

Pediatrics

Pure xanthine pediatric urolithiasis: A cause of acute renal failure

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ARTICLE INFO

Keywords:

Xanthine urolithiasis
Acute renal failure
Pediatrics urolithiasis

ABSTRACT

Xanthine urolithiasis is usually a benign condition, easy to prevent or cure by appropriate alkalinization, forced hydration and restriction of dietary purines if diagnosed early. When unrecognized, xanthinuria can lead to end-stage renal failure or nephrectomy.

Introduction

Nephrolithiasis has become an important cause of childhood morbidity and health care expenditure worldwide.¹ Although considered an adult disease in the past, in the last few decades, it has become increasingly prevalent in children with the incidence increasing by approximately 6–10%.²

Nephrolithiasis is caused by anatomic, metabolic or environmental factors. 40–50% of children with stones have an associated metabolic abnormality. The recurrence rate is between 16 and 67% and is usually due to metabolic abnormalities.^{3,4} Although the exact mechanism of urolithiasis is not known, the most commonly accepted theory is supersaturation, crystallization theory. According to this theory, as the concentration of solutes in urine increases, the solubility product is reached; above which dissolved solutes can form nuclei of its solid phase (the metastable zone). These nuclei can form homogeneously or heterogeneously.³

Xanthinuria type I, a rare hereditary disorder, is characterized by a marked and isolated deficiency of xanthine oxidase activity in the tissues, resulting in hypouricemia and hypouricosuria. Total urinary purine excretion is not increased, whereas excretion of xanthine and hypoxanthine is considerably increased as a response to the elevated serum levels of these oxypurines. Clinical symptoms may include mild abdominal pain, hematuria, dysuria and in severe cases azotemia leading to acute renal failure.

We present a case of classical xanthinuria leading to renal failure in a child, emphasizing the early diagnosis and treatment to prevent further renal damage.

Case report

An 18-month-old boy was referred to our Centre for evaluation of urolithiasis. He was born at term from apparently healthy consanguineous parents (first cousins), and he had no familial history of renal stone disease. His psychomotor development and physical growth was normal. The patient presented with a five-day history of reduced urine production, decreased alertness, peripheral edema, dry mouth and rapid pulse secondary to bilateral obstructive uropathy.

His hematological investigations reveal markedly raised total leukocyte count $22 \times 10^9/L$ and serum creatinine of $817 \mu\text{mol/L}$. Since the patient is in acute renal failure therefore he underwent multiple sessions of peritoneal dialysis under the care of the pediatric nephrology team. Further investigations revealed left mid-ureteric calculus and right renal pelvis calculus causing complete obstruction of bilateral renal tract resulting in acute renal failure and severe azotemia [Fig. 1].

The case was discussed in the multidisciplinary team meeting and it was decided to relieve obstruction from the left side of ureter via therapeutic ureteroscopy or left ureterolithotomy first due to large size ureteric calculus followed by right percutaneous nephrolithotomy (PCNL).

Initially, urine is alkalinized by potassium citrate/bicarbonate in 3–4 divided doses. Left therapeutic ureteroscopy was planned in general anesthesia. The patient was positioned in lithotomy position but urologist was unable to negotiate guidewire beyond the calculus at the level of the ischial spine. Therefore, it was decided to perform left ureterolithotomy. Patient positioned and draped. Gridiron incision given, external and internal oblique muscles were incised. Ureter located extra peritoneal and extensive adhesions encountered. A large ureteric

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<https://doi.org/10.1016/j.eucr.2020.101438>

Received 7 September 2020; Received in revised form 22 September 2020; Accepted 29 September 2020

Available online 1 October 2020

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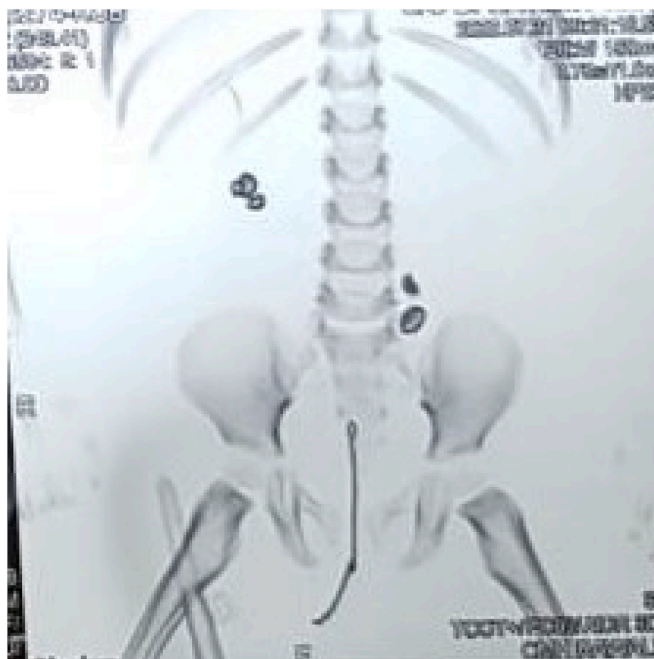


Fig. 1. Computed tomography shows a right renal pelvis calculus and left ureteric calculus.

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Entered At: 06 Aug 2020 10:18
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Lab ID: [REDACTED] Unit: [REDACTED] Panel
Patient:*** [REDACTED]

Stone Analysis
Comment Xanthine 100%. Kindly visit Endo clinic , AFIP with all medical documents.

Fig. 2. Stone analysis reveals 100% pure xanthine calculi.

calculus was removed via ureterotomy. DJ stent placed after checking both proximal and distal patency and confirmed under a fluoroscope. Ureter repaired with PDS 4.0 and the wound was closed in layers.

Right PCNL was performed after one week, lower pole posterior calyx was punctured. Single-step 15Fr tract dilatation done. Calculi were completely fragmented and cleared. Calculus sent for analysis after both surgical procedures revealed 100% xanthine calculi [Fig. 2]. Post-operative recovery was uneventful and the patient was discharged on 3rd postoperative day.

To prevent recurrence, frequent clinical, radiological and laboratory surveillance is advised. The diet should be low in protein and sodium chloride, check urinary pH level and first morning urine for cystine crystals. Regularly check renal functions. Ultrasonography renal and KUB should be routine. Annually 24-h urine testing and multidisciplinary approach involving nephrologist and renal dieticians.

Discussion

Urolithiasis in the pediatric age is related to significant morbidity since renal stone tend to recur and they should not be underestimated.

Sign and symptoms in pediatric groups include gross or microscopic hematuria, urinary tract infections, vague abdominal pain occurs in 10–14% of all pediatric cases.⁵

Xanthinuria is a rare autosomal recessive disorder caused by a mutation in xanthine dehydrogenase gene leading to deficiency or decrease activity of an enzyme. The plasma uric acid is very low and even replaced by xanthine and hypoxanthine. Pure xanthine stones are radiolucent. The diagnosis is based on stone analysis revealing 100% pure xanthine stone in xanthine urolithiasis.

Different management options depend upon the size and location of stone including open pyelolithotomy, nephrolithotomy, ureterolithotomy, vesicolithomy and minimal invasive and endoscopic surgeries like percutaneous nephrolithotomy, retrograde intrarenal surgery and cystolitholapaxy.

Conclusion

The prevention of xanthine stone includes intake of plenty of oral fluids and low purine diet. Alkalinization of urine increase solubility of xanthine thus preventing stone formation. Increased citrate excretion

induced by oral potassium citrate administration prevents the stone formation. Early diagnosis and management is the key to prevent long term complications.

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