Case Report

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Adult Wilms' tumor: Review of literature

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Abstract

Background. The most common renal tumors in adults is renal cell carcinoma. Wilms' tumor in subjects older than 16 years is rare; 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.

Methods. Patient with stage IV adult nephroblastoma with favorable histology was described, current treatment modalities were discussed, and the literature was reviewed.

Results. Nineteen year old female patient is presented with renal mass, abdominal lymphadenopathy, and bilateral pulmonary deposits. Sonar guided biopsy from the renal mass was taken and pathology revealed nephroblastoma. Right nephrectomy was performed and the pathological examination revealed classic histology of nephroblastoma. The case diagnosed as stage IV adult Wilms' tumor with favorable histology. According to National Wilms' Tumor Study Group (NWTS-3), multimodal therapy was initiated immediately after surgery. The patient failed to respond to the first line therapy and died due to disease progression.

Conclusion. Adult Wilms' tumor has no specific guidelines and this may lead to improper or incorrect treatment.

Keywords

Adult Wilms' tumor, nephroblastoma, multimodal therapy

Introduction

The most common renal tumor in adults is renal cell carcinoma. Wilms' tumor, while relatively common in children, is extremely rare in adults representing only 0.5% of all renal neoplasms. Approximately 300 cases of adult Wilms' tumor have been reported in the literature.[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] 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. 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.[2] Prognosis for adult patients with unfavorable histology and stage IV disease (hematogenous metastasis) is poor despite aggressive multimodal therapy.[5] There is no standard therapy for treating patients with adult Wilms' tumor. Some authors suggested an aggressive treatment regardless of the stage, but a therapeutic procedure based on the National Wilms' Tumor Study Group (NWTS) with different treatment depending on tumor stage also has been recommended.[6]

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Case report

Nineteen year old female patient was referred to Clinical Oncology Department, Sohag Faculty of Medicine in December 2008, with right abdominal ill-defined mass, her symptoms starting 6 months prior to referral. The patient complained from abdominal pain, dyspnea, and loss of weight. Physical examination revealed right upper quadrant abdominal mass, not tender, and wheezy chest.

Abdominal computed tomography (CT) showed huge ill-defined mass lesion, which involved the right kidney, measured $8 \times 9 \times 14\,cm^3$, it appeared infiltrating the renal capsule with invasion of the renal vein with further tumor thrombus noted obliterating the proximal extent of inferior vena cava, there were multiple enlarged right renal hilum, and para-aortic lymph nodes (Figure 1). Chest CT showed bilateral pulmonary deposits and enlarged paraspinal lymph nodes (Figure 2). Other investigations, bone scan, and bone marrow aspiration showed no metastasis. Sonar guided biopsy from the renal mass revealed the possibility of Wilms' tumor. Right nephrectomy was performed, and the operative specimen consisted of right kidney with a portion of the ureter and grayish lobulated solid tumor of $13 \times 9\,cm^2$, surrounded by renal parenchyma. The renal capsule was infiltrated by the tumor and there was renal pelvic invasion.

Microscopically (Figures 3–6), the examined growth portion composed of a mixture of primitive blastemal cells, epithelial cells, and mesenchymal elements and these features corresponded to classic nephroblastoma. The tumor was classified as stage IV according to NWTS-3.[6]

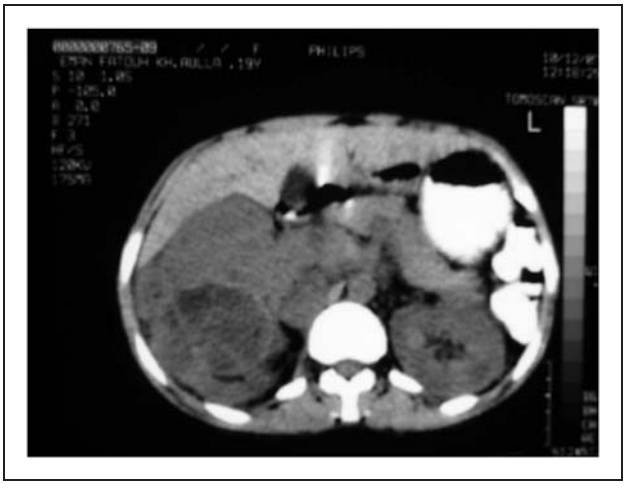

Figure 1. CT image shows huge ill-defined mass lesion involving right kidney.

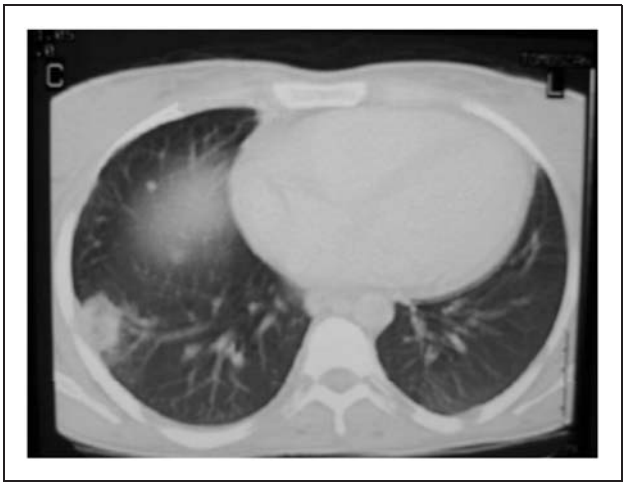

Figure 2. CT image shows pulmonary deposits.

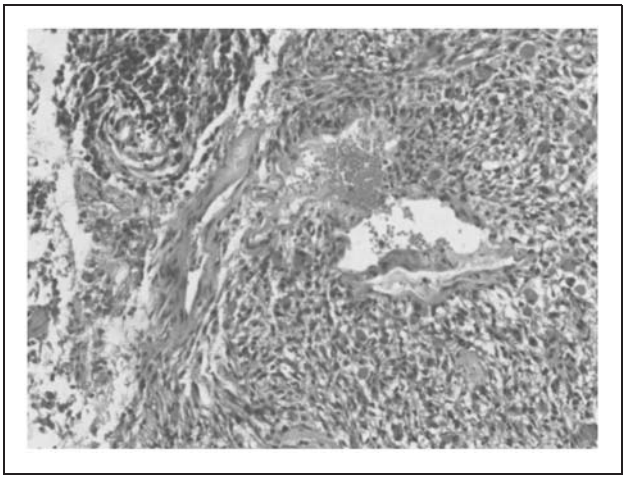

Figure 3. Microscopical picture shows stromal element.

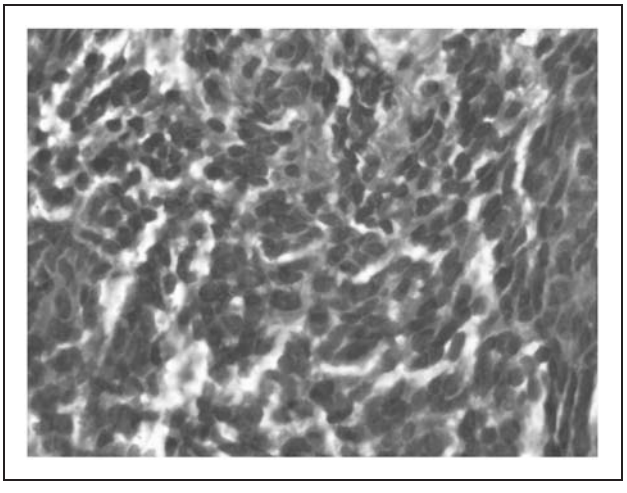

Figure 4. Microscopical picture shows the blastemal cells.

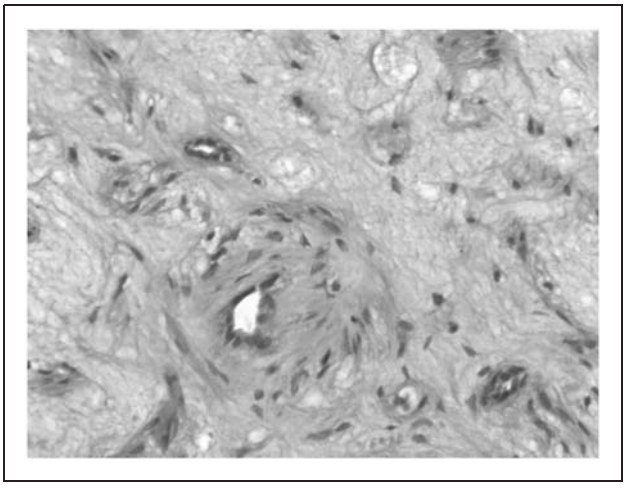

Figure 5. Microscopical picture shows epithelial element.

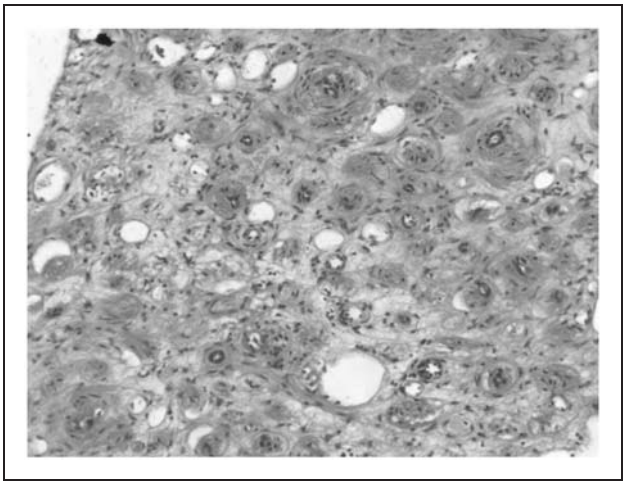

Figure 6. Microscopical picture shows epithelial element.

The patient was started multimodal therapy (combined chemotherapy and radiotherapy) immediately after nephrectomy.

Patient initiated chemotherapy in week 0 as: Dactinomycin 15 μg/kg/day × 5 days IV, Vincristine 1.5 mg/m^2 IV weekly from week 1, Doxorubicin 20 mg/m^2/day × 3 days at week 6, and cyclphosphamide 10 mg/kg/day × 3 days IV at week 6.

Whole lung irradiation was delivered one week after nephrectomy as: 12 GY (150 cGY/ day for 8 days), followed by flank irradiation 1080 cGY (180cGY/ day for 6 days).

The toxicity was mild to moderate (WHO grade 3 alopecia, grade 2 vomiting, grade 2 constipation, grade 1 peripheral neuropathy, and grade 2 myelosupression).

Assessment was performed 1 week after ending the first cycle of the formal therapy, which revealed

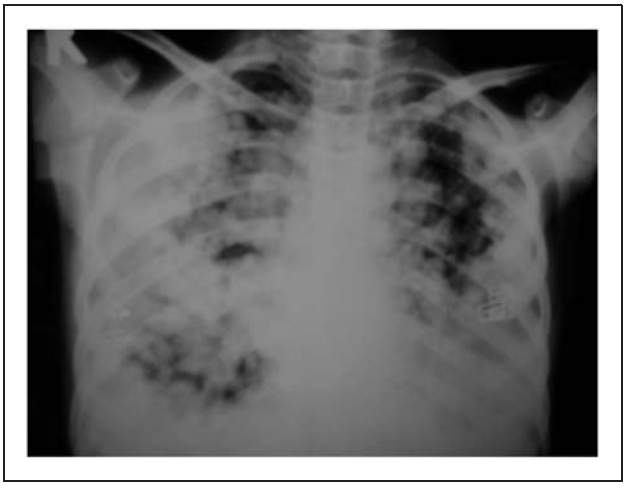

Figure 7. Chest X-ray film shows progression of lung metastasis.

progression of lung metastasis (Figure 7) and abdominal CT assessment revealed liver metastasis.

Deterioration of the patient's general condition began immediately after that, and she eventually died (2 months from the date of starting chemotherapy), because of respiratory distress.

Discussion

The clinical symptoms of our case were pain, dyspnea, and weight loss. Weichert et al.[7] and Reinhard et al.[8] reported that 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 a sudden drop in performance status.[7,8]

The needle biopsy was performed in our study before nephrectomy. Dykes et al.[9] reported that, in Europe percutaneous needle biopsy generally is used in children to assess the nature of massive renal tumors and if performed before nephrectomy, it has been shown to be effective in approximately 90% of cases.

Our case underwent nephrectomy followed by multimodal therapy, according to NWTS protocol. Many investigators agree on the need for multimodality approach in the treatment of adult Wilms' tumor but differ on how aggressive it should be. While some advocate the adoption of current pediatric protocols based on tumor size and grade, others recommended using advanced disease regimens for all stages and grades.[10–12]

The current case failed to respond to the first line therapy and died because of the disease progression; this result is in agreement with Caemic et al.[2] who reported that, the treatment outcome in adult Wilms'

is disappointing and the majority of cases die of metastatic disease.

Conclusions

Adults with Wilms' tumor are at a risk of either under treatment or incorrect treatment because of the rarity of this disease. The retrospective reviews of case reports may provide some guidelines toward effective therapy.

References

1. Alapont JM, Pontones JL and Jimenez-Cruz JF. Wilms' tumor in adults. *Int Braz J Urol* 2003; 29: 40–42.
2. Cameic Turk HM, Erkilic S, Buyuberbe S, Uner x and Ozsarae C. Early multimodal therapy in adult wilms' tumor. *J Chemotherapy* 2002; 14: 530–532.
3. Kilton L, Mathews MI and Cohen MH. Adult wilms' tumor a report of prolonged survival and review of literature. *J Urology* 1980; 124: 1–5.
4. Byrd RL, Evans A and D'Angio GJ. Adult wilms' tumor: effect of combined therapy on survival. *J Urology* 1982; 127: 648–651.
5. Williams G, Colbeck RA and Gowing NFC. Adult wilms' tumor: review of 14 patients. *Brit J Urol* 1992; 70: 230–235.
6. D'Angio GJ, Breslow N, Beckwith JB, Evans A, Baum E and De Lorimein A. Treatment of wilms' tumor. Results of the third national wilms' tumor study. *Cancer* 1989; 64: 349–360.
7. Weichert-Jacobsen K, Papadopoulos I and Skrezek C. Adult wilms' tumor: case report, management, prognosis. *Urol Int* 1995; 54: 99–103.
8. Reinhard H, Aliani S, Ruebe C, Stöckle M, Leuschner I and Graf N. Wilms' tumor in adult: results of the society of pediatric oncology (SIOP), 93/01/society of pediatric oncology and hematology (GPOH) study. *J Clin Oncol* 2004; 22: 4500–4506.
9. Dykes EH, Marwaha RK, Dicks-Mireaux C, Sams V, Ridson RA and Deiffy PG. Risks and benefits of perctaneous biopsy and primary chemotherapy in advanced wilms' tumor. *J Pediatr Surg* 1991; 26: 610.
10. Sharma M and Ahmed MZ. Adult wilms' tumor. *Saudi J Kidney Dis Transplant* 2009; 20: 835–837.
11. Kalapurakal JA, Nan B, Norkool P, et al. Treatment outcomes in adults with favorable histologic type wilms' tumor—an update from the National Wilms' Tumor Study Group. *Int J Radiat Oncol Biol Phys* 2004; 60: 1379–1384.
12. Tawil A, Cox JN, Roth AD, et al. Wilms' tumor in adult-report of a case and review of literature. *Path Res Pract* 1999; 195: 105–111 discussion: 113–114.