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Case Report

Anuria in a 9-Month-Old Infant Resulting from Ureteral Cystine Stones

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Pediatric urolithiasis and calcular anuria in early infancy are rare. Cystine stones may develop in utero or during early infancy. We report the case of a female 9-month-old infant with obstructive anuria resulting from cystine stones in a single functioning unit. She presented to the emergency department owing to the absence of micturition for 3 days. Radiological investigations revealed four left ureteral stones and an atrophic right kidney resulting from a calcular obstruction. Her laboratory values were as follows: serum creatinine 6.7 mg/dl, Na 132 mEq/l, K 6 mg/dl, and hematocrit 32%. An urgent percutaneous nephrostomy tube was inserted into the left side for urinary drainage, and her serum levels of creatinine and K returned to normal within 3 days. A left ureterolithotomy was the final management. Stone analysis revealed pure cystine crystals.

Key Words: *Anuria; Infant; Urolithiasis*

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Pediatric urolithiasis is a relatively rare disease [1]. Anuria in small babies usually has a prerenal cause. Calculus anuria in infants and early childhood is rare but should be considered. The management of pediatric urolithiasis should be individualized with careful consideration of the patients' small body size, delicate tissues, needs for general anesthesia, and risks of long-term complications [2]. We present the case of a 9-month-old female infant who had multiple bilateral ureteral cystine stones. The stones obstructed the middle ureter in a single functioning kidney, causing acute renal failure. The aim of this report is to increase awareness of urinary stones in infants causing obstructive anuria.

CASE REPORT

We report the case of a female 9-month-old infant who was presented to the emergency department because of the absence of micturition for 3 days. The pregnancy and delivery history was normal. Clinical examination showed no congenital abnormalities. The infant's body weight was 7.5 kg. Neither the bladder nor the kidneys were palpable clinically. The infant was quiet in bed. A Foley urethral catheter was inserted and revealed no urine in the bladder. Serum creatinine was 6.7 mg/dl, serum K was elevated to 6 mEq/dl, Na was 132 mEq/dl, and hematocrit was 32%. Abdominal ultrasound showed mild hydronephrosis of the left kidney (Fig. 1A). The right kidney was atrophic with poor corticomedullary differentiation and stones (Fig. 1B). Kidney, ureter, and bladder showed four radiopaque shadows opposite the left sacroiliac joint and one in the renal area on the right side. Moreover, another radiopaque shadow was found at the level of L3 on the right side (Fig. 2). A percutaneous nephrostomy tube was inserted into the dilated left pelvicaliceal system for drainage for several days. After the intervention, the serum creatinine level improved to 1.6 mg/dl and K returned to a normal level on the third day. Exploration of the left middle third of the ureter was performed with the removal of four stones and a double (DJ) stent was fixed. Creatinine returned to 0.5 mg/dl after 2 weeks. The stent was removed after 2 weeks. Chemical analysis of the stones revealed pure cystine crystals. Right kidney function will be evaluated by [DMSA] isotopes. A right nephrectomy may be considered because of the stones and infection. Metabolic workup for cystinuria was performed via a positive nitroprusside test. The diagnosis was

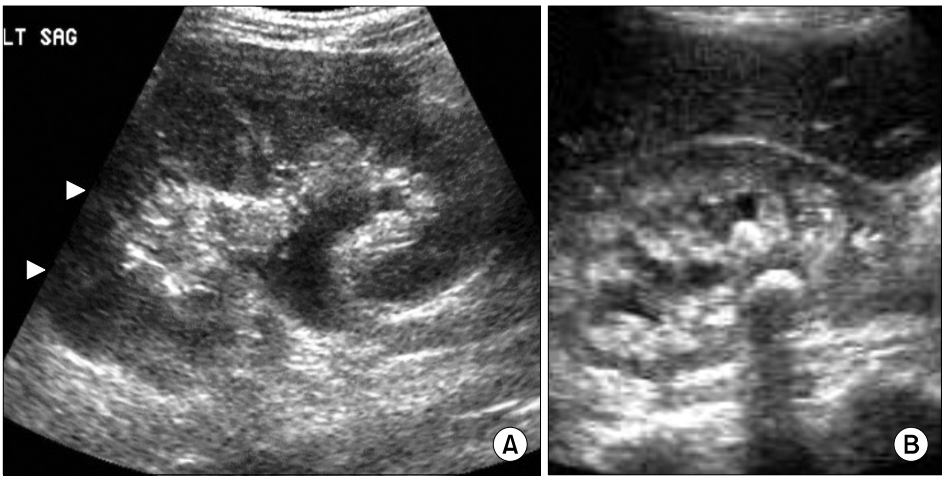

FIG. 1. (A) Preoperative ultrasonography of the left kidney showing mild hydronephrosis due to ureteral stones with good corticomedullary differentiation. (B) Preoperative renal ultrasonography showing right atrophic hyperechoic kidney with loss of corticomedullary differentiation associated with stones.

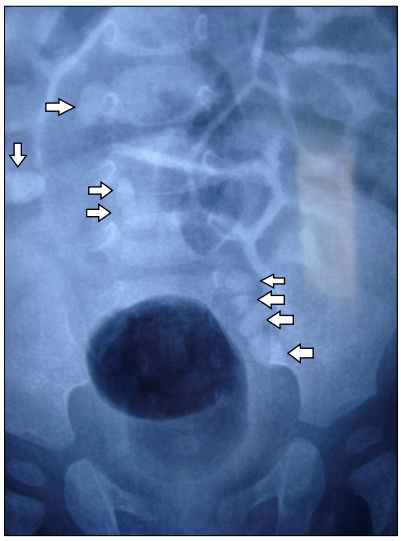

FIG. 2. Preoperative plain abdominal X-ray kidney, ureter, and bladder showing multiple bilateral calculi (arrows).

confirmed by stone analysis by use of chemical methods. Three months later, the results of a renal ultrasound and renal function tests with urine analysis were repeated and were shown to be normal.

A follow-up protocol was given to the parents to prevent the recurrence of stones. It consisted of urine analysis every 2 months and serial ultrasonography for early detection of stone formation every 3 months. Plain abdominal radiography was not necessary. A detailed feeding history of the patient was reevaluated during each visit by the pediatrician. The parents were advised to ensure constant, ample fluid intake during both daytime and nighttime. Potassium citrate (1-1.5 mEq/kg per day) or a mixture of potassium citrate and sodium citrate (Polycitra) was administered with monitoring of urinary pH. Penicillamine was avoided because of its serious side effects, such as hypertension, heart failure, renal failure, and electrolyte imbalance.

DISCUSSION

Stone disease is a rare condition below 1 year of age. The frequent use of ultrasound in the pediatric population associated with more awareness of stones has increased the incidence of stones in recent publications. Predisposing factors for stone formation are genetic inheritance, poverty and poor nutritional status, metabolic abnormalities, and environmental factors such as hot weather during the summer associated with less drinking of water [1]. How early the stone formation begins antenatally, during the newborn period, or in infancy is not clear [2]. Infantile anuria is usually due to prerenal causes such as gastroenteritis, hypotension, and hypoxia. Others have reported cases resulting from ureteropelvic junction obstruction, a blood clot in the renal pelvis of a single kidney, trauma, fungal bolus obstruction, and avulsion of the ureter [3]. Cystine stones account for 6% of pediatric stones [4,5]. They occur only in some cystinuric patients who may develop them either intrauterine or during infancy. Cystine is the least soluble of the urinary amino acids. A recessive genetic abnormality in the renal tubular reabsorptive transport of cystine results in high concentrations of cystine crystals in urine. The crystals tend to precipitate when present in excessive concentrations [5]. Calcular obstructive anuria should be considered in an anuric infant. Few publications have reported pediatric anuria secondary to obstructing cystine stones [5-8]. In this case, right renal and ureteral stones might explain the early obstruction of the right kidney in utero or during early infancy, followed by scarring and nephropathy with complete loss of function. The relative rarity of cystine stones and the association of obstruction bilaterally or unilaterally in a single functioning unit in infants within the first year of life may explain the shortage of publications.

Percutaneous nephrostomy was very useful in the presenting case for preoperative drainage and to improve the patient's general condition with stabilization of serum creatinine and potassium levels. Open ureterolithotomy was performed because of the multiplicity of the stones, small

infant size, increasing serum creatinine and K, and inappropriateness of ureteroscopy or shockwave lithotripsy. A DJ ureteral stent was fixed because of fear of postoperative edema and to remove the nephrostomy tube because it is difficult to maintain tube care in this age group of children.

The differential diagnosis of pediatric anuria should include bilateral obstructive urolithiasis. Calcular obstructive anuria due to cystine stones in early infancy is extremely rare but should be considered. Early recognition of the problem and mention of both stone formation and its recurrence should be goals of pediatricians and urologists.

Conflicts of Interest

The authors have nothing to disclose.

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