

Flank pain radiating to the suprapubic region

A 33 year old woman presented at the urology department with a two year history of intermittent flank pain radiating to the suprapubic region. She had no medical or family history of note. Examination showed that she had microscopic haematuria and proteinuria. Radiological investigations were carried out (figs 1 and 2).

Questions

- (1) What abnormality is shown in fig 1?
- (2) What are the predisposing conditions to this abnormality?
- (3) What investigation is being performed in fig 2?
- (4) What is the diagnosis?

Answers

- (1) Bilateral renal calcification as a result of stones or nephrocalcinosis.
- (2) Metabolic and anatomical abnormalities can predispose patients to bilateral formation of kidney stones.

Anatomical problems—Renal calculi are more common where there are structural anomalies in the kidney, such as tubular ectasia, ureteric obstruction, renal papillary necrosis, and medullary sponge kidney.

Metabolic problems—Hypercalciuria can be idiopathic or occurs secondary to hypercalcaemia (in hyperparathyroidism, for example) sarcoidosis, and hypervitaminosis D. High urinary oxalate usually arises from dietary excess or secondary to a malabsorption syndrome. It is rarely associated with a severe congenital defect or primary hyperoxaluria. Hypocitraturia and distal renal tubular acidosis are also implicated in the formation of renal calculi.

- (3) Intravenous urogram.
- (4) Medullary sponge kidney.



Fig 1 Plain abdominal x ray image, including the kidneys, ureters, and bladder



Fig 2

Discussion

Medullary sponge kidney is a renal malformation characterised by dilation of the collecting ducts. This is associated with the formation of cysts that may be microscopic or visible to the naked eye. These are diffuse and bilateral but do not involve the cortex, although in some patients involvement may be limited to one kidney or only some calyces.^{w1}

Stones may develop in the renal parenchyma or more diffuse calcification (nephrocalcinosis) may be present at the corticomedullary junction or both.^{w2} Medullary sponge kidney affects one person in 5000^{w3} and is also known as tubular ectasia or Cacchi-Ricci disease.^{w4}

Although medullary sponge kidney is autosomal dominant, most cases are sporadic, and in less than 5% of cases is there family history.^{w5} Unlike in autosomal dominant polycystic kidney disease, cysts are not seen elsewhere in the body, and renal failure is uncommon.

Diagnosis

The diagnosis of medullary sponge kidney is normally made by intravenous urogram. Changes seen depend on the severity of the pathological changes. This can be as mild as a “blush” (a faint white border on the inside of the kidney outline), to more obvious linear radiation, to unmistakable cystic dilation in the collecting ducts communicating with the calyces (as in fig 1).^{w6} The kidneys are either normal in size (9-12 cm in length) or mildly enlarged. Magnetic resonance imaging is a useful alternative diagnostic tool in patients with allergy to contrast media.^{w7}

Many patients with medullary sponge kidney are asymptomatic and are discovered incidentally when an abdominal x ray is performed for another indication. The key clinical manifestations of the disorder are related to urine stasis and associated tubular defects, most commonly stone formation and urinary tract infections.^{w8}

Medullary sponge kidney accounts for 12-20% of calcium stones. Haematuria (gross and microscopic) is the second most common symptom. The bleeding is typically painless unless clots lead to ureteric obstruction and subsequent renal colic. Urinary tract infection also occurs more often.

Most patients have a near normal life expectancy. Complications, such as infection, haematuria, and stones affect 10% of patients, and progression to end stage renal disease is rare. Tubular defects are associated with metabolic abnormalities, which are associated with other malformations.^{w9}

Management

The disease has no specific treatment, and management aims to prevent complications. High fluid intake may prevent stones forming and infection, and prophylactic antibiotics are used to treat recurrent infections. If possible, any metabolic abnormalities that predispose a patient to stone formation should be tackled.

Although small calculi may be passed spontaneously, larger calculi need active intervention, such as lithotripsy or surgical removal of stones. Renal abscesses are a rare complication that may require intensive antimicrobial therapy and surgical drainage.

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References w1-w9 are on student.bmj.com.

Learning points

A diagnosis of medullary sponge kidney diagnosis can be made on intravenous urogram or computed tomogram

When uncomplicated the condition has no symptoms and no effect on renal function

The main complications arise from urine stasis and tubular defects. Stone formation leads to colic and infection. Renal tubular acidosis causes a metabolic acidosis. End stage renal disease is rare

Management is focused towards complications