CASE REPORT

A Child with severe Nephrolithiasis and Nephrocalcinosis

by

*BAHRUN, D. *; ALATAS, H. *; WILA WIRYA, I.G.N. * and HIMAWAN, S. ***

(From the Departments of Child Health and Pathology, Medical School, University of Indonesia, Jakarta)

Abstract

A case of severe nephrolithiasis with nephrocalcinosis in a six years old child has been reported.

The diagnosis of nephrolithiasis was made based on the radiological examination and laboratory findings.

The cause of hypercalciuria in this case was still obscure. Necropsy finding revealed a terminal renal failure with severe nephrocalcinosis.

The cause of terminal renal failure in this case was probably multiple renal stones due to prolonged hypercalciuria and severe nephrocalcinosis, complicated by chronic renal infection.

* From the Department of Child Health, Medical School, University of Indonesia, Jakarta.

** From the Department of Pathology, Medical School, University of Indonesia, Jakartta.

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Introduction

Nephrolithiasis with or without nephrocalcinosis is a rare disease in childhood (Daeschner et al., 1960; Troup et al., 1972; Malek et al., 1975).

Its onset may be associated with infection, hypercalciuria, metabolic disorder and urological anomalies (Gazali et al., 1973).

Nephrolithiasis with or without nephrocalcinosis is a frequent situation accounting for numerous emergency room visits and hospital admissions. The urodynamic alterations caused by this disorder may induce problems ranging from urinary infection to renal failure, especially in the recurrent case (Giogliami etal., 1980).

The following case report represents severe nephrolithiasis with nephrocalcinosis which induces progressive renal failure.

As far as we know this case is the first reported in the Indonesian literature.

Case report

R,a six years old boy was admitted to the Department of Child Health, Ciptomangunkusumo Hospital, Jakarta on February, 16, 1981. He was sent by a pediatrician for evaluation of nephrolithiasis.

According to his parents, the patient had an initial complain of dysuria and frequent micturation since two months. Several days before admission he passed two small stones.

The history of diatery intake was normal. The history of excressive administration of vitamin D was denied. He was born spontaneously following normal gestation assisted by an obstetrician. The history of physical and mental development were normal. There was no family history of calculi disease.

Intravenous pyelography examination revealed bilateral multiple renal calices. The calices of both kidneys were already blunt probably due to the stones (Fig 1).

Analysis of the stone showed that it was composed of calcium oxalate.

Physical examination on admission revealed a six years old boy with a body weight of 23 kg and body length of 112 cm. Blood pressure 110/70 mmHg. There was no periorbital edema nor lymphadenopathy. The lungs and heart were normal. The abdomen was supple on palpation and no mass or bruit was detected. Peripheral edema was absent.

Laboratory studies revealed a serum creatinine level of 0,76 mg%; blood urea 27 mg%; the blood albumin 4,09 g%; globulin 2,39 g%; cholesterol 231 mg%, alkaline phosphathase 11,8 Bodansky U. Serum calcium 9,4 mg%; serum phosphorus 4,2 mg%; serum uric acid 5.6 mg %, serum sodium 138 meq/1; serum potassium 3,9 meq/1; serum magnesium 2,1 meq/1; serum chloride 101,6 meq/1.

The hemoglobin content was 12,4 g%; erythrocyte count 4,2 million/cu. mm; leucocyte 1400/cu mm with a normal differential count. Thromboyte count

was normal. Erythrocyte sedimentation rate was 45/h. Blood gas analysis showed a blood PH of 7,379; PC02 34,5 mmHg; HCO3 19,8 meq/l; Base Excess-4,7 meq/1; 02 saturation 97,3% The urea clearance examination was 40 ml/minute/1,73 sqm. Creatinine clearence was 26,7 ml/minute/1,73 sqm. Urinalysis showed a specific gravity of 1,015; PH of the urine was 5,4; proteinuria of 1 + ;RBC of 1-2/hpf; WBC of 10 - 15/hpf. The urinary culture showed Enterobacter aerogenes more than 200.000 colonies/ml The urinary calcium was 290 mg/24 hours or 12,7 mg/kgbw/day. The urinary phophate was 675 mg/24 hours or 29,3 mg/kgbw/day. The procentual tubular reabsorption of phosphate was 79,4%. Aminoaciduria was absent. Radiologic bone survey showed no abnormality.

The working diagnosis was made as nephrolithiasis, pyelonephritis and hypercalciuria with moderately impaired renal function.

He was treated with chlortalidon to prevent recurrent renal stones formation and reduce urinary calcium excretion. He also received a low calcium diet Nalidixic acid was also given to combat the urinary infection. During hospitalization the patient never contracted fever, hypertension or oliguria. He was discharged ten days after admission but did not show up regularly for follow up treatment. Four months later the patient suddenly came with vomiting and diarrhea. Although the general condition was rather bad, the parents refused to hospitalize the patient. One week later they came back again with a severely ill child, pale, apathetic, lethargic and anuria.

He was immediately hospitalized in the intensive care unit. The body temperature was 36°C. Blood pressure 140/95 mmHg. Heart rate 90/minute. Respiration rate 40/minute. Laboratory findings revealed hemoglobin concentration of 4,6 g%; leucocyte count 4600/cumm. Bleeding time, clotting time and thrombocyte count were within normal limits. Blood gas analysis revealed a still compensated metabolic acidosis with a blood PH of 7,3; PCO2 of 23,7 mmHg; PO2 76 mmHg; HCO3 13,7 meq/1 Base excess of - 10,1 meq/1. Bladder catheterization yielded only 10 cc urine. Blood urea was 316 mg%. Blood creatinine was 20,5 mg%.

Based on these findings, a diagnosis of acute on chronic renal failure was made.

The patient received packed red cell blood transfusion and peritoneal dialysis was performed simultaneously. During the next three days the patient's condition improve markedly.

The blood pressure dropped to 120/80 mmHg. Hemoglobin concentration increased to 10 g%, but the thrombocyte count decreased to 22000/cumm. Blood urea was 115 mg%; blood creatinine 11,5 mg% serum sodium 135 meq/1; serum chloride 99,8 meq/1; serum calcium 9,2 mg%;serum phosphorus 4,4 mg%; serum magnesium 2,1 mg%. Urinary production was still low namely

about 50 ml/24 hours. The abdominal X-ray examination revealed that the renal stones became larger and the two kidneys appeared relatively smaller (Fig. 2).

Due to the thrombocytopenia he was transfused with thrombocyte suspension. The thrombocyte was than increased to 262.000/cumm. Thirteen days afterwards the patient's condition further deteriorated. The blood urea increased to 206 mg% and blood creatinine to 13,6 mg%. A second peritoneal dialysis was performed.

However, his condition became further deteriorated and the patient died 4 days afterwards.

Necropsy of the kidney (Fig. 3 : a, b, c) revealed a specimen consisting nine to ten glomeruli. Most of the glomeruli had became total obsolescence, only one to two showed slight sclerosis. The tubuli were atrophic and crystals of calcium salt were found widespread in many dilated tubuli. Examination under polarized light showed birefringence of the crystals. The interstitial tissue was fibrotic and infiltrated by mononuclear cells.

The diagnosis was terminal renal failure with severe nephrocalcinosis.

Discussion

Although calculus disease of the urinary tract is one of the most common urinary problems encountered by the radiologist in adult patient, the condition is uncommon in children (Daeschner et al., 1960). The low incidence of urinary calculi in the pediatric patient is proved by the scarcity of reported cases in the medical literature (Piel et al., 1975).

Daeschner et al., (1960) found in Southeast Texas from 1954 through 1959 in the course of 4442 urologic radiologic procedures performed on approximately 3800 children, 14 patients (0,37%) have urinary calculi. Troup et al., (1972) reported an incidence of renal calculli in childhood in the Milwaukee Children Hospital from January 1950 through December 1969 of 1/7600 admissions. Malek et al., (1975) found 78 children who had upper urinary tract calculi encountered among 145.000 new pediatrics admission within a 20 years period.

Calcium is the most frequent component of stones in all age group occuring in 90% (Prien et al., 1968). Two thirds of all renalstones are composed of either calcium oxalate or calcium oxalate mixed with calcium phosphate in the form of hydroxyapatite (Williams, 1974). Similarly, the analysis of the stone in our case showed that the stone composed of calcium oxalate.

The mechanism that favors deposition of calcium in the kidney is complex and remains unresolved (Chan, 1976). Three major theories have been proposed to explain stone formation and growth (Williams, 1974). First, the precipitation crystallization theory implies that supersaturation of urinary crystaloids eventually leads to their precipitation as a crystal, with subsequent crystal growth. Second, the matrix nucleation

theory based on the assumption that matrix substance forms an initial nucleus for subsequent stone growth by precipitation. Third, inhibitor absence theory : A number of potent inhibitors of crystal formation have been identified in normal urine, including pyrophosphate, mucopolysaccharides, diphosphonates, small polypeptides, urea, citrate, magnesium, certain amino acids and trace metals.

Most investigators agree that the majority of upper urinary calculi in children appear to have unexplained etiology (Troup et al., 1972), however, a number of the cases are considered to be due to infection, urinary tract anomaly, hypercalciuria and metabolic disorder (Gazali et al., 1973).

A definite metabolic cause for calculus was recognized in only 5 children from 120 cases namely cystinuria, oxaluria, distal renal tubular acidosis (RTA) and hyperadrenocortisism (Gazali et al., 1973).

The largest number of children with stones have obstruction of urine flow with stasis and infection primarily from urological anomalies. In some cases from recumbency and immobilization (Piel et al., 1975). Ghazali et al., (1973) found from 34 children with associated urological anomalies in 8 cases the stone was apparently formed in sterile urine and in 26 cases the urine was infected. In 79% of 120stone-forming children, infection with a Proteus species was present, and stone recurred early in 9 whom the only identifiable factor was persistence of Proteus infection (Ghazali et al., 1973).

Ghazali also found using a value of 6 mg/kgbw/day as upper limit of normal calcium excretion in the urine, 12 of 67 children showed hypercalciuria without hypercalcemia. Infection and hypercalciuria with normocalcemia were also found in our case. We assume that the possible origin of stone formation in this case was a combination of these factors.

Looking for the cause of hypercalciuria in our case, we made a differential diagnosis of

1. Renal tubular acidosis of distal tubular origin (RTA type 1).
   The typical chemical findings inclule a normal nonprotein nitrogen, a decreased serum bicarbonate and increased serum chloride, hypophosphatemia, hypocalcemia and alkaline or nearly neutral urine despite systemic acidosis (Melick et al., 1958). In our case repeated urine PH is always acid, excluding the possibility or renal tubular acidosis.
2. Primary hyperparathyroidism
   The diagnosis is based primarily on the repeated laboratory demonstration of hypercalcemia, hypophosphatemia, decreased tubular reabsorption of phosphate and increased serum parathyroid hormone.
   The urinary calcium is usually increased. However, in several cases one or more of these tests were normal, and several repeated determinations were necessary to obtain the diagnosis (Melick, et al. 1958). In our case we found that the concentration of urinary calcium was high, but the serum calcium, phosphorus and procentual tubular reabsorption of phosphate were normal. The X-ray bone survey examination is also normal, so that the diagnosis of primary hyperparathyrodism is excluded.
3. Excessive milk, alkali or vitamin D.
   Excessive milk or vitamin D may result excessive calcium absorption, hypercalciuria and stone formation. In our case we found that the history of dietary intake was normal and excessive administration of vitamin D was denied.
4. Bone disease or immobilization.
   In our case there was no history of immobilization and the result of X-ray bone survey examination was normal.
5. Idiophatic hypercalciuria.
   The diagnosis of idiopathic hypercalciuria in this case was made by urinary calcium excretion above 200 mg/kg bw/day (Gazali et al. 1973) in which no other cause such as RTA, primary hyperthyroidism, excessive milk-alkali or vitamin D and bone disease was found.

The treatment of urinary stone with idiophatic hypercalciuria is still a medical problem. Early recognition of the cause, correction and inactivation of the calculus disease are the essential steps (Malek et al. 1975). Because of the obscure etiology in our case the present treatment has been mostly directed towards reduction of urinary calcium excretion and treatment of the urinary infection. The treatment of idiophatic hypercalciuria which is widely accepted consists of the administration of thiazide diuretics (Coe et al. 1973) and using the cellulose phosphate bind calcium in the food to prevent its absorption (Pak et al. 1974).

Chlortalidon a heterocyclic variant of thiazide and a long acting diuretics has been used by some authors to combat recurrent urinary calcium stones and reduce the urinary calcium excretion in adult patients with success (Sidabutar et al. 1978) Similarly, we also used chlortalidon in our case. However, the drug's effect could not be evaluated because the patient did not show up regularly for follow up examination.

Piel et al (1975) stated that the calcium oxalate stones and nephrocalcinosis could lead to renal failure and early death. In this case, we assume that the prolonged hypercalciuria and chronic infection may be the result of a diffuse nephrocalcinosis and nephrolithiasis with impaired function, leading to terminal renal failure.

Aknowledgement

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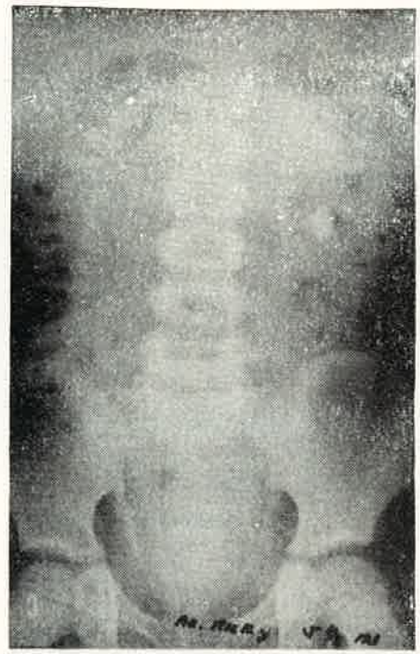

FIG. 1 : *IVP examination showed bilateral multiple renal stones located in the renal calices. The calices of both kidneys were already blunt probably due to the stones.*

FIG. 2 : *The abdominal X-ray examination revealed that the renal stones became larger and the two kidneys appeared relatively smaller*

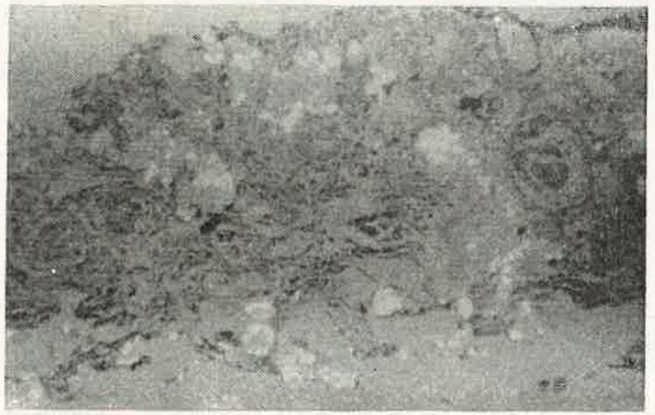

FIG. 3c : *Examination under polarized light showed birefringence of the crystals*

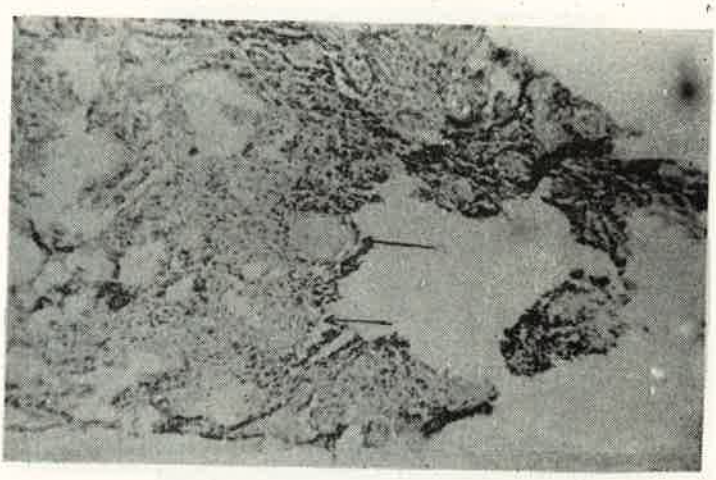

FIG. 3a : *The specimen showed few sclerotic glomeruli (arrows), dilated tubuli containing crystals, interstitial fibrosis and chronic inflamatory cell infiltration.*

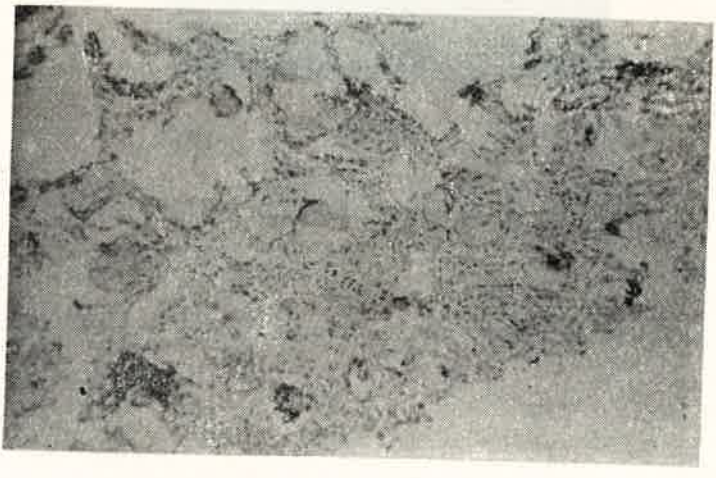

FIG. 3b : *Many calcium crystals were found in dilated tubuli*