

CLINICAL PRESENTATION

A 71-year-old woman presented with subacute kidney failure and mild proteinuria. Two weeks earlier, her physician had noticed slight edema and a urinary tract infection and started therapy with torsemide and ciprofloxacin. She had a long history of rheumatoid arthritis, hypertension, sensory polyneuropathy, chronic pericarditis, depression, and anorexia nervosa. Her medi-

cations included methylprednisolone, sulfasalazine, mirtazapine, and olanzapine. Six months before admission, her serum creatinine level was 1.5 mg/dL (133 μ mol/L). Laboratory findings on admission showed the following values: creatinine, 2.8 mg/dL (248 μ mol/L); blood urea nitrogen, 74 mg/dL (52.8 mmol/L); hemoglobin, 7.8 g/dL (78 g/L); sodium,

130 mEq/L (mmol/L); potassium, 4.1 mEq/L (mmol/L); chloride, 93 mEq/L (mmol/L); bicarbonate, 23 mEq/L (mmol/L); and phosphate, 5.8 mg/dL (1.88 mmol/L). Creatinine clearance was 16 mL/min/1.73 m², and proteinuria had protein of 410 mg/d without light chains. Urine dipstick showed (+) protein, and sediment was negative for cells and cellular casts.

- What is your clinical differential diagnosis?
- What do you see by light microscopy?

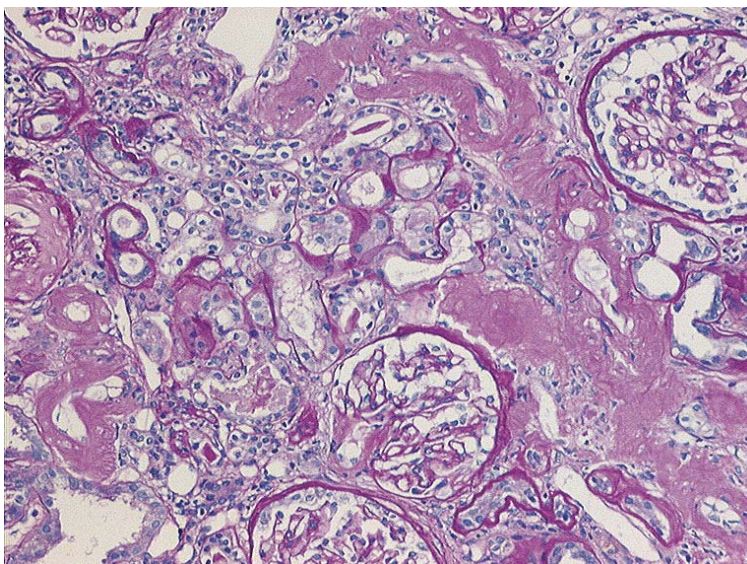


Figure 1. Kidney biopsy (periodic acid Schiff stain; original magnification \times 200).

DISCUSSION

■ What is your clinical differential diagnosis?

The patient was thought likely to have an interstitial nephritis caused by a medication for her rheumatoid arthritis or in association with anorexia nervosa.

■ What do you see by light microscopy?

As seen in Figure 1, there is massive deposition of weak periodic acid–Schiff–positive acellular material in the vascular walls, but no deposits in glomeruli. No significant mesangial proliferation or interstitial accumulation of leukocytes is noticed.

■ What is your interpretation of the Congo red staining?

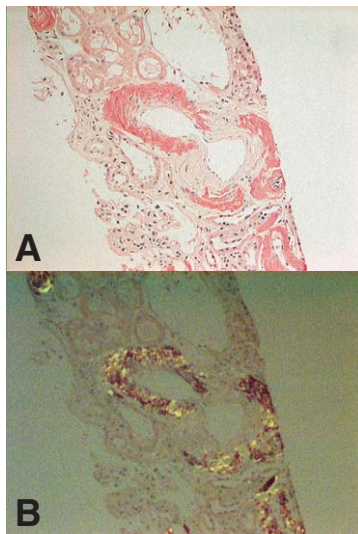


Figure 2. Congo red staining of the kidney biopsy specimen (original magnification $\times 100$).

There is Congo red–positive material in perivascular areas and vascular walls (light microscopy; Fig 2A). Under polarized light,

there is apple-green birefringence, which is pathognomonic of amyloidosis (Fig 2B).

■ What is your clinicopathologic diagnosis?

Our patient showed an interesting pattern of renal amyloid deposition. Deposits were found mainly in vessel walls, whereas glomeruli were hardly affected. This pattern explains the leading clinical presentation with a decrease in renal function, but remarkably low proteinuria. Immunohistochemistry was positive for amyloid A protein and negative for λ and κ light chains (data not shown).

Renal AA amyloidosis is a frequent complication of inadequately treated rheumatoid arthritis, occurring in 20% to 50% of patients. Most patients with renal amyloidosis have mainly glomerular deposits, causing proteinuria in the nephrotic range. However, a subset of patients has mostly vascular and tubulointerstitial amyloid deposits and presents with an early decrease in renal function and only mild proteinuria. Occasionally, renal tubular acidosis and mild diabetes insipidus may occur. Ultrastructural characteristics of the amyloid and genetic predisposition seem to influence the prevailing site of deposition.

Other common renal lesions in patients with rheumatoid arthritis are membranous glomerulopathy, glomerulonephritis, vasculitis, analgesic nephropathy, and thinning of basement membranes. An important clinical differential diagnosis in our patient was interstitial nephritis secondary to the medications or the chronic eating dis-

order. Anorexia nervosa commonly is complicated by interstitial nephritis. Usually this is caused by long-term use of laxatives and chronic hypokalemia, which was not the case in our patient.

FINAL DIAGNOSIS

Renal AA amyloidosis with vascular predominance, secondary to rheumatoid arthritis.

CASE PROVIDED BY

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SUPPORT: None.

POTENTIAL CONFLICTS OF INTEREST: None.

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