patients received adjuvant chemotherapy as per the SIOP risk stratified protocols, and adjuvant radiotherapy was delivered in 46.7% of cases. With a median FU of 4 years, overall survival was 83%, similar to the NWTS results. The authors concluded that adults can be cured in a high percentage by a multimodal treatment according to pediatric protocols.

In summary, we present a case of extrarenal Wilms' tumor in an adult female patient. She underwent percutaneous biopsy of a retroperitoneal mass confirming WT. This was followed by neoadjuvant chemotherapy, complete surgical resection with negative margins, and finally further adjuvant chemotherapy. Despite this, the patient succumbed to the disease. Adult WT is a rare entity which presents with flank pain, weight loss, and decreased performance status. Treatment should follow the established protocols (NWTS or SIOP) for pediatric WT in order to maximize overall survival. With strict protocol adherence, good outcomes can be achieved, even in advanced disease states.

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Footnote

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