

## Pediatrics

## Pelvic neuroblastoma presenting with acute urinary retention and acute kidney injury

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## Introduction

Neuroblastoma is the most common extracranial solid tumor in children under the age of 5 years and is the second most prevalent malignancy in children after acute lymphoblastic leukemia.<sup>1</sup> As this embryonal malignancy arises in cells derived from the neural crest, the primary tumor may initiate where sympathetic tissue is located between the neck and pelvis.<sup>2</sup> Only 2.2% of neuroblastomas are located in the pelvic region and often have a good prognosis.<sup>3</sup> Acute urinary retention due to extrinsic compression from a pelvic mass is a rare phenomenon in infants.<sup>1</sup> We report the first case of a ten-month-old girl who was diagnosed with a pelvic neuroblastoma causing acute urinary retention and acute kidney injury. The detection and management of pelvic neuroblastoma and acute kidney injury are discussed.

## Case report

A ten-month-old girl (length: 68.6 cm; weight: 9.36 kg) presented to the Emergency Department with decreased urine output and oral intake, fussiness, and one episode of emesis one day earlier. She had also experienced diarrhea for several days. There was no fever. A palpable mass was noted in the right abdominal area. Past medical history was negative for urinary tract infection, urological disease, or prenatal hydronephrosis.

A urinalysis revealed 10 mg/dL protein, "large" leukocyte esterase, nitrite negative, 11 WBCs/HPF, and 1 RBC/HPF. A renal function panel was significant for sodium 140 (132–142) mmol/L, potassium 5.8

(3.5–5.1) mmol/L, chloride 105 (98–107) mmol/L, CO<sub>2</sub> 12 (23–29) (mmol/L), blood urea nitrogen (BUN) 52 (4–15) mg/dL, creatinine 1.5 (0.3–0.5) mg/dL, phosphorus 9.2 mg/dL, and C-reactive protein 4.2 (< 1.0) mg/dL. A complete blood cell count showed WBCs 19.64 (6.0–17.5) × 10<sup>3</sup>/μL (neutrophilic predominance 69.9%), hemoglobin 10 (10.5–13.5) g/dL, MCV 81, and platelets 422 (140–440) × 10<sup>3</sup>/μL.

An abdominal ultrasound was significant for distention of the urinary bladder and renal pelvocaliectasis bilaterally. A pelvic ultrasound demonstrated a large soft tissue mass (7.5 × 4.1 × 4.8 cm) in the left pelvis with internal vascularity and numerous scattered calcified foci (Fig. 1A). A pelvic CT scan revealed a large heterogeneous presacral mass invading the sacral spinal canal and the left posterior paraspinous soft tissues (Fig. 1B). Bladder outlet obstruction from the tumor with marked circumferential bladder wall thickening and moderate bilateral hydronephrosis was observed. The rectum was also partially obstructed, with fecal stasis in the colon. No distant metastatic disease was apparent. A lumbar MRI scan confirmed a large presacral mass extending into and filling the spinal canal between S1–S5 via the neural foramen S2–S3, S3–S4, and S4–S5 on the left and S3–S4 on the right. Small contiguous extension posterior to the spinal canal at S1–S3 on the left was appreciated.

The patient underwent urinary catheterization with 260 mL of clear urine and subsequent Foley catheter placement. A biopsy of the presacral mass was performed on the fourth hospital day. Histological examination demonstrated tumor cells with scant cytoplasm which were PGP9.5+, Synaptophysin+, and Chromogranin+. Less than 5% of the tumor cells exhibited differentiation. There was abundant

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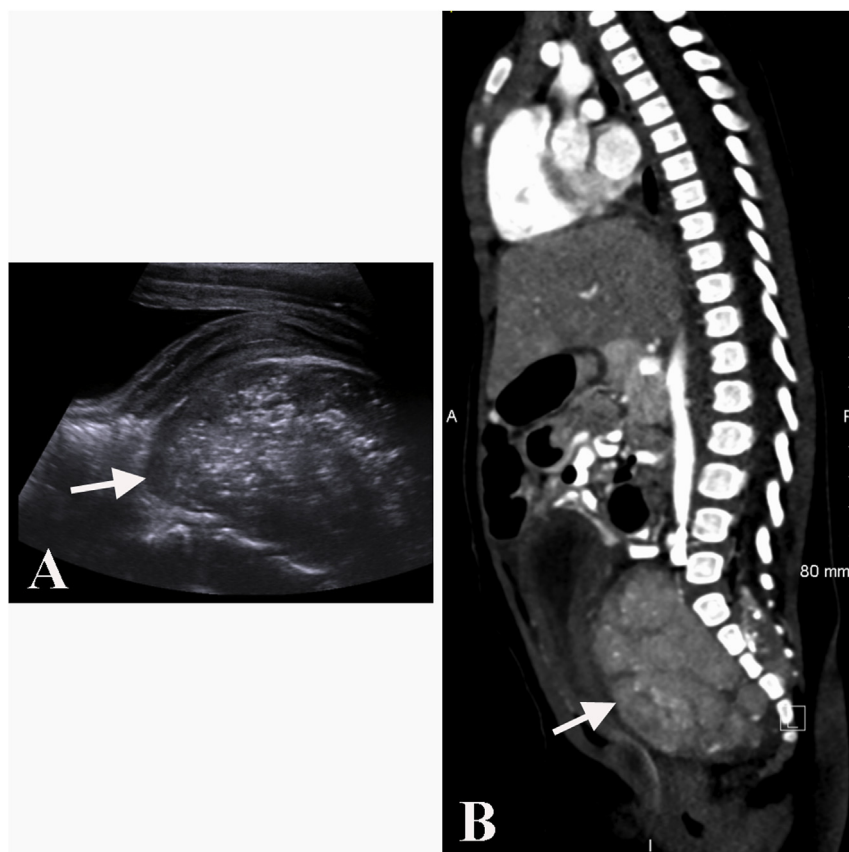
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**Fig. 1.** (A) A pelvic ultrasound demonstrated a large soft tissue mass (arrow) in the left pelvis with internal vascularity and numerous scattered calcified foci. (B) A CT scan revealed a large heterogeneous presacral mass (arrow) invading the sacral spinal canal and the left posterior paraspinal soft tissues.

neuropil but scant S100 + Schwannian stroma and a low mitotic-karyorrhectic index. Undifferentiated neuroblastoma was excluded due to the abundant neuropil, and ganglioneuroblastoma was ruled out due to the paucity of Schwannian stroma or cell maturation. The histological findings were consistent with a neuroblastoma, MYCN negative.

The patient was discharged on the 15th hospital day with the Foley catheter in place. The patient was evaluated by our urology service two weeks after hospital discharge, at which time there was no evidence of hematuria or decreased urine output. Clean intermittent catheterization was encouraged for management of the urinary retention. She initiated treatment for intermediate-risk neuroblastoma per ANBL0531. She underwent two cycles of chemotherapy consisting of carboplatin/cyclophosphamide/doxorubicin followed by four cycles of carboplatin/doxorubicin/etoposide.

A CT angiogram of the abdomen and pelvis three months after her biopsy demonstrated significant mass effect with bilateral hydro-ureteronephrosis. She underwent resection of the neuroblastoma two months later.

## Discussion

Neuroblastomas have been referred to as mimickers of other malignancies due to their diverse clinical behavior.<sup>1,2</sup> Patients may present with benign disease with spontaneous regression or metastatic and rapidly fatal disease.<sup>3</sup> Early detection and surgical resection are associated with improved survival.<sup>3</sup>

Patients with neuroblastomas often have nonspecific urinary signs and symptoms which pose a challenge to a timely and accurate diagnosis. Neuroblastomas are not commonly included in the differential diagnosis of acute urinary retention which usually encompasses urinary tract infections/stones, neurologic disorders such as tumors,

constipation, and voiding dysfunction.<sup>1,4</sup> Acute kidney injury refers to an abrupt decrease in kidney function, marked by both injury (structural damage) and impairment (loss of function).<sup>5</sup> Patients with acute kidney injury may exhibit sepsis, ischemia, and nephrotoxicity. Neuroblastomas have rarely presented with acute kidney injury and proteinuria.<sup>2</sup>

The current Children's Oncology Group (COG) protocol ANBL0531 refers to the Response- and Biology-based Therapy for Intermediate-risk Neuroblastoma.<sup>6</sup> This protocol provides the minimal therapy needed to achieve excellent outcomes for patients with intermediate-risk neuroblastoma. The chemotherapy consists of cycles of cyclophosphamide, doxorubicin, carboplatin, and etoposide every 3 weeks. Patients with an intermediate-risk neuroblastoma are categorized based on clinical and biologic factors, with a reduction in therapy as the overall goal.

Our case represents the first in the literature of an infant presenting with both acute urinary retention and acute kidney injury and subsequently diagnosed with pelvic neuroblastoma. Her acute kidney injury was manifested by the increased BUN and creatinine, hyperkalemia, and metabolic acidosis. Furthermore, the infant's elevated BUN and creatinine may reflect her urinary tract obstruction due to the neuroblastoma combined with dehydration from emesis and diarrhea. Pediatric urologists should consider a pelvic neuroblastoma when an infant has decreased urine output and oral intake, an abdominal mass, proteinuria, abnormal renal function, and acute urinary retention. A comprehensive history and physical examination, urinalysis, and imaging studies such as an ultrasound, CT scan, and MRI scan are mandatory in cases of acute urinary retention and may uncover a rare pelvic mass requiring surgical intervention, radiation, and/or chemotherapy.

### Informed consent

The patient's parents provided written consent to the publication of the photos and details of the case. The Chair of the University of Louisville Institutional Review Board (IRB) determined that this study does not meet the “Common Rule” definition of human subjects' research. Therefore, our project did not require IRB review. The IRB number was 18.0857.

### Declarations of interest

None.

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