









# Botryoid Wilms Tumor in a Neonate Presenting with Fetal Hydronephrosis: A Case Report

수신증으로 발견된 신생아의 포도상의 윌름스 종양: 증례 보고

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Botryoid Wilms tumor, a very rare variant of Wilms tumor, arises from the pelvocalyceal system, and its occurrence in the fetal or neonatal period has never been reported in the literature. Herein, we report an exceedingly rare and challenging case of botryoid Wilms tumor in a neonate who initially presented with fetal hydronephrosis. Postnatal ultrasonography revealed multiple lobulating hypoechoic masses with varying degrees of intralesional vascularity within the dilated pelvocalyceal system. To our knowledge, this is a case report of botryoid Wilms tumor of the youngest child in English literature.

**Index terms** Hydronephrosis; Magnetic Resonance Imaging; Wilms Tumor; Neonate; Ultrasonography

## INTRODUCTION

Although Wilms tumor (WT) which typically originates from the renal parenchyma is the most common renal neoplasm in childhood, its occurrence in neonatal period is extremely rare, accounting for 0.16% of all WT cases (1). Botryoid WT arises from the pelvocalyceal wall with pathologic appearance similar to botryoid sarcoma. As a variant of WT, it is very rarely reported in children beyond neonatal period (2-5).

Here, we report an exceedingly rare case of botryoid WT diagnosed in a neonate presented with fetal hydronephrosis with imaging features on various modalities, especially its sonographic findings.

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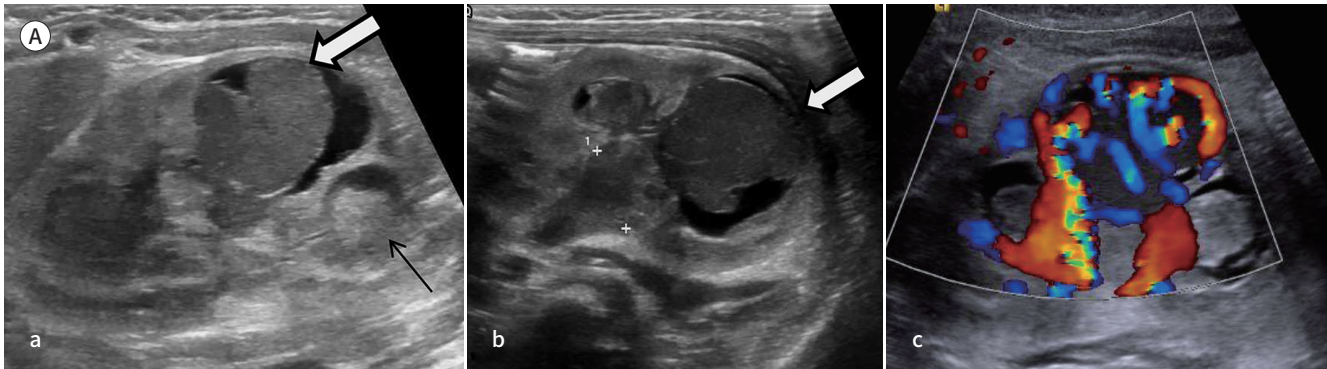
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**Fig. 1.** A 7-day-old male neonate with botryoid Wilms tumor.

**A.** Longitudinal (**a**) and transverse (**b**) ultrasound scans of the left kidney reveal multi-lobulated and irregular contoured hypoechoic masses filling the dilated collecting system with severe parenchymal thinning. Some hyperechoic nodular lesions (thin arrow in **a**) are also seen in the lower pole calyx. The intracalyceal mass of the mid-pole (thick arrows in **a** and **b**) shows increased vascularity in color Doppler ultrasonography (**c**), while other lesions within the pelvis and calyces do not show vascularity. The calipers (in **b**) indicate the anteroposterior dimensions of the renal pelvis occupied by mixed echoic lesions.



## CASE REPORT

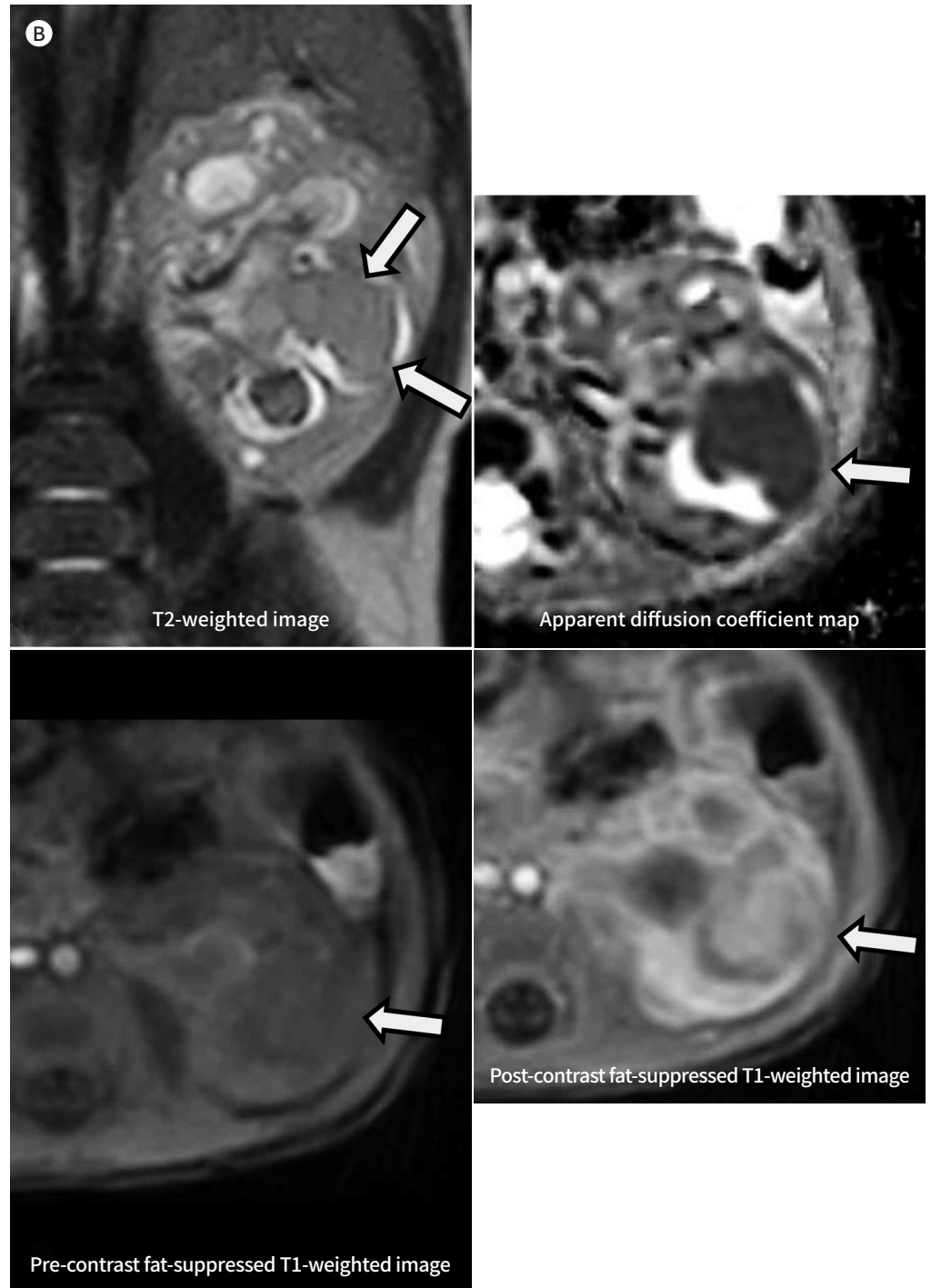
A 7-day-old male neonate born as a full-term baby without specific medical or family history presented for further evaluation of fetal hydronephrosis. Urinalysis results showed 11–20 white blood cells per high-power field (HPF) and 0–2 red blood cells per HPF. On peripheral blood analysis, results were unremarkable.

Ultrasonography (US) revealed lobulating contoured hypoechoic masses filling and distending the pelvocalyceal system of the left kidney associated with severe parenchymal thinning (Fig. 1A). Color Doppler study showed abundant intralesional vascularity of the mass in the mid-pole calyx. No vascularity was seen in other mass lesions of the renal pelvis or calyces of upper and lower pole. There was no gross involvement of the renal parenchyma by masses nor extension into the ureter. When compared with fetal US at 35 weeks of gestation (not shown here), these mass-like lesions increased in the extent growing into calyces. They had initially occupied the renal pelvis, causing hydronephrosis. Presence of hypervascular solid mass and rapid growing for a relatively short term raised a concern for malignant tumor without clinical evidence of urinary tract infection. Urine cytology also revealed cluster of small round cells with marked degeneration suggestive of neoplasm.

MRI with contrast enhancement demonstrated a multilobulated polypoid lesion in the dilated calyx of the mid pole extending the renal pelvis with intermediate signal intensity both on T1- and T2-weighted images (Fig. 1B). Other lesions filling the upper and lower pole calyces showed high or low signal intensities on T2 and T1-weighted images. The mass in the mid-pole showed strong enhancement after gadolinium injection while other lesions in the upper and lower pole showed no contrast enhancement, suggesting hematoma or necrotic change. On diffusion-weighted image, the mid-pole lesion demonstrated diffusion restriction while others did not. Based on these imaging findings, a malignant renal tumor arising from pelvocalyceal system was suggested, with differential diagnosis including botryoid WT, sarcoma botryoides, and rhabdoid tumor. Work up for distant metastasis including chest CT and brain US was negative.

**Fig. 1.** A 7-day-old male neonate with botryoid Wilms tumor.

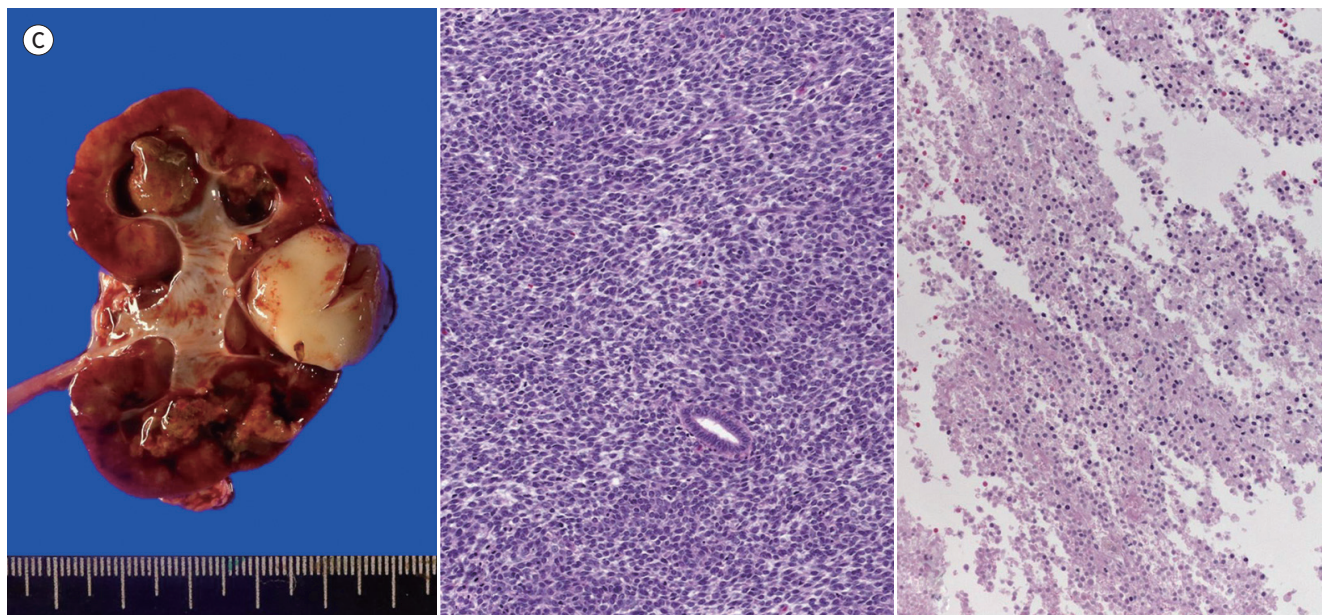
**B.** Coronal T2-weighted image (right upper box) demonstrates a multilobulated polypoid mass (arrows) with intermediate signal intensity in the dilated calyx of the mid-pole. Other lesions filling the upper and lower pole calyces show high or low signal intensities. On apparent diffusion coefficient map (left upper box), the mass (arrow) demonstrates diffusion restriction in comparison with the diffusion weighted image (not shown here). Pre- and post-contrast axial fat-suppressed T1-weighted images (lower boxes) at the mid-pole level demonstrate avid enhancement of the intracalyceal mass (arrows). Other lesions in the collecting system showed no contrast enhancement (not shown here).



**Fig. 1.** A 7-day-old male neonate with botryoid Wilms tumor.

**C.** On a cut section of the kidney, a yellowish soft-to-firm solid mass is present in the calyx of the mid-portion, and several dark reddish friable masses are identified in the upper and lower calyces (left image). Microscopically, the yellowish solid mass is composed of round to ovoid blue blastemal cells with a few primitive tubules (H&E stain,  $\times 200$ ) (middle image). The reddish friable mass is mostly necrotic with some pyknotic neoplastic cells (H&E stain,  $\times 200$ ) (right image).

H&E = hematoxylin & eosin



Radical nephroureterectomy was performed. Gross specimen revealed a well-demarcated, bulging, yellowish, solid mass in the calyx of the mid-portion and several dark reddish friable masses in the upper and lower calyces (Fig. 1C). Microscopically, the solid mass of mid-pole calyx was composed of blastemal cells while reddish friable masses were mostly necrotic with some pyknotic neoplastic cells. No ureteral involvement was noted. Histopathologic examination confirmed the diagnosis of botryoid WT with positive WT-1 (Wilms tumor 1 protein). Immunohistochemistry for CD99, desmin, and CD56 in the small round cell and ETV6 gene study were all negative (Fig. 1C). The patient was subsequently treated with post-operative adjuvant chemotherapy. He has been followed up without recurrence or distant metastasis for one year.

## DISCUSSION

The presented neonatal case of botryoid WT suggests its congenital origin causing fetal hydronephrosis, which has never been reported in the literature. In general, renal tumors detected in neonates are very rare. Congenital mesoblastic nephroma (CMN) is the most common primary renal neoplasm in neonates and infancy (5). CMN usually presents as a homogeneous and slightly hyperechoic renal mass located near the renal hilum with renal parenchymal involvement, although cellular variant CMN, similar to fibrosarcoma, can be presented as a heterogenous renal mass with frequent necrosis and hemorrhage. The occurrence of WT is extremely rare in neonates, representing only 0.16% of all WT cases (1). Rhab-

doid tumor which is a highly malignant and aggressive renal tumor may also occur (although very rarely) in neonates with similar imaging features to WT and synchronous or metachronous brain lesion can be present. Ossifying renal tumor of infancy which is typically presented as a calcified renal pelvis mass in infancy should also be included in differential diagnosis of renal mass developed in infants. Although ossifying renal tumor of infancy may present as extensive tumor calcifications, gross calcification may not be noted in relative young ages (6).

In contrast to WT which originates from the renal parenchyma, botryoid WT arises within the pelvocalyceal wall and occupies the collecting system with pathologic appearance similar to botryoid sarcoma (2). It has been reported very rarely as a case report in children aged from 4 months to 8 years (19 males, 7 females), with 14 occurring in the right, 9 in the left, and 3 in both collecting systems. They almost always present with hematuria and frequently extending to the ureter and bladder (2-4, 7). Although our case initially showed no gross hematuria, hematuria with blood clot developed during admission.

Sonographic finding for botryoid WT has been reported as a renal pelvic mass with mixed echogenicity associated with hydronephrosis (2, 7). Our presented case also showed multilobulated hypoechoic masses filling the calyces which well corresponded with those of previous reports. Necrosis or hemorrhage has been reported in previous cases like our case. US without radiation nor sedation clearly showed the extent and characteristics of the renal mass. Furthermore, color Doppler study is very useful in discriminating tumor from non-tumorous conditions such as fungal ball or hematoma which usually does not demonstrate vascularity. In particular, fungal ball in an appropriate clinical setting should be included for differential diagnosis of intrapelvic or intracalyceal mass-like lesions with hydronephrosis. Regarding color Doppler study features of botryoid WT, Park et al. (3) have described that there is no demonstrable vascularity in the mass, hence mimicking a blood clot rather than tumorous condition. However, in our study, the lesion in the mid-pole showed abundant vascularity within the mass while there was no vascularity for lesions in upper and lower poles (Fig. 1A). It turned out to be necrotic tissue. Sonographic findings also correlated well both with CT (not shown here) and MRI including diffusion-weighted image.

According to National Wilms Tumor Study/Children's Oncology Group, of stage I groups, patients under age 2 with favorable histology and tumor weighing less than 550 g are considered as very low risk group. They can be managed by nephrectomy alone without adjuvant chemotherapy (8). Although the tumor in our case was confined in the unilateral kidney, possibility of microscopic seeding through the urinary tract including bladder could not be excluded due to tumor location and positive urine cytology. Therefore, adjuvant chemotherapy after nephroureterectomy was performed like cases described in previous reports. In previous cases, radiation therapy was added in two cases: one case with the bilateral involvement and another case with extension to the bladder (4).

In conclusion, we report the youngest case of botryoid WT presented with fetal hydronephrosis. As our case shows, congenital presentation of this rare subtype of WT is possible, necessitating careful evaluation of any presence of mass lesion within the collecting system in cases of fetal hydronephrosis.

### Author Contributions

Conceptualization, Y.S.; data curation, Y.S., K.J., B.M.; formal analysis, K.C.H., K.J.; investigation, Y.S., K.C.H.; methodology, Y.S., K.C.H.; supervision, K.J.H., J.T.Y.; validation, K.J.H., J.T.Y.; visualization, K.C.H., K.J.; writing—original draft, K.C.H.; and writing—review & editing, Y.S.

### Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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This case study was approved and obtaining informed consent was waived by the Institutional Review Board of our medical center.

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## 수신증으로 발견된 신생아의 포도상의 윌름스 종양: 증례 보고

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포도상의 윌름스 종양은 신우에서 발생하는 윌름스 종양의 희귀한 변이형이며 태아 혹은 신생아에서 진단된 경우는 아직까지 보고된 바 없다. 저자들은 산전 초음파에서 우연히 발견된 수신증으로 내원한 신생아의 포도상의 윌름스 종양을 경험하여 이를 보고하고자 한다. 출생 후 초음파 검사에서 확장된 신우 및 신배를 채우는 다수의 소엽상의 저에코 종괴가 관찰되었고 종괴 내부는 다양한 정도의 혈류 증가 소견을 보였다. 저자들이 아는 한, 영문으로 보고된 가장 어린아이에서 발생한 윌름스 종양의 증례이다.

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