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Partial Nephrectomy in Wilms Tumor: A Rarely Implemented Surgical Method

Wilms Tümöründe Parsiyel Nefrektomi: Çok Nadir Uygulanan Bir Cerrahi Yöntem

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Abstract

Nephroblastoma (Wilms Tumor/WT) is the rare but the most common primary kidney tumor in children. The WT is generally diagnosed between ages of 1 and 5 and the most common diagnosis is the age of 3. Up to 95% of WTs are diagnosed correctly with imaging (ultrasound, computed tomography and magnetic resonance). Histopathologicaly tissue examination is implemented and subtypes are determined. Surgery is one of the key factors in WT treatment. Transperitoneal radical nephrectomy is the standard operation for unilateral WTs. Nephron sparing surgery (partial nephrectomy) is suggested only in selected cases with a single kidney or bilateral WT. Other treatment combination for Wilms tumor involves chemotherapy and radiation treatment. A patient who was diagnosed with Wilms tumor in the right kidney and underwent partial nephrectomy which is rarely implemented or recommended in selected cases is represented in this article.

Keywords: Wilms tumor, child, partial nephrectomy

Özet

Nefroblastoma (Wilms tümörü/WT) çocuklarda nadir fakat en sık görülen primer böbrek tümörüdür. WT tanısı genellikle 1 ile 5 yaş arasında konur ve en sık tanı 3 yaştır. WT'lerin %95'e kadarı görüntüleme (ultrason, bilgisayarlı tomografi ve manyetik rezonans) yöntemleri ile doğru şekilde teşhis edilir. Histopatolojik olarak ise doku incelemesi yapılır ve alt tipleri belirlenmektedir. Cerrahi WT tedavisinin temel taşlarından biridir. Tek taraflı WT'ler için transperitoneal radikal nefrektomi standart operasyondur. Nefron koruyucu cerrahi (parsiyel nefrektomi) yalnızca tek böbrekli veya iki taraflı WT'li seçilmiş hasta vakalarında uygulanması önerilmektedir. Wilms tümörünün diğer tedavi kombinasyonu ise kemoterapi ve radyasyon tedavisini içermektedir. Bu yazıda sağ böbrekte Wilms tümörü tanısı alan, çok nadiren yapılan veya seçilmiş vakalarda yapılması önerilen parsiyel nefrektomi yapılmış hasta sunulmaktadır.

Anahtar kelimeler: Wilms tümörü, çocuk, parsiyel nefrektomi

Introduction

Wilms tumor (WT) is the most common renal tumor in childhood period, affecting one in 10,000 children [1–3]. It is mostly seen between the ages of 2-4. WT is an embriological tumor that classicaly shows a triphasic histological complex structure originating from blastem, epithelial and stroma components. Besides that it may also includes cartilage, osteoid and neuronal elements [4].

The WT treatment is implemented with two distinctive methods: The Europe's International Society of Pediatric Oncology (SIOP) method which adopts the principle of initiating chemotherapy (CT) without tissue diagnosis and then surgical application and North America's National Wilms Tumor Study (NWTS), now known as the Children's Oncology Group method (COG) which carries out a treatment plan with tissue diagnosis. Primarly providing CT contributes to preventation of phase escalation due to tumor cells being shed during surgery and surgical complications that may occur in the presence of large-sized or thrombus but besides that has a disadvantage of providing unnecessary CT to the cases diagnosed histopathologically other than WT. In NWTS/COG method, although tissue diagnosis is the main criterion, the principle of providing CT first is adopted in very large tumors, bilateral cases and the presence of thrombus extending into the IVC or atrium. However, the most important factor determining the prognosis in both methods is the phase of tumor, whether it contains anaplasia and the positivity of biological indicators such as 1p, 16q LOH veya 11p15q LOH [2,3,5].

Surgery is one of the key factors in WT treatment. Transperitoneal radical nephrectomy is standard operation for unilateral WTs. Nephron sparing surgery is suggested to be implemented in selected patient cases with single kidneys or bilateral WT. In this case report, a patient who diagnosed with Wilms tumor in right kidney at the age of 4 and underwent partial nephrectomy which is rarely implemented or suggested to be implemented in selected cases is represented.

Case

Our case is a 4-years old male patient who was brought to our clinic by his parents with the complaint of bleeding in the urine which had been going on for 1 month. Full blood cell count and biochemical values of the patient were within normal limit, there was 3+ hematuria in the urinalysis. In the urinary track ultrasound examination a 4 cm suspicious mass was seen in the lower pole of right kidney. Whole abdominal magnetic resonance imaging performed on the patient was reported as 'a mass lesion approximately 42x37 mm in size in the lower pole of the right kidney, with a smoothly circumscribed, encapsulated appearance, containing cystic areas, extending towards the renal pelvis, but not creating an appearance in favor of significant invasion findings and showing lower but heterogeneous contrast uptake compared to the renal parenchyma and other organs in the abdomen are normal **(Figure 1).** Partial nephrectomy decision was made due to the fact that the tumor was located in a single pole, infiltrated less than 1/3 of the kidney, there was no renal vein invasion, and there was surgical experience in this regard. The patient underwent right partial nephrectomy with the provisional diagnosis of renal mass. Partial nephrectomy was 4,5x3,5x3 cm in size and in the form of yellowish brown tumor tissue with a diameter of 3,5 cm at a distance of 0,2 cm from the nearest surgical margin in cross sections. Microscopically, in the examination of the sections, a partially cystic tumor tissue with a generally solid pattern that is seperated from the kidney tissue with a distinct but irregular border line is seen. The tumor is comprised of 3 different components; epithelial component which is differentiated to generally tubular, partially glomerulus structures, blastomatous component characterized with primitive fusiform cells surrounding them and stromal component which formed bundles between them and partly showed prominent rhabdoid differentiation. In immunohistochemical studies, it was reported that positive colouration was detected in epithelial component with Pan-CK, in both the stromal and partly blastomatous components with vimentin, in rhabdomyoblastic cells with desmin. As a result of these histopathological findings, tumor was reported as WT with classical, triphasic features and appropriate histology **(Figure 2).** The patient then underwent chemotherapy and radiotherapy. No relapse was observed during patient's 8-years follow-up. No problems occured in the kidney that underwent partial nephrectomy within years **(Figure 3).**

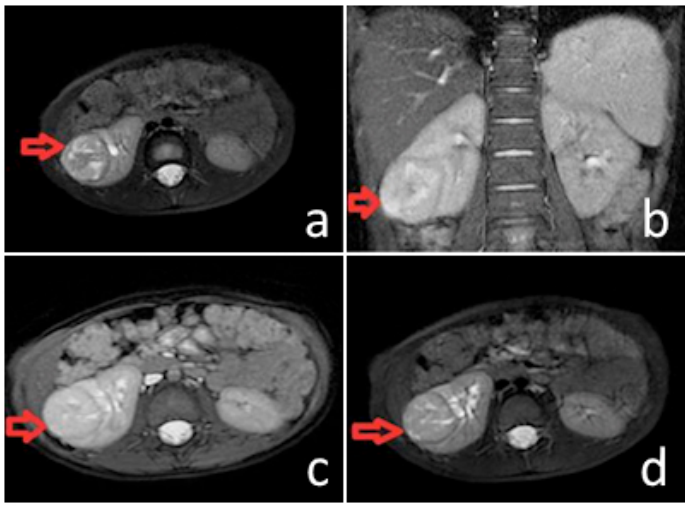

Figure 1. Mass lesion approximately 42x37 mm in size in the lower pole of the right kidney, with a smoothly circumscribed, encapsulating appearance, containing cystic areas, extending toward the renal pelvis, but not creating appearance in favor of significant invasion findings and showing lower but heterogeneous contrast uptake compared to the renal parenchyma

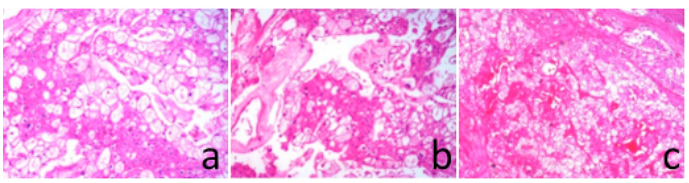

Figure 2. H&EX20 epithelial component which is differentiated to generally tubular, partially glomerulus structures, blastomatous component characterized with primitive fusiform cells surrounding them and stromal component which formed bundles between them and partly showed prominent rhabdoid differentiation

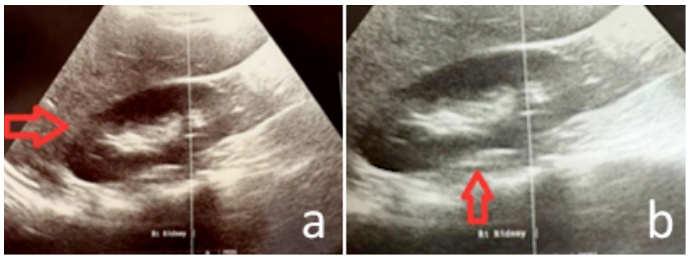

Figure 3. View of the kidney that underwent partial nephrectomy at the last check-up

Discussion

Surgery is one of the key factors in WT treatment. The SIOP method in Europe suggests chemotherapy before surgery while COG in North America suggests operation before chemotherapy [6]. The direction of WT requires a multidisciplinary approach with the participation of pediatric radiologist, oncologist, surgeon and radiotherapist. Firstly, in order to determine the origin of tumor, its position as regards to surrounding tissue and vascular uptake the patient should be carefully evaluated in pre-operative period by using appropriate imaging techniques.

Transperitoneal radical nephrectomy is standard operation for unilateral WTs. Nephron sparing surgery is suggested only in selected patient cases with single kidney or bilateral WT [7]. It is discussed in unilateral cases whether it should be implemented by partial nephrectomy or enucleaction [8,9]. This approach is suggested only in synchronous or metachronous bilateral cases or single kidneys. Only less than 5% of unilateral WTs are appropriate for partial nephrectomy because most of the kidneys are locally progressed during diagnosis [8,10]. For partial resection surgical criteria are as follows: tumor is placed in a single pole and infiltrated less than 1/3 of the kidney; there is no renal vein invasion, surgeon has experience in pediatric oncology [8,11]. We think that our case will contribute to literature in a way that it is a case presentation diagnosed with Wilms tumor in the right kidney and underwent partial nephrectomy that is rarely implemented or suggested to implemented in selected cases.

Conclusion

Although radical nephrectomy was standard in cases which Wilms tumor was detected we would like to specify that partial nephrectomy operation may give successful results in selected cases.

Ethics Committee Approval: N / A.
Informed Consent: An informed consent was obtained from the patient's parents.
Publication: The results of the study were not published in full or in part in form of abstracts.
Peer-review: Externally peer-reviewed.
Authorship Contributions: Any contribution was not made by any individual not listed as an author. Concept – K.Y.; Design – K.Y.; Supervision – K.Y.; Resources – K.Y.; Materials – K.Y.; Data Collection and/or Processing – K.Y.; Analysis and/or Interpretation – K.Y.; Literature Search – K.Y.; Writing Manuscript – K.Y.; Critical Review – K.Y.

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