Nephrology
Dialysis
Transplantation

Nephroquiz for the Beginner
(Section Editor: M. G. Zeier)

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Diabetes mellitus, arterial hypertension and hilar adenopathy

Case

A 48-year-old female presented with progressive weakness, oedema of the ankle, and a worsening hypertension. The previous medical history included insulin-dependent diabetes for 15 years, arterial hypertension, and nodular goitre. Clinical examination of the patient did not reveal other abnormalities. Routine laboratory results showed elevated serum creatinine of 3.2 mg/dl, mild anaemia with a haemoglobin of 11.3 g/dl. Serum electrolytes were in normal ranges. Proteinuria was 440 mg/l.

The routine chest X-ray showed bihilar adenopathy, so a CT scan of the lung was performed. This showed bihilar and mediastinal adenopathy and round–stellar opacities (Figs 1 and 2).

Questions

What is the reason for the proteinuria? What is the reason for the hilar adenopathy? What is your diagnosis?

Fig. 1. Chest X-ray of the patient. (Arrows showing bihilar adenopathy.)

Fig. 2. CT scan of the thorax. (Arrows showing enlarged mediastinal lymph nodes.)
Answer to the quiz on preceding page

The diagnosis is sarcoidosis, the reason for proteinuria was a renal involvement. The chest X-ray and the CT scan of the lung showed typical manifestations of thoracic sarcoidosis.

Because of the elevated creatinine levels despite a well-controlled diabetes mellitus and no signs of a diabetic retinopathy, a kidney biopsy was performed. This showed no diabetic changes but epithelioid cell granulomas were found (Fig. 3). In the lymph nodes obtained by mediastinoscopy epithelioid granulomas were also found. This confirmed the diagnosis of sarcoidosis.

The patient was treated with oral steroids at a dose of 1.5 mg/kg body weight. Eight months later kidney function had improved slightly but the general condition remained unchanged. Abdominal ultrasound revealed a large tumour in the pelvis. The histology showed a highly malignant B-cell lymphoma and the patient died of an infection after the initiation of chemotherapy.

Sarcoidosis is a systemic disease in which every organ can be involved. Its characteristic features are epithelioid cell granulomas. Acute onset of the disease is known as Löfgren’s syndrome; however, chronic disease is more common. Symptoms of renal involvement are deterioration of renal function, proteinuria, and erythrocyturia. The literature reports renal involvement in up to 27% of patients with sarcoidosis. Renal function usually improves after steroid therapy.

In addition to the development of epithelioid cell granuloma in the kidneys, nephrocalcinosis can occur as a cause of hypercalcaemia due to the increased production of 1–25 dihydroxyvitamin D3 by stimulated alveolar macrophages.

The occurrence of malignancies, especially lymphomas, in patients with sarcoidosis is more common than in healthy persons. This might be the effect of the proposed T-cell disorder in sarcoidosis.

Suggested reading


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Fig. 3. Histology of the renal biopsy showing two small epithelioid cell granulomas within the renal medulla, PAS × 360.