Diagnosis: Wilson’s disease (hepatolenticular degeneration). Wilson’s Disease is an inborn error of copper metabolism which may present as a predominantly hepatocellular disease (chronic hepatitis or cirrhosis) in 40% of patients, as a psychiatric illness in 10%, or as a neurologic disorder in 35%. More unusual presentations include painless hematuria, glycosuria or hemolytic anemia and, in the pediatric age group, as a syndrome of acute hepatitis with liver failure, hemolysis and renal insufficiency. While patients with hepatic involvement alone may not have a Kayser-Fleischer ring, even with slit-lamp evaluation, virtually every patient with a neuropsychiatric disorder exhibits this sign.

Diagnosis can be confirmed thus: serum total copper level > 118 mg/dl and serum ceruloplasmin < 20 mg/dl.

Treatment for the patient’s condition: Z)-penicillamine, a copper-binding agent, 1–2 gm/day, plus potassium sulfide, 20 mg t.i.d., and a diet low in copper.

The prognosis without treatment is death within 2 years.

Kidney pathology: proximal tubular dysfunction (incomplete Fanconi’s syndrome) from copper deposition and toxicity in the proximal tubules. A wide variety of renal abnormalities has been described in Wilson’s disease, including decreased renal plasma flow, decreased glomerular filtration rate, renal tubular abnormalities and renal stones. Proximal renal tubular dysfunctions include aminoaciduria, hyperphosphaturia, hypercalciuria and glucosuria. Distal renal tubular dysfunctions include potassium-wasting and concentration defects. Combinations of proximal and distal tubular dysfunctions include defective excretion of uric acid and acid-base disturbances. The latter is almost always due to distal renal tubular acidosis. Infrequently, there may be a proximal renal tubular acidosis pattern with defective bicarbonate reclamation.

(6) Bone pathology: Renal rickets or osteomalacia. The pivotal defect in this syndrome is a proximal renal tubular phosphate leak resulting in hypophosphatemia. This in turn stimulates renal 25-OHD3 1α-hydroxylase and the production of 1,25-(OH)2 cholecalciferol, which causes increased calcium and phosphate absorption, parathyroid suppression and hypercalciuria. Prolonged hypophosphatemia would diminish osteoid mineralization, causing osteomalacia or renal rickets.

References
The etiology of this patient’s pancreatitis cannot be known with certainty. Tetracycline is associated with acute pancreatitis in patients with liver disease. There has only been 1 report of a patient developing pancreatitis following the administration of tetracycline in the absence of liver disease. Since tetracycline is partially excreted by the kidney, it should not be administered to patients with renal insufficiency without reducing the dose. Since there are so many alternatives to this antibiotic, there is no reason to give it to patients with renal disease. It is possible that this patient’s pancreatitis was the result of tetracycline which accumulated to toxic levels due to the preexisting renal insufficiency. On the other hand, may be that the pancreatitis was unrelated to the tetracycline and was the result of a viral infection which also caused the respiratory symptoms. In any case, tetracycline should not be administered to patients with renal insufficiency.

The acute renal failure was likely caused by the severe volume contraction which resulted from the pancreatitis. While tetracycline is well known to produce azotemia secondary to its antianabolic effect, especially in patients with renal insufficiency, it does not produce the type of acute renal failure seen in this patient.

The permanent loss of renal function following this patient’s acute illness is of considerable interest. The expected course of acute renal failure in a patient who survives the acute illness is recovery of renal function. This patient made no such recovery. The reason for the failure to recover any renal function is not clear. If the patient’s severe volume contraction was sufficient to cause cortical necrosis, then no recovery would be expected. On the other hand, it is becoming increasingly clear that patients who suffer a bout of acute renal failure superimposed on chronic renal insufficiency may not recover renal function in the fashion expected from those patients who develop acute renal failure on the background of normal kidneys. For this reason, it is particularly important to avoid exposing patients with chronic renal insufficiency to those events which may cause acute renal failure.

References

Announcements
First International Conference on New Actions of Parathyroid Hormone
A conference dealing with new actions of parathyroid hormone, such as its effects on the heart, hematopoietic system, blood vessels, blood pressure, skeletal muscle, and lipid, protein, and carbohydrate metabolism. New actions of the hormone on the kidney and the skeleton will also be included. There will be invited presentations, poster sessions, and oral presentation of selected abstracts submitted by interested investigators.
The conference will take place in Kobe, Japan, during October 27–31, 1987. It is being organized by an international committee under the chairmanship of Prof. Takuo Fujita (Japan). The members of the committee include Shaul G. Massry (USA) as Secretary General, Etsuro Ogata (Japan), Michael Rosenblatt (USA), David A. McCarron (USA), and R. Dieter Hesch (FRG).
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Uremic Toxins Symposium
Ghent, Belgium, October 3–4, 1986