

Medullary sponge kidney presenting with hypokalaemic paralysis

K. S. A. JAYASINGHE*
M.B.B.S.

R. MOHIDEEN
M.B.B.S.

M. H. R. SHERIFF
M.B.B.S., M.D.(Ceylon),
M.R.C.P.(UK)

B. L. J. MENDIS
M.B.B.S.

R. EKANAYAKE
M.B.B.S.

K. DHARMADASA
M.B.B.S., M.D.(Ceylon),
F.R.C.P.(Lond), F.R.C.P.(Edin)

University Medical Unit, University of Colombo, Sri Lanka

Summary

Medullary sponge kidney associated with a defect in urine acidification is rare and usually of no clinical significance. We report a case presenting as hypokalaemic paralysis due to associated congenital distal renal tubular acidosis.

KEY WORDS: quadreparesis, renal, tubular acidosis.

Introduction

Medullary sponge kidney is a congenital disease in which the renal medulla is replaced by numerous small cysts. The patients are usually asymptomatic. Those with symptoms usually present with renal or ureteric colic, haematuria and polyuria (Black and Jones, 1979). A few cases with impaired urinary acidification have been reported (Deck, 1965; Grandberg, Layergren and Theve, 1971; Morris *et al.*, 1965), but the associated mild impairment of urinary concentration or acidification is of no clinical concern in the majority of patients (Yendt, 1982).

Here we report a case of medullary sponge kidney with impaired urinary acidification and potassium wasting who presented with hypokalaemic paralysis.

Case report

A 26-year-old male was admitted to the neurosurgical unit of the General Hospital Colombo in June 1979 with a history of weakness of the lower limbs which spread rapidly within 24 hr to involve the upper limbs. He had flaccid areflexic quadreparesis

without sensory involvement. The higher cerebral functions, cranial nerves and ophthalmoscopy were normal and there was no bladder involvement. The cardiovascular system, respiratory system and the abdominal examinations were normal.

Despite normal cerebrospinal fluid, he was treated with dexamethasone on a clinical diagnosis of ascending myelitis. The recovery was complete within 48 hr. In July 1979, he had a similar episode which also apparently responded to dexamethasone.

In September 1979 he was admitted with rapid onset of quadreparesis with associated dyspnoea. Routine biochemical screening revealed a serum potassium of 2.2, 2.3 and 2.8 mmol/l at different intervals with electrocardiographic features suggestive of hypokalaemia. He recovered rapidly with parenteral and oral potassium. Biochemical investigations revealed sodium 140 mmol/l, potassium (as above), chloride 112 mmol/l, standard bicarbonate 17.9 mmol/l, urea 5 mmol/l, creatinine 120 μ mol/l, creatinine clearance (Feb. 1981) 110 ml/min, 24 hr urinary calcium 7.0 mmol.

On a single occasion a centrifuged sample of urine showed 20-30 pus cells and 2-3 red blood cells per high power field, but repeated microscopic examinations were normal and the urine was sterile on culture. No albuminuria or glycosuria was detected. Potassium balance studies were carried out on a 20 mmol sodium and a 20 mmol potassium diet. The serum potassium varied between 3.1 and 3.4 mmol/l on the 4 consecutive days while the excretion of potassium in the urine over 24 hr on these four occasions were 52, 44, 31 and 49 mmol. This showed an inability to conserve potassium despite its restriction in the diet. During an ammonium chloride

*Present address: Department of Medicine, Bristol Royal Infirmary, Bristol.

loading test, (0.1 g/kg body weight), the urine pH never fell below 6.5 in spite of an arterial blood pH of 7.25. Urine osmolality after water deprivation to effect a loss in body weight of 3%, was 307 mOsm/kg (normal—greater than 800 mOsm/kg) suggesting a defect in urinary concentration. Tests using vasopressin were not done. A plain X-ray of the abdomen showed evidence of renal medullary calcification, and intravenous urography showed large kidneys, with a number of pyramidal cavities. The contrast filled these cavities before filling the calyces. Residual filling of the cavities occurred even after the calyces were empty, and there was an increase in the number of radiodense opacities in the pyramids in comparison to the plain X-ray. The calyceal pattern and the rest of the urinary tract were normal. These radiological features are characteristic of medullary sponge kidney (Black and Jones, 1979). The renal arteriogram did not reveal a tumour in the suprarenals and was normal.

Thus our patient had radiological evidence of medullary sponge kidneys with an impaired urinary acidification and excessive urinary potassium loss. He was commenced on oral sodium bicarbonate, 1 g three times a day, and advised to take a generous intake of high potassium-containing foods. No supplementary potassium was prescribed.

After commencing on this regimen he has been completely asymptomatic. His repeat urogram did not show any change in the calcific opacities.

Discussion

Medullary sponge kidney is essentially a radiological diagnosis (Beeson, Mcdermott and Wyngaarden, 1979) and can be associated with congenital defects such as renal tubular acidosis (RTA) or acquired tubular dysfunction from long standing urinary tract infection or obstructive uropathy due to nephrolithi-

asis. Deck (1965) reported three patients with medullary sponge kidneys who had renal tubular acidosis.

Our patient had diagnostic radiological features and an impaired ability to acidify urine with an ammonium chloride load, with acidosis, suggestive distal renal tubular acidosis. Excessive wasting of potassium in the urine probably caused the presentation of hypokalaemic paralysis. Though this could be an acquired defect, the absence of a history suggestive of a chronic urinary tract infection, the presence of sterile urine on repeated examinations, and the lack of deterioration over the observed 2 years are against this possibility. The defect in urinary concentration could be a result of the hypokalaemic nephropathy.

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