CASE REPORT
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Percutaneous Biopsy in Adult Wilms Tumor and A Review of the Literature

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Abstract

Wilms tumor is rare in adults, constituting only 0.5% of all renal masses. Tumors in adults are diagnosed later than those in children, and survival after treatment is still low. Some reasons for this are considered as patients not receiving neoadjuvant chemotherapy before surgery and late initiation of treatment due to delays in receiving pathology results. In this paper, we present a 38-year-old male patient diagnosed with Wilms tumor after percutaneous biopsy and review the related literature.

Keywords: Renal mass, adult Wilms tumor, percutaneous biopsy

Introduction

Wilms tumor constitutes 5-6% of all pediatric tumors. It is the most common renal tumor seen in childhood, with most being diagnosed within the first five years of life (1). In adults, Wilms tumor is rare, accounting for only 0.5% of all renal masses (2) and having an incidence of 0.2 per million (3). Current treatment approaches have increased survival in children up to 90%; however, tumors in adults are diagnosed later than those in children and remain have a lower survival rate. The main reasons for this situation are delays in diagnosis due to the rarity of the disease and lack of complete treatment protocols for adults. Other reasons are considered as patients not receiving neoadjuvant chemotherapy before surgery and late initiation of treatment due to delays in receiving pathology results (4). However, there are publications in the literature that recommend percutaneous biopsy (PCB) before surgical treatment in young adult patients with renal masses suspected to be Wilms tumors (3).

In this paper, we present a 38-year-old male patient who was suspected to have lymphoma and Wilms tumor in the preliminary diagnosis based on imaging characteristics and was diagnosed with Wilms tumor as a result of PCB performed for differentiation.

Case Report

A 38-year-old male patient presented to an external healthcare center with the complaint of left flank pain and was referred to our tertiary hospital with a preliminary diagnosis of a renal mass. Dynamic renal computed tomography (CT) was examined on the patient. CT revealed a mass lesion originating from the left kidney, occupying a large area in the left retroperitoneal area, measuring approximately 20 cm at its largest dimension, partially extending into the bony pelvis, extending from the midline to the right, and containing necrotic areas in the central part (Figure 1A, 1B). In the dynamic examination, the mass showed progressive enhancement in the solid parts, except for the necrotic parts. The mass was observed to significantly compress the renal vein, but thrombosis was not present. Pathological lymph nodes with the largest having a 2-cm short axis, were detected in the right and left paraaortic areas. Magnetic resonance imaging was performed with a preliminary diagnosis of lymphoma and demonstrated mass originating from the left kidney containing cystic necrotic areas with hypointensity in T1-weighted images and hyperintensity in T2-weighted images, and contrast enhancement of solid parts (Figure 2A, 2B). Since a differentiation between lymphoma-Wilms tumor and mesenchymal lesion could not be made, percutaneous tru-cut biopsy was performed from the mass. As a result, the mass was diagnosed as a Wilms tumor. In positron emission tomography

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CT performed for staging purposes, mass lesions with high standardized uptake value:12 values were observed in the mass and aortic lymph nodes. No distant metastasis was detected. Based on the diagnosis of Wilms tumor, the patient was started on neoadjuvant chemotherapy that consisted of ifosfamide, etoposide. After observing a reduction in mass size and tumor regression in control imaging, left radical nephrectomy was performed (Figure 3A, 3B). The operative specimen consisted of left kidney with a portion of the ureter and on gross examination the tumor was 13x9.5x11.5 cm large, white- grayish, solid. The renal capsule, the renal sinus, renal vein were infiltrated by the tumor and the tumor involved lymphovascular spaces. Pathology showed blastemal predominant WT, with a minor epithelioid component. No features of anaplasia were found. The radical nephrectomy pathology result of the patient was evaluated as Stage 3 Wilms tumor. The patient was followed up and no recurrence or metastasis was detected in imaging during the first two years. The patient was followed up with thorax, abdomen and pelvic CT for a 3-month period.

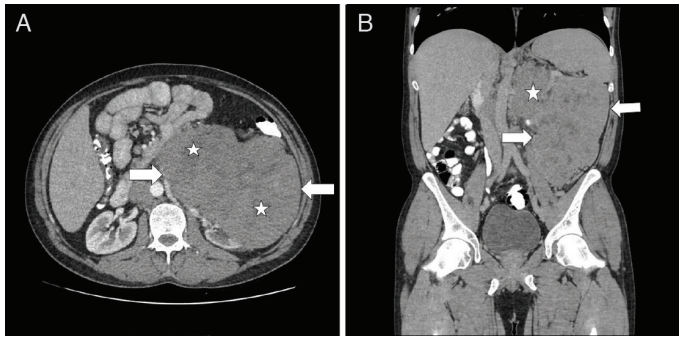

Figure 1. CT examination of the patient before chemotherapy at the time of diagnosis A. Axial and B. Coronal plane CT shows a mass lesion originating from the left kidney (arrow) and containing necrotic areas in the central part (star)

CT: Computed tomography

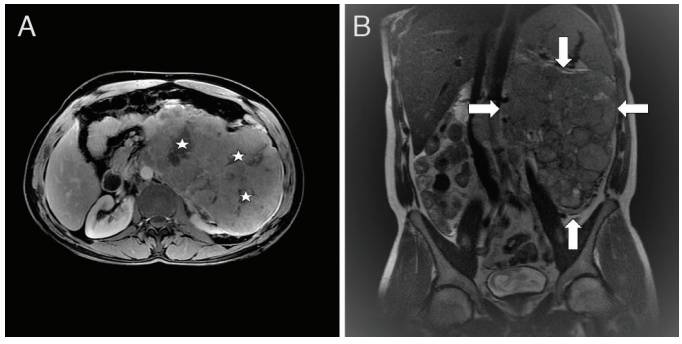

Figure 2. MRI examination of the patient at the time of diagnosis A. Axial T1-weightened MRI shows a mass originating from the left kidney containing cystic necrotic areas with hypointensity (star) and contrast enhancement of solid parts and B. Coronal T2-weightened T2 MRI plane shows mass lesion extending into the bony pelvis, extending from the midline to the right (arrows)

MRI: Magnetic resonance imaging

Liver metastasis emerged in imaging performed at the end of the second year. Abdominal CT examination revealed a newly developed mass in the right lobe of the liver, which was primarily evaluated as a metastasis. The patient underwent right hepatectomy. The pathology metastectomy result of the patient was evaluated as a Wilms tumor metastasis. No recurrence was detected in the next 3 months follow-up abdomen CT. Written and informed consent from the patient was obtained for publishing.

Discussion

Wilm tumor has a poorer prognosis in adults than in children. However, recently, there has been an increase in relative survival. The combination of radiotherapy, chemotherapy and surgery is recommended for treatment. Some studies have reported that the five-year survival rate has increased to 82.6% in the group with favorable histological characteristics (5).

However, despite many developments, the lack of protocols for adults makes the diagnosis, treatment and follow-up of Wilms tumor difficult. Similar to children, treatments vary according to tumor stage and histological type in adults. Due to its lower incidence, no phase 3 studies or treatment guidelines are available for adult Wilms' tumors, and in most isolated cases, the management is extrapolated from pediatric guidelines. The standard pediatric treatment has been proposed by two groups; the International Society of Paediatric Oncology (SIOP) and the National Wilms Tumor Study (NWTS) (6). One of the main differences between the two protocols is that the NWTS approach advocates up-front nephrectomy, whereas the SIOP protocols emphasize neoadjuvant chemotherapy (7). The most effective chemotherapeutics in treatment of nephroblastoma are vincristine, actinomycin D, ifosfamide, carboplatin, cyclophosphamide, etoposide and doxorubicin. According to SIOP, neoadjuvant chemotherapy reduces the risk of tumor rupture and reduces the probability of recurrence (8). This

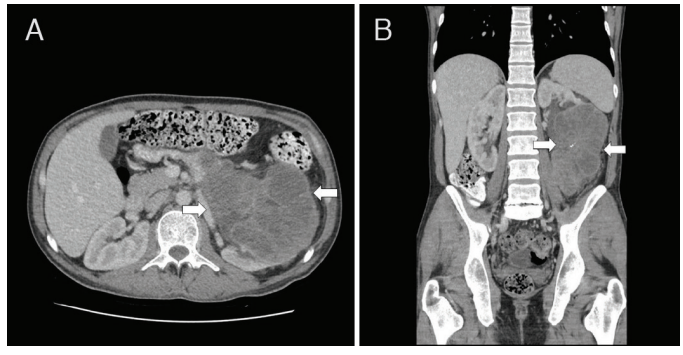

Figure 3. CT examination of the patient after neoadjuvant chemotherapy A. Axial and B. Coronal plane CT shows a reduction in mass size and tumor regression (arrow)

CT: Computed tomography

makes it necessary to make a diagnosis before the operation and accurately stage the tumor (9). But the use of PCB in patients with renal masses is a method that is not widely preferred due to the possibility of tumor seeding and frequent complications, such as bleeding. After imaging in patients with renal masses, the diagnosis is usually made by performing total or partial nephrectomy (10).

Another critical point in adult Wilms tumors is the lack of guidelines on how to follow-up patients after surgery. The current protocol of SIOP surveillance for Wilms' tumor recommends that abdominal imaging and chest X-ray should be performed every 3 months for the first 2 years. imaging is repeated every 4-6 months in the third and fourth years and annually in the fifth year (11). The same recommendation is advised for adults in studies since most of the relapses occur within 2 years of completion of therapy (9).

It has been reported that there may be inadequacies in the differential diagnosis of renal cell carcinoma (RCC) from benign masses, such as lipid-poor angiomyolipoma and renal oncocytoma and malignant masses (12). There are publications showing that clear cell RCC can be distinguished from other masses to a large extent, but the differential diagnosis of other subtypes is mostly impossible (13). In addition, rare mass lesions, such as Wilms tumors are often not considered in differential diagnosis. Mostly, PCB is used only when primary diagnoses, such as lymphoma and urethral carcinoma, are considered based on imaging characteristics or in cases where a differential diagnosis of infection or non-renal mass cannot be made (10).

In PCB studies conducted early in the 21st century, the rate of non-diagnostic PCB was reported as 31%, and the false negativity rate was as high as 25%. Based on these and similar results, PCB was not included in most localized renal mass algorithms (14,15). However, in a more recent study, the diagnosis rate of PCB was reported as 72% for fine-needle biopsy, 87% for core biopsy, and 92% for both (16). In addition, Ozambela et al. (17) determined the rate of PCB-related complications as 5.18% for hematuria and 1.75% for pneumothorax. The rate of others, including perirenal hematoma, pseudoaneurysm, and arteriovenous fistula, was reported to be 0.1%. Despite all these recent developments, when the same authors examined the rate of percutaneous renal mass biopsies (RMB) performed in the USA between 2006 and 2017, they observed that although the number of RMB procedures had increased over the years, this was not sufficient. They also stated that less than 15% of patients with renal masses underwent PCB (17). The American Association of Urology and the American Society of Clinical Oncologists report that the PCB technique is reliable and has high diagnostic value. PCB is indicated in small-sized renal masses that are probably benign, in patients who will be actively monitored due to low life expectancy, and in these cases scheduled for thermal ablation, and when the mass is considered to be of hematological, inflammatory, or infectious origin. In addition, PCB is recommended in cases with Wilms tumor, in which a pathological diagnosis would change the treatment protocol of the patient and impact survival (18,19).

It is known that the earlier chemotherapy is started in adult patients with Wilms tumor, the better survival is, and chemotherapy is recommended before surgery in this patient group. In a previous study, patients who started treatment in the first 30 days after surgery and those who received delayed treatment were compared. The five-year survival rate was 80% in patients who received early treatment, while it was 28.6% in the other group (20). Based on this information, the diagnosis of Wilms tumor based on PCB may start treatment at the earliest stage in these patients. In this study, because of the preliminary diagnosis of lymphoma and Wilms tumor, PCB was performed, which allowed for the early diagnosis of Wilms tumor. Thus, chemotherapy was applied before surgery, and because of the reduction in the size of the mass, the operation was safer. With this case report, we wanted to emphasize the current position and importance of the PCB method in patients with Wilms tumors and renal masses in general.

Ethics

Informed Consent: Written and informed consent from the patient was obtained for publishing.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: E.G., Concept: E.E., Design: E.E., E.G., Data Collection or Processing: E.E., Analysis or Interpretation: E.G., Literature Search: E.E., E.G., Writing: E.E.

Conflict of Interest: No conflict of interest was declared by the authors.

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References

1. De Vries-Brilland M, Sionneau B, Dutriaux C, Blay JY, Ravaud A, Gross-Goupil M. Successful Treatment of Metastatic Adult Wilms Tumor With Anti-BRAF Treatment: A Case Report and a Brief Review of the Literature. Clin Genitourin Cancer 2019;17:e721-e723.
2. Reinhard H, Aliani S, Ruebe C, Stöckle M, Leuschner I, Graf N. Wilms' tumor in adults: results of the Society of Pediatric Oncology (SIOP) 93-01/ Society for Pediatric Oncology and Hematology (GPOH) Study. J Clin Oncol 2004;22:4500-4506.
3. Ratajczyk K, Czekaj A, Rogala J, Kowal P. Adult Wilms tumor with inferior vena cava thrombus and distal deep vein thrombosis - a case report and literature review. World J Surg Oncol 2018;16:38.
4. Huszno J, Starzyczny-Słota D, Jaworska M, Nowara E. Adult Wilms' tumor - diagnosis and current therapy. Cent European J Urol 2013;66:39-44.

5. Kalapurakal JA, Nan B, Norkool P, Coppes M, Perlman E, Beckwith B, Ritchey M, Breslow N, Grundy P, D'angio GJ, Green DM, Thomas PR. 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.

6. Modi S, Tiang KW, Inglis P, Collins S. Adult Wilms' tumour: case report and review of literature. J Kidney Cancer VHL 2016;3:1-7.

7. Brown JT, Harik LR, Barbee MS, Esiashvili N, Badell ML, Goldsmith KC, Master VA, Bilen MA. Multidisciplinary Care of Adult Wilms' Tumor During Pregnancy: A Case Report and Review of the Literature. Clin Genitourin Cancer 2020;18:e1-e4.

8. De Kraker J, Graf N, van Tinteren H, Pein F, Sandstedt B, Godzinski J, Tournade MF. Reduction of postoperative chemotherapy in children with stage I intermediate risk and anaplastic Wilms, tumor (SIOP 93–01 trial) randomized controlled trial. Lancet 2004;364:1229-1235.

9. Segers H, van den Heuvel-Eibrink MM, Pritchard-Jones K, Coppes MJ, Aitchison M, Bergeron C, de Camargo B, Dome JS, Grundy P, Gatta G, Graf N, Grundy P, Kalapurakal JA, de Kraker J, Perlman EJ, Reinhard H, Spreafico F, Vujanic G, Warwick AB; SIOP-RTSG and the COG-Renal Tumour Committee. Management of adults with Wilms' tumor: recommendations based on international consensus. Expert Rev Anticancer Ther 2011;11:1105-1113.

10. Lim CS, Schieda N, Silverman SG. Update on Indications for Percutaneous Renal Mass Biopsy in the Era of Advanced CT and MRI. AJR Am J Roentgenol 2019:1-10.

11. Brok J, Lopez-Yurda M, Tinteren HV, Treger TD, Furtwängler R, Graf N, Bergeron C, van den Heuvel-Eibrink MM, Pritchard-Jones K, Olsen ØE, de Camargo B, Verschuur A, Spreafico F. Relapse of Wilms' tumour and detection methods: a retrospective analysis of the 2001 Renal Tumour Study Group-International Society of Paediatric Oncology Wilms' tumour protocol database. Lancet Oncol 2018;19:1072-1081.

12. Ljungberg B, Bensalah K, Canfield S, Dabestani S, Hofmann F, Hora M, Kuczyk MA, Lam T, Marconi L, Merseburger AS, Mulders P, Powles T, Staehler M, Volpe A, Bex A. EAU guidelines on renal cell carcinoma: 2014 update. Eur Urol 2015;67:913-924.

13. Canvasser NE, Kay FU, Xi Y, Pinho DF, Costa D, de Leon AD, Khatri G, Leyendecker JR, Yokoo T, Lay A, Kavoussi N, Koseoglu E, Cadeddu JA, Pedrosa I. Diagnostic Accuracy of Multiparametric Magnetic Resonance Imaging to Identify Clear Cell Renal Cell Carcinoma in cT1a Renal Masses. J Urol 2017;198:780-786.

14. Dechet CB, Zincke H, Sebo TJ, King BF, LeRoy AJ, Farrow GM, Blute ML. Prospective analysis of computerized tomography and needle biopsy with permanent sectioning to determine the nature of solid renal masses in adults. J Urol 2003;169:71-74.

15. Lane BR, Samplaski MK, Herts BR, Zhou M, Novick AC, Campbell SC. Renal mass biopsy--a renaissance? J Urol 2008;179:20-27.

16. Yang CS, Choi E, Idrees MT, Chen S, Wu HH. Percutaneous biopsy of the renal mass: FNA or core needle biopsy? Cancer Cytopathol 2017;125:407-415.

17. Ozambela M, Wang Y, Leow JJ, Silverman SG, Chung BI, Chang SL. Contemporary trends in percutaneous renal mass biopsy utilization in the United States. Urol Oncol 2020;38:835-843.

18. Campbell S, Uzzo RG, Allaf ME, Bass EB, Cadeddu JA, Chang A, Clark PE, Davis BJ, Derweesh IH, Giambarresi L, Gervais DA, Hu SL, Lane BR, Leibovich BC, Pierorazio PM. Renal Mass and Localized Renal Cancer: AUA Guideline. J Urol 2017;198:520-529.

19. Finelli A, Ismaila N, Bro B, Durack J, Eggener S, Evans A, Gill I, Graham D, Huang W, Jewett MA, Latcha S, Lowrance W, Rosner M, Shayegan B, Thompson RH, Uzzo R, Russo P. Management of Small Renal Masses: American Society of Clinical Oncology Clinical Practice Guideline. J Clin Oncol 2017;35:668-680.

20. Terenziani M, Spreafico F, Collini P, Piva L, Perotti D, Podda M, Gandola L, Massimino M, Cereda S, Cefalo G, Luksch R, Casanova M, Ferrari A, Polastri D, Valagussa P, Fossati-Bellani F. Adult Wilms' tumor: A monoinstitutional experience and a review of the literature. Cancer 2004;101:289-293.