Unusual course of nephrotic syndrome associated with atypical pneumonia

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Keywords: nephrotic syndrome; pneumonia

Introduction

Focal segmental glomerulosclerosis (FSGS) is the most common cause of idiopathic nephrotic syndrome. As secondary forms may have a better prognosis than the primary form of FSGS it is important to exclude an underlying disease.

Case report

A 28-year-old man presented in March 2000 with a 1-week history of increasing leg oedema. Surprisingly, he had also experienced an involuntary weight loss of 6 kg during the preceding weeks. There was no relevant medical past history. Physical examination revealed a slightly elevated blood pressure of 145/90 mmHg and marked pretibial oedema. As laboratory values included a proteinuria of 14.5 g/day, a serum albumin of 1.85 g/dl, and a serum cholesterol of 357 mg/dl, a nephrotic syndrome was diagnosed. Serum creatinine was 0.9 mg/dl and creatinine clearance 148 ml/min. The urinary sediment showed no abnormalities.

Kidney biopsy revealed obliteration of the glomerular capillary lumen in segmental parts of some of the glomerula and adhesions of portions of the glomerular tufts to the Bowman’s capsule (Figure 1), establishing the diagnosis of FSGS. The immunofluorescence revealed segmental IgM deposits in one of the visualized glomerula. Electron microscopy showed marked foot process fusions of the podocytes in a light microscopically normal glomerulum.

Serum electrolytes and differential blood count were unremarkable. Screening for hepatitis B and C, and HIV was negative and an immunoserologic workup failed to show any abnormalities despite a moderately elevated C-reactive protein (2.0 mg/dl). The initial chest X-ray showed pneumonia in the right upper lung field and right mediastinal enlargement (Figure 2). Computed tomographs visualized multiple mediastinal and retroperitoneal lymphnodes (Figure 3). Therefore, mediastinoscopic biopsy was performed. Morphology revealed the presence of Hodgkin’s disease of the nodular-sclerosing type (clinical stage III₂B). Chemotherapy was started according to the enhanced BEACOPP-protocol [3], including procarbacine, etoposide, cyclophosphamide, doxorubicine, vincristine, bleomycine, and prednisone. As renal function was normal, no dose reduction of the drugs was necessary. The patient had no major side effects despite hair loss. Four months after the start of therapy a control CT was performed and showed a remarkable regression of the pathological lymphnodes (Figure 4). Furthermore, the proteinuria decreased after 4 months of therapy (0.41 g/day), leg oedema were absent and the patient was feeling well.

Discussion

The primary form of focal-segmental glomerulosclerosis has an acute onset and usually presents with a persistant nephrotic syndrome and hypertension as was the case in our patient. Secondary forms of FSGS can be associated with intravenous drug abuse, lithium therapy, HIV-infection, malignancy or Hodgkin’s disease. Usually, Hodgkin’s disease rarely affects the kidney. Minimal change nephropathy occurs in only 0.4% of these patients and amyloidosis,
as a later complication of Hodgkin’s disease, in 0.1%. The association of FSGS and Hodgkin’s disease seems to be more uncommon [1]. From a pathogenic point of view, a circulating T-cell derived factor seems to play a role in the pathogenesis of FSGS [4]. Hodgkin’s disease is a B-cell derived tumour, but polyclonal T-cells are present in close vicinity to the malignant B-cells [5]. Those T-cells may produce a ‘soluble factor’ targeted to glomerular cells, thereby causing glomerulopathy. This factor might be a chemokine which induces inflammation and subsequent scarring, leading to enhanced protein-permeability of the glomerulum [6].

Nephrotic syndrome can be associated with several complications. Loss of clotting inhibitors and particular immunoglobulins makes the patient prone to complications like thrombosis and infection. Our patient presented with a clinically silent pneumonia (Figure 2), which may have been promoted by the nephrotic syndrome.

A high-proteinuric state as it was present in our patient is associated with a particularly bad prognosis in idiopathic FSGS. The initial therapy of FSGS is prednisone, which is only able to induce a remission in 20% of cases [2]. In secondary FSGS, the discovery of an underlying disease can change the therapeutic approach and therefore improve the prognosis of the nephrotic syndrome as in our case.

Teaching point

In a patient with focal-segmental glomerulosclerosis (FSGS), a secondary form of FSGS should be considered.

It is necessary to establish the diagnosis of an underlying disease like Hodgkin’s lymphoma as early
as possible since therapeutic intervention can induce a remission of the nephrotic syndrome.

References


