

# MEDULLARY SPONGE KIDNEY IN THE RADIOLOGICAL DIFFERENTIAL DIAGNOSIS OF NEPHROCALCINOSIS

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The term "medullary sponge kidney" was coined by Lenarduzzi (5) in 1939 for this condition which was first fully investigated and described by him. Though this condition is uncommon a few authors have published fairly large numbers of cases of their own, collected over a number of years—Lhez (6), Lindvall (7), Evans (2), and Abehouse (1).

Recently a case of medullary sponge kidney was seen locally and the author takes this opportunity to enhance local interest and awareness of this condition and thus possible early diagnosis.

The term "nephrocalcinosis" as used in radiology signifies diffuse calcification in the renal parenchyma. This excludes localised or circumscribed area of calcification as in hypernephroma, tuberculous disease or renal infarction.

Kreel (4) in his excellent article on nephrocalcinosis gives causes of radiologically demonstrable nephrocalcinosis as follows:—(Table I).

Of the above seven known causes of nephrocalcinosis, medullary sponge kidney alone shows no systemic changes.

The condition is diagnosable on radiology alone and the findings are:—

## (1) Plain Radiograph: (Fig. 1a)

There may be one to several hundreds of small calculi either unilateral or bilateral or confined to one pole of a kidney alone. In only two of thirty-five cases of Lindvall and five of twelve cases of Evans were calculi not present. The calculi are dense and sometimes arranged in groups. They are confined to the medulla of the kidney. The appearance is sometimes so characteristic that the diagnosis is strongly suggested on plain film alone. In cases where the calculi are bilateral and diffusely arranged, it has to be differentiated from other causes of nephrocalcinosis. In this respect enlargement of the kidneys is in favour of the diagnosis of medullary sponge kidneys.

## (2) I.V.P. (Fig. 1b)

There would be present cavities of various sizes and shapes in the medulla and if calculi are present, these cavities contain the calculi. These cavities fill before the calyces and on release of ureteric compression, retain the dye longer than the calyces. In early cases, before there is definite cavity formation, there is enlargement of the renal papillae.

TABLE I

Conditions	Systemic Changes
1. Hyperparathyroidism	Osteoporosis; cysts; sub-periosteal resorption; "rugby jersey" vertebrae.
2. Hyperchloraemic acidosis (or tubular acidification defect)	Osteomalacia or rachitic changes.
3. Idiopathic hypercalciuria	Osteomalacia.
4. Sarcoidosis	Pulmonary and osseous changes.
5. Chronic glomerular nephritis	Osteomalacia.
6. Medullary sponge kidney	Nil.
7. Myeloma Cushing's & steroid } rare therapy	Osteoporosis; rib fractures with hypertrophic callus formation.
8. Unknown	Nil.

The differential diagnoses besides those of nephrocalcinosis are renal tuberculosis, calyceal diverticulum and cysts, papillary necrosis and so-called tubular stasis. Amongst these, renal tuberculosis is most important as it is often wrongly misdiagnosed in cases of medullary sponge kidney and unnecessary prolonged chemotherapy instituted.

The underlying pathology is cystic dilatation of the collecting tubules. It is confined to the medulla. The degree of changes varies from very small to large cavity and from a few to innumerable numbers. Calculus formation is due to stasis as there is no biochemical changes in uncomplicated cases and no renal dysfunction. The cause of the cystic dilatation is undecided.

The condition itself does not give rise to any clinical manifestation. The patients present themselves because of symptoms and signs of superimposed infection or stone formation.

#### A CASE OF MEDULLARY SPONGE KIDNEY

K.M.F., a Chinese female was first seen on January, 1958 at the age of 19, for minor illnesses. On 12.9.63, she had R.I.F. pain radiating down to the inguinal region. There was nausea but no vomiting.

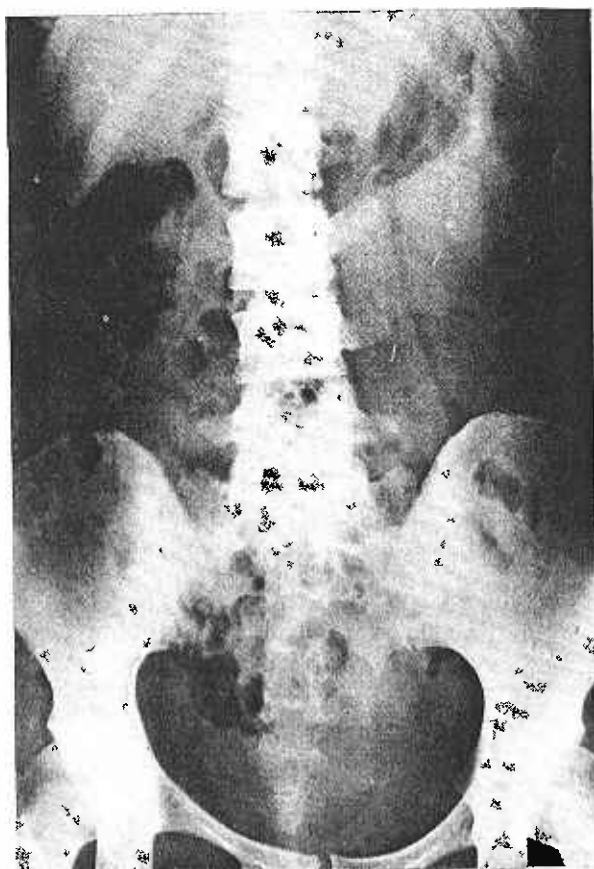


Fig. 1a. Medullary sponge kidney showing unilateral nephrocalcinosis. Normal bony structure.

The laboratory findings were:—

Urine REME: 10-12 RBC, 15-20 WBC  
Albumin +.

Chest X-ray revealed no abnormality.

X-ray abdomen showed nephrocalcinosis on the left (Fig. 1a).

Serum calcium: 10.4 mgm %

Serum inorganic phosphate: 3.4 mgm %

She had persistent pain on follow up, and on 4.10.63 she was admitted for frank hematuria. Physical examination revealed nothing of significance.

The laboratory findings were:

Hb 75% T.W. 4,800 with normal differential count.

Urine FEME: 200-300 RBC  
2-3 WBC

Blood urea: 22 mgm %

Blood uric acid: 4.7 mgm %

Serum calcium: 9.8 mgm %

Serum inorganic phosphate: 3.8 mgm %

BSR: 3 mm (7.10.63)  
2 mm (8.10.63).

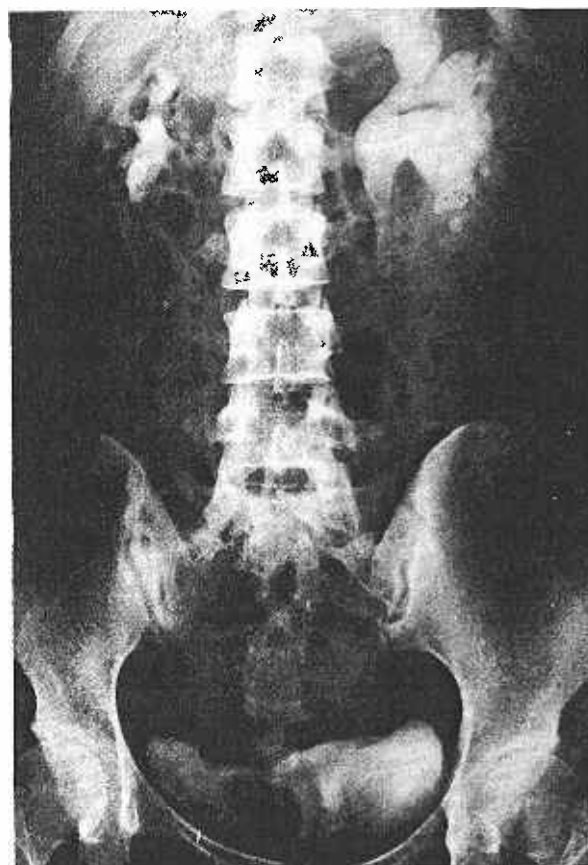


Fig. 1b. I.V.P. film showing an enlarged left kidney with large renal papillae and cysts in the medulla.

24 hours specimens of urine for culture for tuberculous bacillus collected on 5.10.63, 7.10.63 and 8.10.63 were all negative.

I.V.P. on 21.10.63 (Fig. 1b) showed a normal right kidney. The left kidney was enlarged compared to the right. Discrete calcifications were noted in the renal medulla, and surrounded by opaque contrast medium excreted. The renal papillae were greatly enlarged and so were the calyces and pelvis. However, the fornices of the calyces remained sharp.

Cystoscopy on 18.11.63 revealed nothing of significance. I.V.P. was repeated on 3.4.64 and the picture remained the same as before.

The patient continued to have occasional RBC in the urine up to December 1964. A further I.V.P. on 15.8.66 showed that the condition had remained static.

## DISCUSSION

The above case is a good example of medullary sponge kidney, both clinically and radiologically. Its presentation in young adulthood with symptoms and signs of calculus formation, the persistent hematuria and failure to establish tuberculous infection are typical. The normal

serology rules out systemic causes of nephrocalcinosis.

There is no specific treatment for this condition. In severe unilateral cases with gross hematuria, nephrectomy either total or partial, has been attempted.

Kerr et al (3) have pointed out that cases of congenital hepatic fibrosis have renal lesion resembling medullary sponge kidney, but nevertheless have failed to show that medullary sponge kidney is associated with hepatic fibrosis. However, they do advise a close watch for symptoms and signs of hepatic fibrosis in cases of medullary sponge kidney.

Figs. 2a and 2b are the radiographs of a case of acquired renal tubular acidosis with nephrocalcinosis. They are shown for comparison with the radiographs of this case of medullary sponge kidney. The features of note are the bilaterality of the calcinosis, the demineralisation of the bony structures, the presence of ureteric calculus and the absence of enlargement of renal outlines, the papillae and the calyces.

## ACKNOWLEDGEMENT

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Fig. 2a. Plain film of a case of renal tubular acidosis showing bilateral nephrocalcinosis. There are also right ureteric calculi. The bony structure is demineralised.

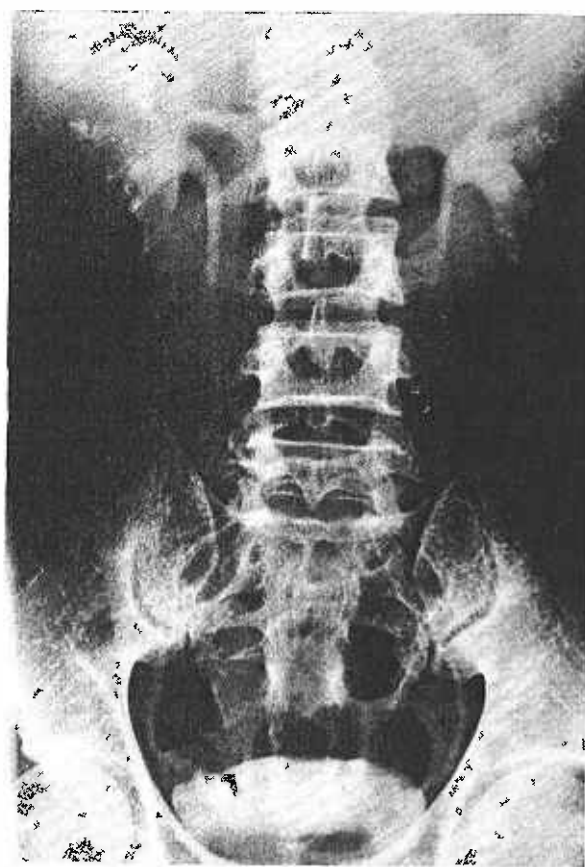


Fig. 2b. I.V.P. film of same case of renal tubular acidosis showing no enlargement of the kidneys nor its papillae.

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