CASE REPORT

Bilateral supernumerary kidneys: how much is too much?

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SUMMARY

A middle aged African-American woman with a stable history of carnitine palmitoyl transferase II (CPT II) deficiency presented with myalgias for 1 week. Physical examination and laboratory findings were consistent with severe sepsis secondary to pyelonephritis leading to rhabdomyolysis. Subsequent CT of the abdomen revealed bilateral supernumerary kidneys with non-obstructive calculi within the supernumerary kidneys. Abnormal ureteral development of the supernumerary kidneys likely led to an increased risk for urinary tract infections (UTIs) and renal calculi resulting in pyelonephritis. The stress of this infection overwhelmed the muscle CPT II enzyme load, putting her in a state of rhabdomyolysis. In addition to fluids and antibiotics, she was provided a diet rich in carbohydrates and low in fats so as to limit long-chain fatty acid oxidation. Supernumerary nephrectomy was not considered during this admission. During follow-up, she developed obstructive ureteral calculi requiring placement of a right-sided ureteral stent.

rhabdomyolysis by a mechanism of impaired long-chain fatty acid oxidation during periods of stress in which there is increased muscle and lipid metabolism. In the setting of CPT II deficiency and concomitant infection, abnormal lipid metabolism results in an excess of β oxidation intermediates within the mitochondria that subsequently damage mitochondrial membranes and sarcoplasmic reticulum. This, in turn, destabilises membrane-bound calcium channels resulting in prolonged muscle contraction and rhabdomyolysis.² The risk of CPT II deficiency-related rhabdomyolysis increases significantly with the presence of supernumerary kidneys and their predilection for development of urinary tract infections (UTIs) and renal calculi.

CASE PRESENTATION

A middle-aged African-American woman presented with low back pain and diffuse myalgias. She was unable to perform activities of daily living for 1 week. Her medical history was significant for obesity, nephrolithiasis and CPT II deficiency causing one documented episode of rhabdomyolysis 16 years ago. She reported no family history of renal disease or CPT II deficiency. She described experiencing progressive back, hip and shoulder pain along with dysuria, subjective fever and malodorous dark-brown urine for the past 1 week. She did not report a history of moderate or even vigorous exercise prior to the onset of symptoms. On admission, she had a temperature of 38.6°C and a

BACKGROUND

Supernumerary kidneys are a unique embryological anomaly defined as organised renal tissue with a distinct collecting system, blood supply and well-defined capsule. To the best of our knowledge, bilateral supernumerary kidneys are even rarer, with only four cases being reported in humans.¹ We found no case reports of any patients with bilateral supernumerary kidneys and carnitine palmitoyl transferase II deficiency (CPT II). CPT II leads to

Figure 1 The image on the left is a non-contrast abdominal CT in the coronal plane displaying native kidneys with inflammatory changes along the inferior pole of the right kidney. The image on the right displays the patient's bilateral supernumerary kidneys with non-obstructive nephrolithiasis and venous drainage visible along the right supernumerary kidney.

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heart rate of 95 bpm. She appeared lethargic on initial assessment. Her physical examination was significant for diffuse tenderness of her upper and lower extremities, bilaterally with severe costovertebral angle tenderness.

INVESTIGATIONS

Laboratory findings were significant for a urinalysis positive for leucocyte esterase, nitrites and 10 red blood cells per high-power field. Creatine phosphokinase peaked at 162 177 IU/L and her serum creatinine peaked at 299.68 $\mu\text{mol/L}$. The urine culture was positive for *Escherichia coli*. CT scan of the abdomen without contrast showed scarred and atrophic bilateral supernumerary kidneys with non-obstructive renal calculi in the native and supernumerary kidneys (figure 1). In addition, fat stranding was noted adjacent to the right native and supernumerary kidneys, indicative of pyelonephritis.

DIFFERENTIAL DIAGNOSIS

Prior to investigations, our differential diagnosis was focused on the patient's enzyme deficiency, dysuria and diffuse myalgias. The most likely aetiology contributing to her symptoms was rhabdomyolysis secondary to sepsis from pyelonephritis. However, in the setting of CPT II deficiency, the patient could have developed rhabdomyolysis from vigorous exercise or viral illness.

TREATMENT

CPT II is an enzyme found along the inner membrane of the mitochondrial matrix and is essential for long-chain fatty acid oxidation. Therefore, the patient was prescribed a diet rich in carbohydrates and low in fats at a ratio of 80:20, so as to limit long-chain fatty acid oxidation. Pyelonephritis and rhabdomyolysis with acute kidney injury was successfully treated with high-rate intravenous fluid resuscitation and a 14-day course of ceftriaxone. Supernumerary nephrectomy was considered; however, due to her isolated symptomology, surgical intervention would have been considered premature. However, with the risk of stress-induced rhabdomyolysis and repeated UTIs, laparoscopic supernumerary nephrectomy may be considered in the future if she continues to present with recurrent UTI and nephrolithiasis.

OUTCOME AND FOLLOW-UP

This patient was admitted for 11 days for severe rhabdomyolysis and acute kidney injury. She was successfully treated with intravenous fluids and antibiotics. She was instructed to make a long-term dietary modification in which her consumption of long-chain fatty acids was limited to less than 15–20% of her diet. Follow-up at 1 month showed complete resolution of rhabdomyolysis. Four months following discharge, she again developed similar symptoms; however, at this time, she was found to have obstructive uropathy of the right common ureter with mild hydronephrosis of the native kidney. Right-sided ureteral stents were placed temporarily and removed following clearance of calculi. She was instructed to follow-up with her primary care provider on a bi-annual basis to assess kidney function.

DISCUSSION

To the best of our knowledge, bilateral supernumerary kidneys are an extraordinarily rare embryological anomaly with only four cases being reported.¹ The true prevalence of this condition is difficult to predict as the occurrence of such a phenomenon in the community is extremely infrequent. Recently, treatment of symptomatic unilateral supernumerary nephrectomy has been expanded to include laparoscopic supernumerary

Learning points

- ▶ Carnitine palmitoyl transferase (CPT II) is an enzyme found along the inner membrane of the mitochondrial matrix and is essential for long-chain fatty acid oxidation. Adult onset CPT II deficiency is a predominantly autosomal recessive disorder that affects skeletal muscle, thus commonly presenting with intermittent myalgias, myoglobinuria and rhabdomyolysis secondary to stress.^{2 4} The exact mechanism by which rhabdomyolysis occurs in the setting of CPT II deficiency is unclear; however, the prevailing theory proposes that high concentration of β oxidation intermediates causes damage to the sarcoplasmic reticulum and inner mitochondrial membranes, leading to increased intracellular calcium, allowing for sustained muscle contraction.²
- ▶ Bilateral supernumerary kidneys, an extraordinarily rare anatomic finding, arise due to ureteric buds interacting with additional metanephric blastemal masses during nephrogenesis.⁶ It is postulated that abnormal ureter formation predisposed this individual to an increased risk of nephrolithiasis and pyelonephritis that subsequently led to severe rhabdomyolysis. The infection is likely to have increased long-chain fatty acid oxidation leading to downstream effects on the calcium channels within the inner membrane of the mitochondria, ultimately resulting in severe rhabdomyolysis.² Renal complications of rhabdomyolysis include acute kidney injury and possibly acute tubular necrosis due to oxidative damage and cast deposition from myoglobinuria.⁷
- ▶ Conservative versus invasive treatment of supernumerary kidneys is primarily driven by symptomatology. In asymptomatic patients, the presence of supernumerary kidneys does not seem to pose an increased risk for major sequela and therefore does not require further intervention. However, in patients presenting with chronic flank pain, recurrent UTIs and/or nephrolithiasis, treatment options vary. Usual care, including antibiotics and hydration, should be administered first to all patients with a UTI or pyelonephritis. In patients with large calculi or signs of hydronephrosis, the need for lithotripsy and ureteral stent placement should be evaluated further. In those that fail standard medical and surgical therapy, supernumerary nephrectomy may be considered as a definitive treatment option. Recently, Innocenzi and colleagues reported a case of successful laparoscopic unilateral supernephrectomy with minimal complication in a patient with chronic UTIs secondary to a unilateral supernumerary kidney.³ Complications of such an intervention include haematoma and urinoma formation, the incidence of which cannot be accurately assessed due to the limited number of similar cases.³ Prior to such intervention, the supernumerary renal collecting system should be imaged with a voiding cystourethrogram, and the vascular anatomy should be delineated with MRI or CT of the abdomen.³
- ▶ Currently accepted dietary modifications in patients with CPT II deficiency include limited fat intake with increased carbohydrate intake. Novel treatment strategies including anaplerotic dietary modifications, in which medium-chain fatty acids, such as triheptanoin, are used as they will not require CPT II for β oxidation. Augmenting 30–35% of total caloric intake with triheptanoin has been shown to minimise the incidence of spontaneous rhabdomyolysis and exercise-induced rhabdomyolysis in patients with adult onset CPT II deficiency.⁸

nephrectomy.³ CPT II deficiency was initially described in 1973 by DiMauro *et al* in adults with exercise-induced rhabdomyolysis.⁴ Subsequent studies have outlined variations in phenotype based on genotypic differences.⁵ In addition, recent studies have given insight into diagnosis and management of CPT II deficiency based on physical examination findings and muscle biopsy results.⁵

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Competing interests None.

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