experienced teams such as that of Dr Andrews to adopt the policy of not treating this group of patients. This attitude, however, has been criticized by other groups [1] since presentation of anti-GBM nephritis with oliguria or severe renal insufficiency may not always be accompanied by advanced histological lesions. In some cases, acute tubular necrosis lesions have been observed which are probably associated with the haematuria of glomerular origin and these could be responsible for the oliguria-renal insufficiency rather than the glomerular lesions themselves. On the other hand, there are reports of patients with marked oliguria or anuria who have recovered sufficient renal function to avoid long-term dialysis or transplantation [1–4]. We have the same experience in an unpublished case. We feel that the determining factor may be the nature of the crescent, i.e., cellular crescents as shown by our patients are more likely to be reversible.

Although all of our patients admitted to the hospital with acute renal failure it is very difficult to make the decision not to do anything or offer any type of therapy. This may lead to irreversible renal failure with no hope for the patient. We prefer to administer low dose immunosuppression and wait. Not only did our patients lack complications due to therapy, but 2 months later one underwent an episode of pulmonary haemorrhage, probably because of the insufficient intensity of the treatment.

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Letters

Sjögren’s syndrome complicated by MPO-ANCA positive crescentic glomerulonephritis

Sir,

Although Sjögren’s syndrome (SS) is occasionally complicated by tuberculosis interstitial nephritis as one of the extraglandular manifestations, the association of crescentic glomerulonephritis is not common [1,2]. We report here a female patient with SS associated with rapidly progressive glomerulonephritis and myeloperoxidase-specific anti-neutrophil cytoplasmic antibodies (MPO-ANCA). Renal biopsy revealed crescentic glomerulonephritis with scant immune-deposit, in addition to tubulointerstitial nephritis. She developed progressive renal failure, but this was reversed by steroid therapy and plasma exchange.

Case. A 67-year-old woman was admitted to our department on October 7, 1997 for nasal bleeding and progressive renal dysfunction. One year prior to admission, she noted the numbness and livedo of lower extremities. On March, 1997, she was found to have proteinuria and renal dysfunction (BUN 24.9 mg/dl, creatinine 1.7 mg/dl). One month prior to admission, she had noted recurrent nasal bleeding. The patient was admitted for further examination. On admission, her temperature was 36.6 °C, pulse, 80 beats/min; blood pressure; 160/90 mmHg. Examination of chest revealed fine crackles present at the lower back of chest. There was livedo on the limbs. Findings included, marked anaemia (Hb 5.2 g/dl), elevated serum BUN (33 mg/dl) and Cr (2.8 mg/dl), positive nuclear antibody (X320, speckled pattern), antibody for SS-A 32.5 U/ml (normal range: 7–20 U/ml), positive rheumatoid factor (RF) and cryoglobulins.

Immunoelectrophoresis of serum protein demonstrated no monoclonal band and cryoglobulins were classified as type III. High concentrations of serum MPO-ANCA was noted (603 ÉLISA U/ml) serum proteinase 3 specific anti-neutrophil cytoplasmic autoantibodies (PR-3-ANCA) were not detected. Urinalysis showed proteinuria (0.43 g/day) and microhaematuria.

Chest computed tomography (CT) scan showed irregular opacities, interstitial fibrosis, and mild pericardial effusion. Schirmer’s test demonstrated 5 mm (right eye) and 5 mm (left) tearflow at 5 min. Minor salivary gland biopsy showed chronic inflammation with lymphocyte infiltration and acinar atrophy consistent with SS. A renal biopsy specimen contained 24 glomeruli, 11 of which were completely sclerosed, and four of which had fibrocellular crescents. There was mild mesangial sclerosis, occasional capillary collapse and interstitial infiltration of lymphocytes (Figure 1). Direct immuno-

Fig. 1. Renal biopsy (light microscopy): in addition to the diffuse interstitial mononuclear cells infiltrations, 20% of glomeruli obtained showed fibrocellular crescents. No findings of arteritis was identified. (PAS stain, ×70).

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fluorescent staining of renal biopsy specimens revealed no deposition of IgG, IgM or C3 in the glomeruli (data not shown). Examination of the nasal cavity revealed an oozing ulcer of the septum. Nasal bleeding was stopped after nasal tamponade.

An initial diagnosis of primary SS with crescentic glomerulonephritis was made and she was treated with intravenous methylprednisolone (125 mg/day, three successive days) and followed by plasma exchange (3 l/day, total 6 l). Thereafter she was given oral prednisolone (40 mg/day). These therapies resulted in a marked decrease of MPO-ANCA. Low dose of cyclophosphamide (25 mg/day) was also tried during, but had to be stopped because of myelosuppression. Renal function had markedly improved (BUN 32 mg/dl, Cr 1.8 mg/dl) when she was discharged after 99 days.

Comment. The most common renal lesion of SS is interstitial nephritis with interstitial lymphocytic infiltration, fibrosis and tubular atrophy. In contrast, glomerulonephritis is relatively rare [1,2]. The present case had extraglandular manifestations affecting the lungs and kidneys. The present case fulfilled the criteria of primary SS [3], i.e. dry eyes with positive Schirmer’s test, abnormal biopsy findings of salivary gland and presence of specific autoantibodies. MPO-ANCA, the serological marker for systemic vasculitis, was also present. Cryoglobulin-mediated glomerulopathy is occasionally found in primary SS [4], although cryoglobulins were detected in our patient, but immunofluorescence study showed no immunoglobulin deposits in the glomeruli. Antibodies to MPO have been reported in collagen disease such as SLE [5,6], but only two cases of SS with MPO-ANCA-related glomerulonephritis have been reported [7,8].

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Seizures as the presenting feature of post-streptococcal glomerulonephritis

Sir,

During the last decades many diseases have become progressively rare mainly due to better hygiene and vaccination of the population. For this reason diagnosis can be difficult, especially when presenting symptoms are confusing or unusual.

Case. A 33-year-old female was admitted to the emergency ward because of seizures and blurred vision. The previous week she had been treated with tablet tetracycline for sinusitis, but was otherwise healthy. On admission she was somnolent, with mild periorbital oedema, and diastolic hypertension, 110–120 mmHg. The rest of the physical examination was unremarkable. A CT (computed tomography) of the brain and a lumbar puncture were performed, followed by an MR (magnetic resonance). Localized oedema was noticed in the occipital, temporal, frontal and parietal lobes (Figure 1). Cerebro spinal fluid (CSF) cell count was normal. Virus encephalitis was suspected and treatment with acyclovir parenterally was initiated but discontinued 2 days later when virologic examination of blood and CSF did not support this diagnosis. A chest X-ray showed bilateral perihilar pulmonary congestion and oedema, particularly in the right lower lobe, while sonography of the kidneys, including renal artery circulation, were normal. The sedimentation rate was 50 mm (0–20), C-reactive protein 33 mg/l (<9 mg/l), serum creatinine 71 µmol/l (60–120 µmol/l), serum albumin 27 g/l (36–48 g/l). Urinalysis showed albuminuria reaching 1.3 g/l and microscopic haematuria. An acute inflammatory reaction and hypoalbuminaemia were found in plasma electrophoresis the following day, with complement C3 and C4 within the lower normal range. Serologic tests for ANA, p- and c-ANCA, anti-GBM antibodies, and RF gave no evidence of connective tissue or vasculitic diseases. A 24 h urine cathocolamines sample was normal. Renal crisis due to scleroderma was excluded on the basis of the clinical and laboratory findings. Eye fundoscopy and urinary bladder cystoscopy disclosed no abnormalities. The initial patholo-

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Fig. 1. Axial MR T2 weighted images show focal hyperintensity in the temporal, occipital, frontal and parietal lobes, consistent with oedema (arrows).