Balkan Med J 2021;38:135-136

Primary renal neuroblastoma mimicking Wilms' tumor

Vedat Akçaer[1], Tuba Eren[2], Ebru Taştekin[3]

[1]Department of Pediatric Surgery, Trakya University School of Medicine, Edirne, Turkey
[2]Department of Pediatric Hematology and Oncology, Trakya University School of Medicine, Edirne, Turkey
[3]Department of Pathology, Trakya University School of Medicine, Edirne, Turkey

A 22-month-old girl was referred to our clinic with a prediagnosis of Wilms' tumor from the health care center where she presented with persistent fever. On examination of the patient, a painless, firm, smooth-surfaced, and immobile mass, which was sized approximately 15 x 10 cm and filling the left upper quadrant of the abdomen, was detected. Apart from having anemia, the remaining laboratory findings of the patient, in whose systemic examinations no other clinical feature was found, were normal. On the basis of an analysis of the ultrasonography (USG) and computed tomography (CT) images, a heterogeneous mass with a size of 8 x 9 x 12 cm crossed the midline, which was considered to be generated by the left kidney, was interpreted as Wilms' tumor (Figure 1. a, b). The patient underwent a left nephroureterectomy and periaortic lymph node excision. The patient whose pathology result was reported as primary renal neuroblastoma (NB) (PRNB) was assessed to have LN invasion. The neuroendocrine marker synaptophysin was positive, and N-Myc amplification was positive in our patient, whereas WT-1 was found to be negative on analysis. Chemotherapy was administered to the patient whose bone marrow biopsy was negative.

NB is the most frequent type of extracranial solid tumor during childhood. It has an incidence rate of 1 per 100,000. More than 40% of the patients are in the metastatic or high-risk group at the time of diagnosis. Despite multimodal treatment, the survival rate among these patients is 31.2%.[1] Intrarenal NB is a very rare primary renal malignancy, similar to our case. It is considered to originate from renal sympathetic ganglia or adrenal medullary tissue within the renal parenchyma.[2] Localization of PRNB might be confused with Wilms' tumor, which is much more prevalent owing to its similar clinical and radiological features. Patients who have PRNB also present with a firm, nonsensitive abdominal mass, such as Wilms' tumor. Furthermore, they might have nonspecific systemic symptoms, such as fever, weight loss, and anemia.[3] Hypertension could be crucial evidence for diagnosis. Hypertension is observed in 27% of extrarenal NB cases, whereas this rate increases to 66-100% in PRNB cases.[3,4] It has been revealed that hypertension develops between 20% and 25% of patients with Wilms' tumor.[1] It is considered that hypertension develops because of the pressure on renal vessels or owing to a rise in the level of renin and catecholamine.[3] Hypertension was not observed in our patient, although there were persistent fever, anemia, and intra-abdominal mass.

USG and subsequent CT or magnetic resonance imaging performed in the radiographic evaluation are helpful in assessing primary tumor and local invasion and treatment planning.[5] It has been

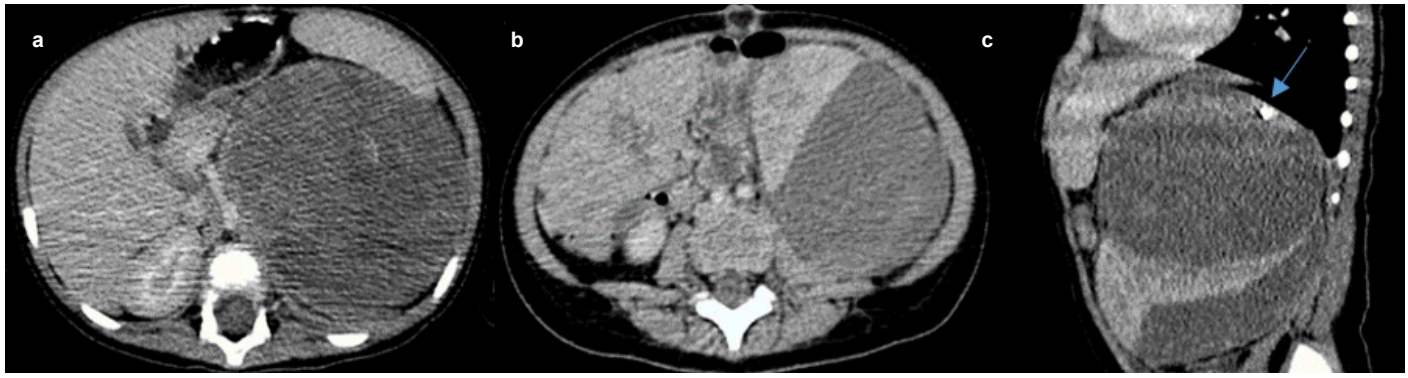

FIG. 1. (a-c). Preoperative CT images of the patient. The mass crossed the midline (a). There is a large heterogeneous mass arising from within the left kidney (b). Intratumoral calcification was seen (blue arrow) (c). The left adrenal gland was not able to be identified.
CT: computed tomography.

Address for Correspondence: Vedat Akçaer, Department of Pediatric Surgery, Trakya University School of Medicine, Edirne, Turkey
e-mail: vedatakcar@gmail.com
Received: July 22, 2020 Accepted: November 6, 2020 • DOI: 10.4274/balkanmedj.galenos.2020.2020.7.204

Available at www.balkanmedicaljournal.org

ORCID iDs of the authors: V.A.; 0000-0002-1152-3216; T.E. 0000-0001-5471-0663; E.T. 0000-0002-7686-7765.
Cite this article as:
Akçaer V, Eren T, Taştekin E. Primary renal neuroblastoma mimicking Wilms' tumor. *Balkan Med J.* 2021;38(2):135-136.

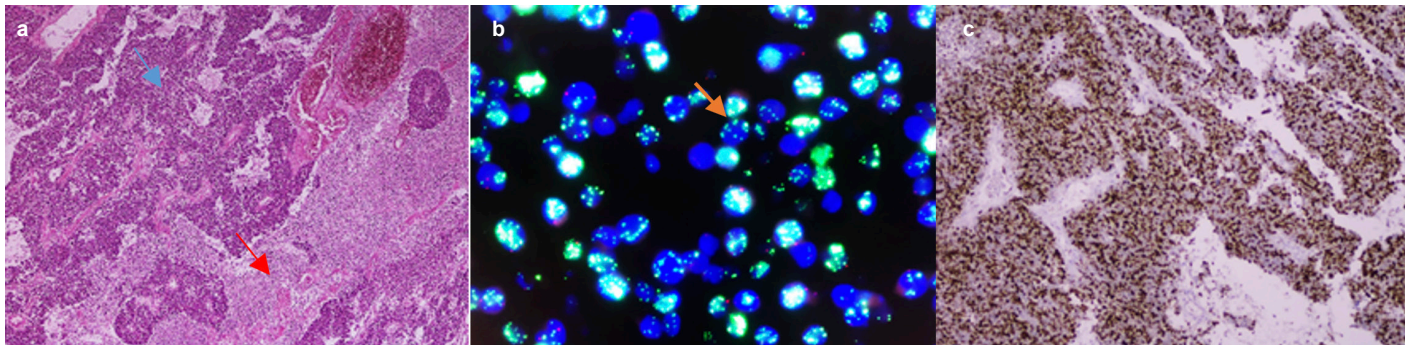

FIG. 2. (a-c). Undifferentiated tumor cells (blue arrow) and necrotic areas (red arrow) on hematoxylin eosin-stained slides (× 40) (a). N-myc amplification was shown with fluorescence in situ hybridization method (arrow: signals in amplified tumor cells) (b). Synaptophysin positivity in tumor cells (c).

shown that calcification was found in 11% of patients with Wilms' tumor and 67% of patients with PRNB.[3,6] However, it was suggested that calcification was not detected in patients studied by Fan et al., which consisted of 8 patients[7], whereas calcified regions within the tumor were monitored in our patient using CT (Figure 1c).

NB is histopathologically defined as favorable and unfavorable.[1] PRNB is generally in unfavorable histology, and its rate of anaplasia is higher than that in adrenal NB and Wilms' tumor. Our case falls into the unfavorable histology category of the International Neuroblastoma Pathology Classification. Undifferentiated tumor cells (blue arrow) and necrotic areas (red arrow) were visible on hematoxylin eosin-stained slides (Figure 2a). Hence, patients with PRNB are typically detected at advanced stages at the time of admission.[4] The most significant prognostic factors in histopathological terms are the presence of N-myc and DNA ploidies.[8] Similarly, synaptophysin and N-myc amplification were positive in our patient on the basis of the analysis (Figure 2. b, c).

In the presence of hypertension and radiologically intratumoral calcification in renal masses, PRNB should be considered before the Wilms' tumor in prediagnosis. Albeit, both tumors have analogous features clinically and radiologically; radiological and pathological correlations are vital in structuring the diagnosis and treatment because their treatment responses are distinct and their prognoses are not similar. Immunohistochemical analyses contribute considerably to verifying the diagnosis and planning the treatment. The prognosis of PRNB is worse than that of Wilms' tumor, requiring a more painstaking chemotherapy and adjuvant therapy, and monitoring of patients should be performed more meticulously.

Patient Consent for Publication: Informed consent was obtained from the patient.

Author Contributions: Concept - V. A., T. E. ; Design - V.A.; Supervision - T. E., E. T.; Resources - V. A., T. E., E. T.; Data Collection and/or Processing - V. A., T. E., E. T.; Analysis and/or Interpretation - Literature Search - V. A., T.E., E.T.; Writing - V. A., T. E., E. T.; Critical Review - V. A., T. E., E. T.

Conflict of Interest: The authors have no conflicts of interest to declare.

Funding: The authors declared that this study has received no financial support.

REFERENCES

1. Holcomb GW, Murphy JP, Peter SDS . Holcomb and Ashraft pediatric surgery. 7th edition. In Davidoff AM, Nuchtern JG, editors. Neuroblastoma . Philadelphia: Elsevier; 2019: 1010-30.
2. Shende A, Wind ES, Lanzkowsky P. Intrarenal neuroblastoma mimicking Wilms' tumor. *N Y State J Med.* 1979;79(1):93.
3. Kessler OJ, Sıegel JF, Brock WA. Intrarenal neuroblastoma masquerading as Wilms' tumor. *Urology.* 1998;51:313-316. [Crossref]
4. Shamsian BS, Kajizadi M, Rezaei N , et al. Primary intrarenal neuroblastoma with hypertension and disseminated intravascular coagulation. *Case Rep Oncol Med.* 2013;2013:684939. [Crossref]
5. Newman EA, Abdessalam S, Aldrink JH, et al. Update on neuroblastoma. *J Pediatr Surg.* 2019;54(3):383-389. [Crossref]
6. Dickson PV, Sims TL, Streck CJ, et al. Avoiding misdiagnosing neuroblastoma as Wilms tumor. *J Pediatr Surg.* 2008;43(6):1159-1163. [Crossref]
7. Fan R. Primary renal neuroblastoma-a clinical pathologic study of 8 cases. *Am J Surg Pathol.* 2012; 36(1):94-100. [Crossref]
8. Riley RD, Heney D, Jones DR, et al. A systematic review of molecular and biological tumor markers in neuroblastoma. *Clin Cancer Res.* 2004;10(1 Pt 1):4-12. [Crossref]