Nephrology
Dialysis
Transplantation

Nephroquiz for the Beginner
(Section Editor: T. J. Rabelink)

Watch out for lumbosciatalgy

A 51-year-old man with a history of progressive renal insufficiency that developed over a period of 4 months was referred to our unit in May 1998. Recent medical history revealed malaise, backache, and hypertension. Hydrocele was found on physical examination. Laboratory studies on admission showed: serum creatinine 6.6 mg/dl (583 μmol/l), blood urea nitrogen 60 mg/dl (21.4 mmol/l), uric acid 7.2 mg/dl (428.2 μmol/l), white blood cell count 6.6 × 10³/mm³ with a normal differential, haemoglobin 10.3 g/dl, platelet count 234 000. Urinalysis showed 80–100 erythrocytes and 2–4 leucocytes per high power field without proteinuria or casts. Renal ultrasound revealed bilateral hydronephrosis. Neither renal ultrasound nor abdominal radiography revealed the presence of stones along the urinary tract. A CT scan of the abdomen was performed.

Question
What is the most likely diagnosis? What treatment should be given to the above patient?
Answer to the quiz on preceding page

The cause of renal insufficiency in this patient turned out to be retroperitoneal fibrosis, which had led to bilateral obstruction and hydronephrosis. The ureters were embedded in dense fibrous tissue. The obstruction of retroperitoneal lymphatic or venous vessels, reducing the absorption of the fluid normally contained in the vaginal tunic, caused the hydrocele.

Retroperitoneal fibrosis is an uncommon disease (although presumably more common than hitherto appreciated) that often presents in an insidious fashion. Early symptoms, as in our case, may include malaise, anorexia, weight loss, fever, hypertension and backache originating in the lumbosacral region and described as a dull ache with girdle distribution. Symptoms due to the obstructive uropathy or to uraemia usually are manifest in advanced stages.

It is assumed that retroperitoneal fibrosis is an autoimmune condition. Our patient had idiopathic retroperitoneal fibrosis since evidence for drug-induced retroperitoneal fibrosis (methysergide, beta-blockers, bromocriptine, methyldopa) or systemic collagen diseases was not found [1,2]. Imaging studies are required to establish the diagnosis of retroperitoneal fibrosis [3,4]. Excretory urography will show proximal hydrourerteronephrosis, medial deviation of the ureters (which is characteristic but not diagnostic since it may be seen in a variety of conditions) and extrinsic compression of the ureters. CT scan is considered the examination of choice to visualize the extent of fibrosis. In our patient, CT scanning showed a solid formation enveloping the abdominal aorta, the inferior vena cava and the ureters (Figure 1). Extension to the common iliac blood vessels or into the mesenteric root and mediastinum, involving neighbouring organs may be seen. The differential diagnosis includes syphilitic aortitis, lymphoma and various forms of cancer, and may require laparotomy or CT-guided needle biopsy.

Treatment of retroperitoneal fibrosis usually consists in surgical relief of obstruction by ureterolysis. After surgery, long-term steroid treatment has been recommended in order to maintain remission [2]. In our patient, such combined management was associated with a marked improvement of renal function (serum creatinine 1.6 mg/dl, 141.4 μmol/l) and regression of retroperitoneal fibrosis (Figure 2). Other therapeutic options include the use of methylprednisolone pulse therapy alone, azathioprine, or the association of mycophenolate mofetil and prednisone [5].

The described case points out that lower backache is not always of rheumatologic origin.

Suggested reading


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