SEVERE RENAL INVOLVEMENT IN PRIMARY SJÖGREN'S SYNDROME

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Primary Sjögren's syndrome is a chronic inflammatory autoimmune disorder that is characterized by a mononuclear cell infiltration of the exocrine glands. Similar lymphocytic infiltrates may invade visceral organs, and this results in several extra glandular manifestations, including renal disease.

Various rare renal lesions have been reported, such as membranous and membranoproliferative nephritis (7), necrotizing glomerulonephritis (13), renal lymphoma and pseudolymphoma (1).

The characteristic histological finding is a focal interstitial nephritis which occurs in about 50 percent of the patients. Its frequency may be underestimated. The interstitial nephritis results in latent or overt tubular defects: (table I) distal renal tubular acidosis type one (20 to 25% of the patients) (13), with possible nephrocalcinosis (2), nephrogenic diabetes insipidus (reported in up to 50 percent of the patients) (13), rarely proximal renal tubular acidosis (13) and Fanconi syndrome. Aseptic leucocyturia (11), proteinuria (with levels below 0.5 g/day) and mild renal impairment (8) are quite common.

Three cases of rapidly evolutive and severe tubulo-interstitial nephritis (creatinine clearance below 30 ml/mn) are described in the literature. Tu and al in 1968 (10) and Gerhard and al in 1978 (5) Cobo Reinoso (4) reported the observations of three females who had a PSS and whose creatinine clearance were respectively 16, 20 and 23 ml/mn. No therapeutical trial was initiated.
We are going to report here 5 cases of PSS with marked deterioration in renal function.

5 females, mean age 55.2 years (range : 25 to 68 years) admitted for tiredness satisfied the criteria for the diagnosis of PSS (Homma's criteria), (table II).

- xerophtalmia
- abnormal Schirmer test : less than 5 mm of filter paper was moistened after 5 mn).
- keratoconjunctivitis sicca with Rose Bengal staining (11)
- xerostomia
- and a characteristic biopsy of the minor salivary glands : grade IV in the classification of Chisholm and Mason with more than one focus of lymphocytes for 4mm² gland section (3).

Furthermore 4 patients had an elevated erythrocyte sedimentation rate. Hypergammaglobulinemia was present in three cases. Antibodies to SSA and to SSB were always negative. None of the patients met clinical or serological criteria for an additional auto-immune disease (rheumatoid arthritis, systemic lupus erythematosus, systemic sclerosis, polymyositis, polyarteritis nodosa). They didn't take any nephrotoxic drug. Three patients had other disease features Raynaud’s phenomenon, arthritis, peripheral neuropathy, pulmonary fibrosis.

Laboratory and renal morphological data are summarized in table N°III.

Arterial blood pressure was normal. The mean creatinine clearance was 11.1 ml/mn (range 4 to 25 ml/mn), the mean urinary protein excretion was 980 mg/d (range : 500 to 1660 mg). Microscopic hematuria and aseptic leucocyturia were present in each case.

Two patients had distal renal tubular acidoses ; there was no impairment of proximal tubular function. The kidneys were normal in 3 cases, nephrocalcinosis was present in patient N°1, and small sized kidneys were seen in patient N°5.

All renal biopsy specimens revealed a dense mononuclear cell interstitial infiltrate, and fibrosis. The tubular pattern was destroyed by tubular dilatation and atrophy. Segmental glomerular sclerosis was seen.