**Letters**

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**Alpha-1-antitrypsin deficiency and mesangio-capillary glomerulonephritis in an elderly patient**

Sir,

Alpha-1-antitrypsin deficiency is a frequent genetic disorder with an incidence at around 1/1500 to 1/3000. In adults, it mostly presents first with panacinar emphysema. Liver cirrhosis and hepatocellular carcinoma are less common and nephritic syndrome is an exceptional finding at presentation. In infants and children, it is the most common genetic cause of liver disease and in this age group glomerulonephritis is a frequent association.

We describe a 73-year-old woman who presented with renal failure, related to alpha-1-antitrypsin deficiency. A 73-year-old woman was admitted to the hospital because of fatigue, loss of appetite and weight loss of more than 10 kgs. Her medical history was limited to a thyroidectomy. She stopped smoking more than 20 years ago (10 pack years). Physical examination revealed a cachectic and pale woman. The abdomen was swollen (due to ascites) and both legs showed bilateral pitting oedema.

Blood count: platelets 191,000/mm$^3$, haemoglobin 8.4 g/dl; white blood cells 9,000/mm$^3$ (92% neutrophilic granulocytes). Serum creatinine was 2.41 mg/dl, serum urea 276 mg/dl and urinary protein 5.5 g/g creatinine. Sodium was 130 mEq/l, potassium 5.1 mEq/l and albumin 1.58 g/dl (normal 3.6–5.1 g/dl). Abdominal echography showed bilateral pitting oedema. The abdomen was swollen (due to ascites) and both legs showed bilateral pitting oedema.

Renal biopsy showed the typical findings of type I membranoproliferative glomerulonephritis, on light and electron microscopy and immunofluorescence examination [1]. Liver biopsy showed micronodular cirrhosis.

Haemodialysis was started for the nephritic syndrome with renal failure. After an initial period of improvement, she refused food. As a result, her condition deteriorated fastly with renal failure. After an initial period of improvement, she refused food. As a result, her condition deteriorated fastly.

Kidney disease as the presenting sign of alpha-1-antitrypsin deficiency is exceptional.

The relationship between glomerulonephritis and alpha-1-antitrypsin deficiency is not well understood. Possible explanations are the disturbance of enzymes in the kidney or the presence of liver failure. The loss of balance between protease and anti-protease activity may cause tissue damage to organs other than the lungs [2]. The abnormal PiZ protein may induce formation of circulating immune complexes, causing vasculitis and glomerulonephritis [3] or may act as an antigen responsible for local immunological response, resulting in glomerulonephritis. The finding of IgM antibodies against Epstein–Barr virus indicates a recent infection with possibly liver damage, which could have been the trigger for this late manifestation of glomerulonephritis [4].

In conclusion, although renal failure and nephrotic syndrome are uncommon in adults with alpha-1-antitrypsin deficiency, our case report underlines the importance of considering alpha-1-antitrypsin deficiency in the work-up of nephritis in combination with chronic liver disease.

Conflict of interest statement. None declared.


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**Protein-losing gastroenteropathy in a patient with systemic lupus erythematosus and antiphospholipid antibody syndrome simulating nephrotic syndrome**

Sir,

Systemic lupus erythematosus (SLE) is an autoimmune disorder which can affect multiple organs [1]. Protein-losing enteropathy (PLE) in patients with SLE is uncommon [2], typically occurring in young women, and characterized by the onset of profound oedema and hypoaalbuminaemia [3], clinically indistinguishable from nephrotic syndrome. In many cases, it is the first obvious manifestation of SLE [3]. Diarrhoea is present in about 50% of cases, but steatorrhoea is absent [3]. We report a case of PLE associated with SLE and antiphospholipid antibody syndrome (APS).

A 34-year-old white woman was referred to our service to investigate a nephrotic syndrome. Three months previously, she presented with abdominal cramps, nausea and oedema of the lower limbs that evolved to generalized oedema. She also had malar rash, photosensitivity, hypothyroidism and a sister with SLE. She denied miscarriage or oral contraceptives use.

Clinical examination revealed normal vital signs, generalized oedema and ascites. Laboratory data included: normal complete blood count/LDH/sodium/potassium/creatinine/albumin was 0.8 g/dl, globulin 2.4 g/dl, cholesterol 284 mg/dl, C3 79 mg/dl (normal 84–167), C4 15 mg/dl (16–30), ANA >1/200, anti-DNA 49 UI (<30), anti-Ro/La/Ena/Sm were negative, C-reactive protein 49 mg/ml (<5).

Surprisingly, the urinalysis was normal with negative proteinuria. Investigation of the oedema showed no renal or liver disorder, nor malnutrition. Nuclear imaging study employing chromium-labelled albumin revealed an enteric loss of 1878 ml of protein per day (normal <14.6 ml/day). Endoscopy was unrevealing. Colonoscopy with biopsies...