Clinical Imaging

Congenital nephrogenic diabetes insipidus presenting with bilateral hydronephrosis and dilatation of the ureter and bladder

A 21-year-old man was admitted to our emergency department with complaints of dysuria. Computed tomography showed bilateral hydronephrosis and dilatation of the ureter and bladder (Fig. 1). Blood laboratory data indicated mild renal dysfunction (creatinine 1.18 mg/dL). His medical history included polyuria and polydipsia. His symptoms improved dramatically after urine drainage. Treatment with hypertonic saline and desmopressin failed to increase urine osmolality. Considering these results and his family history (his uncle and nephew were already diagnosed with nephrogenic diabetes insipidus [NDI]), he was diagnosed with congenital NDI. Congenital NDI is a rare disorder resulting in a decrease in urinary concentrating ability that results from resistance to the antidiuretic hormone. A mutation in the x-linked vasopressin V2 receptor (*V2R*) gene is the most common cause of congenital NDI (approximately 90%).[1,2] Patients with hydronephrosis resulting from congenital NDI are rare in the emergency department.[2–5]

DISCLOSURE

Approval of the research protocol: N/A.
Informed consent: Informed consent was obtained from the patient.

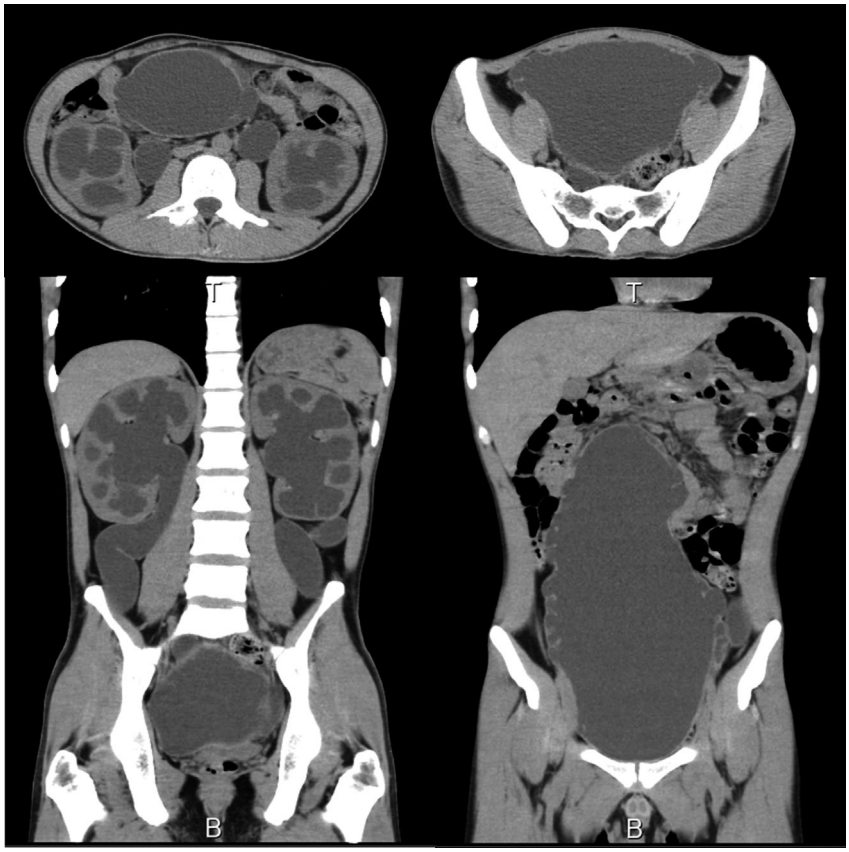

Fig. 1. Computed tomography scans of a 21-year-old man diagnosed with congenital nephrogenic diabetes insipidus, showing bilateral hydronephrosis and dilatation of the ureter and bladder.

Registry and registration no. of the study/trial: N/A.
Animal studies: N/A.
Conflict of interest: None.

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