Autopsy Findings of Primary Sjögren's Syndrome with End-Stage Renal Failure

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Dear Sir,

We describe a case of primary Sjögren's syndrome (pSS) with end-stage renal failure. A 51-year-old woman experienced dry mouth, bone pain and general fatigue. She was admitted to another hospital, where pSS was diagnosed on the basis of Shirmer's test and salivary gland biopsy. Tests showed deterioration of renal function, with a serum creatinine level of 2.3 mg/dl. She refused to accept specific forms of medication such as steroid therapy, and no renal biopsy was performed. She was admitted to our hospital 3 years later, suffering from the same complaints. She exhibited marked metabolic acidosis and hypo-calcemia. Laboratory parameters included hemoglobin 7.6 mg/dl, white blood cell count 8,600/mm³ (eosinophilia 11%), blood urea nitrogen 27 mg/dl, creatinine 3.6 mg/dl, calcium 6.9 mg/dl, inorganic phosphate 2.9 mg/dl, IgG 3,090 mg/dl, β_2 -microglobulin (β_2 MG) 29.1 mg/l (0.6-2.1), base excess -12.4 mmol/l. Complement components were normal. Rheumatoid factor was positive, while anti-nuclear antibody, lupus erythematosus (LE) cell, LE test, and anti-DNA antibodies were negative. Urine protein excretion was 0.5 g/day with renal tubular proteins. Creatinine clearance was 9.6 ml/min. A bone marrow biopsy revealed normocellular marrow with slight enhancement of eosinophils. The patient received oral drugs to resolve the renal tubular acidosis and hypocalcemia. Administration of prednisolone 5 mg/day improved eosinophilia, erythrocyte sedimentation rate, and symptoms of bone pain. Renal function remained stable for 5 years, but then acute exacerbation of renal insufficiency occurred with a fever of unknown etiology, severe ileus, and pathological bone fractures. Maintenance hemodialysis (mHD) was continued for 2 years. However, polyuria was persistent because of renal tubule dysfunction during mHD. Finally, the patient died of bacterial peritonitis caused by perforation of the small intestine, which was rich in lymphatic glands. Autopsy revealed multiple ulcers on the tongue and caries of the teeth, keratoconjunctivitis, atrophic salivary glands, chronic esophagitis, chronic hepatitis with hepatomegaly (1,740 g), old pulmonary tuberculosis, and atrophic kidneys (30 g/30 g). A microscopy study revealed that almost all extraglandular organs were replaced by fatty tissue. Renal tubules were almost totally destroyed and marked infiltration of inflammatory cells, predominantly lymphocytes, plasma

cells and histiocytes, was observed in the renal interstitium. However, the extent of global glomerular sclerosis was limited to only 30% (fig. 1). The structure of the remaining glomeruli was relatively well preserved compared with the typical findings usually seen in patients on mHD.

Cases of severe renal insufficiency resulting from chronic interstitial nephritis (ON) associated with pSS are quite rare. Only 2 cases treated by mHD have been reported [1], and both showed marked increases in serum β_2 MG and γ -globulin. The presence of infection and organic involvement of the liver or lung indicates a poor prognosis in pSS [2]. Patients with severe renal involvement usually die of complicating insufficiency of organs other than the kidneys before HD is acquired. Therefore, cases treated by mHD are quite rare. In the present patient, there was no evidence that the unusual population of T cells in the renal interstitium (helper/sup-

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 $\frac{3}{8}, \frac{1}{8}, \frac{5}{8}, \frac{5}{8} = \frac{1}{8}$.
 HP $\hat{a}\hat{1}$

Fig. 1. Renal specimen obtained at autopsy reveals that the extent of global glomerular sclerosis is limited to only 30%.

pressor cell ratio 3:1) was responsible for the unusual features of the disease. We had not administered high-dose prednisolone therapy because of the presence of severe osteoporosis and renal involvement. Our patient did not have a past medical history of pyelonephritis, exposure to radiation or continuous nonsteroidal anti-inflammatory drug therapy, as possible causes of CIN.

References

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