Balkan Med J 2024;41:147-8

Unexpected Radiologic Findings of a Botryoid Wilms Tumor with Extension into the Bladder

Merve Yazol[1], Mehmet Arda İnan[2], Öznur Leman Boyunağa[1]

[1]Department of Radiology, Gazi University Faculty of Medicine, Ankara, Türkiye
[2]Department of Pathology, Gazi University Faculty of Medicine, Ankara, Türkiye

A 4-year-old boy with gross hematuria, painful urination and a palpable abdominal mass on physical examination was referred to us. The initial abdominal ultrasound (US) revealed a 9 x 8 x 5 cm hypoechoic mass originating from the right pelvicalyceal system. The mass had a distinct lobulated shape and extended into the distal ureter, causing considerable dilation (Figure 1a, b). Contrast-enhanced computed tomography (CT) revealed significant displacement of the renal parenchyma and corroborated the US findings (Figure 1c). Magnetic resonance imaging (MRI) revealed a multilobular polypoid mass that did not invade the parenchyma. Additionally, the following complex features were observed: T1 hypointense and T2 hyperintense areas, indicating indicating necrosis with cystic areas (Figure 2a); peripheral T1 hyperintensity, indicating a hemorrhage; and evidence of diffusion restriction (Figure 2b, c).

The tumor's unique features, pelvicalyceal system origin and extension into the ureter and pelvis, led us to reconsider our initial diagnosis of a classic Wilms tumor. It prompted us to consider a botryoid subtype of Wilms tumor.[1] Furthermore, the complex radiological findings widened our differential diagnoses to include other rare pediatric renal tumors such as malignant rhabdoid tumor, rhabdomyosarcoma, and xanthogranulomatous pyelonephritis.[2,3] A radical nephrectomy was performed. Histopathological examination revealed a large tumor with blastemal, epithelial, and stromal components, which infiltrated the pelvicalyceal system and extended into the distal ureter (Figure 3a). The blastemal component included small- to medium-sized undifferentiated cells with relatively small, regularly shaped nuclei and small nucleoli. The stromal component exhibited rhabdomyoblasts, while the epithelial component exhibited abundant rosette-like structures (Figure 3b). The tumor lacked anaplastic features, which is crucial for determining the prognosis. WT1, the most helpful marker, was generously expressed in the blastemal component. The epithelial components strongly expressed WT1, while the stromal components weakly expressed it (Figure 3c).

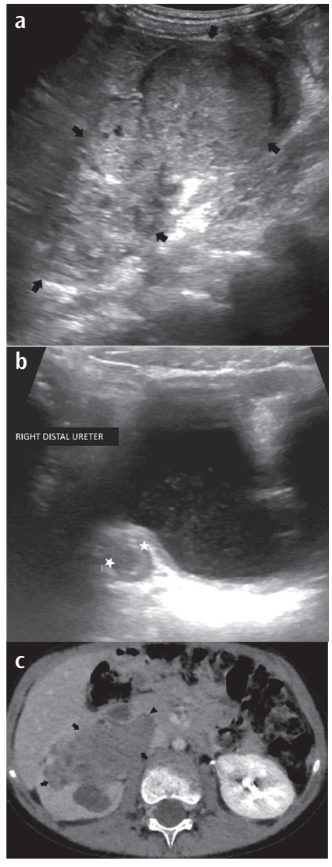

FIG. 1. (a) Ultrasonography of the abdomen showing a large hypoechoic heterogeneous mass originating from the right renal pelvicalyceal system (black arrow). (b) The mass extends into the distal ureter and bladder (stars). (c) Computed tomography showing a solid mass with heterogeneous enhancement in the pyelocaliceal system (arrows) extending into the proximal ureter (arrowhead) and displacement of the remaining thin renal parenchyma.

Corresponding author: Merve Yazol, Department of Radiology, Gazi University Faculty of Medicine, Ankara, Türkiye
e-mail: myazol@gmail.com

Received: September 27, 2023 **Accepted:** November 02, 2023 **Available Online Date:** February 29, 2024 • **DOI:** 10.4274/balkanmedj.galenos.2023.2023-9-93
Available at www.balkanmedicaljournal.org

ORCID iDs of the authors: M.Y. 0000-0003-1437-8998; M.A.İ. 0000-0002-6179-2828; Ö.L.B. 0000-0002-5200-1588.

Cite this article as: Yazol M, İnan MA, Boyunağa ÖL. Unexpected Radiologic Findings of a Botryoid Wilms Tumor with Extension into the Bladder. Balkan Med J.; 2024; 41(2):147-8.

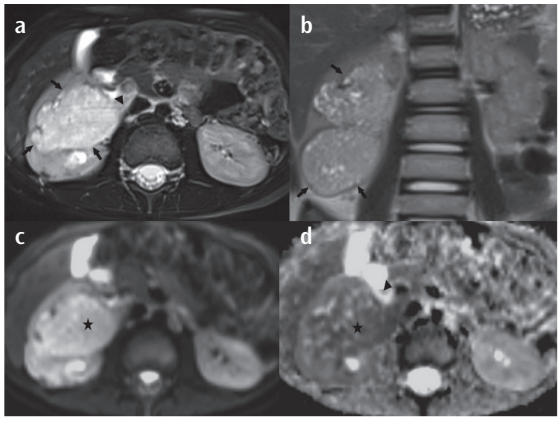

FIG. 2. (a) Axial T2-weighted image (WI) demonstrates a multilobulated soft-tissue mass (arrows) with high signal intensity in the dilated right renal pelvis, calyx, and proximal ureter (arrowhead). (b) In the coronal T2-WI, the mass (arrows) can be visualized occupying the renal pelvis and all the calyces, causing hydronephrosis. The lesion shows diffusion restriction, appearing hyperintense on the DWI (c) and hypointense on the ADC map (d) (star).

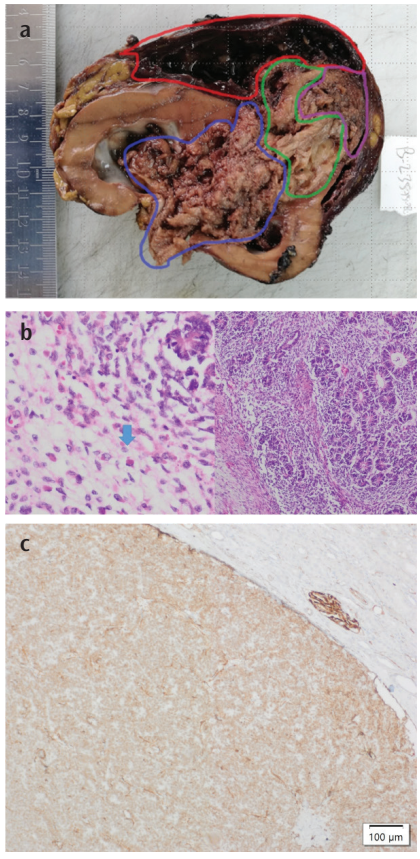

FIG. 3. (a) Gross pathological examination revealed a large tumor obliterating the renal collecting system, which was composed of fragile brown tissue, cystic hemorrhagic regions, and yellow necrotic areas. (b) Triphasic tumor depiction: The left side shows the stromal component containing rhabdomyoblasts (blue arrow), while the right side highlights both the epithelial and blastemal components. (c) Immunohistochemistry (x 100 magnification): Wilms tumor-1 expression in the tumor. Glomeruli are the internal positive controls.

The incidence of classic Wilms tumor is 10.4 cases in 1 million children aged < 15 years and 0.2 for every 10,000 infants. Approximately 10% of Wilms tumors demonstrate a distinct botryoid growth pattern into the renal pelvis and calyces, as seen in our patient.[4] The classic Wilms tumors have a 90% survival rate, and those with botryoid growth patterns have a favorable prognosis following complete excision without the need for additional radiotherapy or chemotherapy. National Wilms Tumor Study Group database demonstrates that only 2% of patients have ureteral extension, and even fewer tumors reach the bladder as was seen in our patient.[4] Radiological examination, particularly CT and MRI, play a crucial role in distinguishing between the classic and botryoid subtypes of Wilms tumor. The unique characteristics of a botryoid Wilms tumor, such as its origin in the renal pelvicalyceal system and extension into the ureter, help differentiate it from the classic type. In children aged < 5 years with gross hematuria and no inflammation, a pelvicalyceal lobulated mass, absence of parenchymal invasion, heterogeneous enhancement on CT and MRI due to varied tissue composition, and diffusion restriction, hydroureteronephrosis is a key radiologic sign for tumor differentiation.[1,5] Thus, a thorough radiological analysis is essential for the accurate diagnosis and differentiation of these tumor subtypes.

Informed Consent: Written consent was obtained from the patient.

Authorship Contributions: Design- M.Y., O.B., M.A.I.; Analysis or Interpretation- M.Y., O.B., M.A.I.; Writing- M.Y., O.B., M.A.I.

Conflict of Interest: The authors declare that there is no conflict of interest.

REFERENCES

1. Lowe LH, Isuani BH, Heller RM, et al. Pediatric renal masses: Wilms tumor and beyond. *Radiographics.* 2000;20:1585-1603. [CrossRef]
2. Tu BW, Ye WJ, Li YH. Botryoid Wilms' tumor: a report of two cases. *World J Pediatr.* 2005;21:43-46. [CrossRef]
3. Honda A, Shima M, Onoe S, et al. Botryoid Wilms' tumor: a case report and review of the literature. *Pediatr Nephrol.* 2013;11:1-5. [CrossRef]
4. Bahoush G, Pahlavani R, Salarinejad S, Zarei E. Bilateral Botryoid Wilms Tumor presented with acute renal failure. *Radiol Case Rep.* 2021;16:166-170. [CrossRef]
5. Park CJ, Im YJ, Shin HJ, Kim MJ. Botryoid Wilms' tumor in a child presenting with gross hematuria: a case report. *Journal of the Korean Society of Radiology.* 2016;75:198-202. [CrossRef]