End-Stage Renal Disease after Necrotising Glomerulonephritis in an Elderly Patient with Temporal Arteritis

J.	Julio	Pascual ^a
C.	Carlos	Quereda ^a
F.	Fernando	Liaño ^a
M.J.	María J.	García-Villanueva ^b
F.	Francisco	Mampaso ^c
J.	Joaquín	Ortuño ^a

Servicios de ^aNefrología, ^bReumatología y ^cAnatomía Patológica, Hospital Ramón y Cajal, Madrid, Spain

Dr. Julio Pascual, Servicio Nefrología, Hospital Ramón y Cajal, Carr. Colmenar km 9,100, E-28034 Madrid (Spain)

Dear Sir,

Small and Brisson [1] and Canton et al. [2] have recently reported patients with temporal arteritis (TA) and renal involvement. In addition, biopsy-proven glomerular disease and/ or intrarenal vasculitis has been reported in a few patients with TA [3-8]. It is debated whether these are related disorders or only chance associations. We would like to report a patient with TA who subsequently developed necrotizing glomerulonephritis and end-stage renal disease.

A 76-year-old man was first seen in February 1985 after several weeks of diffuse headache and jaw claudication. Both temporal arteries were palpable and tender with diminished pulses and the rest of physical examination was unremarkable. Laboratory studies revealed a hemoglobin of 143 g/l, erythrocyte sedimentation rate (ESR) 74 mm/h and serum creatinine 70 µmol/l. Urinalysis showed 7-8 red blood cells per field and no proteinuria. Antinuclear antibody, rheumatoid factor, cryoglobulins, hepatitis B antigen and immunoglobulins were absent or normal. Temporal artery biopsy showed intense arteritis with multinucleated giant cells; the patient was thus diagnosed as having TA. Immunohistochemical study with monoclonal antibodies showed a predominance of T helper lymphocytes in the inflammatory infiltrate. He began to receive prednisone 60 mg daily which induced complete resolution of his symptoms within a week. ESR decreased to 20 mm/h. He was maintained on 20 mg of prednisone daily, tapered to 8 mg daily and remained asymptomatic for 2 years.

In May 1987, after the diagnosis of duodenal ulcer and left aseptic femoral osteone-crosis, steroids were stopped. He did not come to clinical follow-up during the subsequent 2 years and 7 months. On readmission, he complained of a subjective decrease in diuresis, intense upper and lower extremity weakness and a weight loss of 2 months duration. He denied headache, jaw claudication or visual symptoms. Examination was unremarkable except for absence of left coxo-femoral joint movements. Laboratory studies revealed hemoglobin 100 g/l, ESR 126 mm/h, serum creatinine 1,408 µmol/l. Urinalysis showed proteinuria 2.8 g/l, numerous red blood cells, 20 white blood cells per high-power field and hyaline and granular casts. He underwent

percutaneous renal biopsy. Light-microscopic examination revealed a renal cortex with 20 glomeruli, 13 of them globally sclerotic and the rest showing focal segmental necrotizing glomerulonephritis (FSNGN). All glomeruli showed marked epithelial proliferation with crescents and there were also regions of tubular atrophy with associated interstitial fibrosis and chronic mononuclear inflammation with a predominance of T helper lymphocytes. Vasculitic lesions were not apparent. On the basis of the biopsy findings and previous side effects of steroids, he was maintained only on prednisone 30 mg daily and was considered as having terminal renal failure. Hemodialysis was performed on an alternate-day basis; creatinine clearance was never > 1 ml/min. When steroids were tapered to 10 mg every other day, he developed intense headache so he was put on prednisone 10 mg daily. An internal arteriovenous fistula was performed on the forearm; he has now completed 15 months of hemodialysis treatment.

While it is known from autopsy studies that TA is a disease which can affect almost any artery [9], renal function is usually normal in patients with this disease and there have been very few reports of renal insufficiency occurring in patients with demonstrated TA. Renal disease usually is not mentioned in large series of TA, or is considered not to be associated with renal vasculitis [6]. There are several descriptions of patients with TA and abnormalities in urinalysis with no biopsy verification of the renal damage [11]. Wegener and Hollenhorst [3] described a patient with suspicion of TA (not biopsied), who subsequently died in renal failure, autopsy study disclosing focal glomerulonephritis and polyarteritis nodosa. Lupus nephritis developing in a patient who initially presented with classic TA has been reported in a large series of TA [4]. This patient died in renal failure unresponsive to steroid therapy. The first patient with biopsy-proven TA and renal vascular disease was described by O'Neill et al. [5] who speculated about the possibility of common association. Unfortunately, treatment schedule or evolution were not specified. In their review of glomerulonephritis and vasculitis, Droz et al. [6] included a patient with TA and focal crescentic glomerulonephritis without vasculitic lesions. No clear data on evolution were provided either. Elling and Kristensen

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[7] reported a patient with classical signs of TA who suddenly developed fatal renal failure, despite treatment with steroids. Autopsy showed disseminated giant cell arteritis in the kidneys, pancreas, oesophagus and larynx, with the most pronounced changes in the kidneys, where all arteries and arterioles were affected. Glomeruli were absolutely preserved. Small and Brisson [1] have recently reported a patient with Wegener's granulomatosis affecting the kidneys presenting as TA. Finally, Canton et al. [2] have reported a patient similar to ours, but with a good renal outcome after immunosuppressive treatment with cyclophosphamide and steroids.

Our patient had biopsy-proven TA of 5 years duration, easily controlled with low-dose steroids. After 2 years without steroids, he developed rapidly progressive glomerulonephritis with extensive glomerular necrotizing lesions and sclerosis. Renal biopsy in systemic necrotizing vasculitis commonly ends

in the diagnosis of FSNGN either with or without evidence of vessel necrosis beyond the glomerular level [12]. Wilkowski et al. [13] studied 170 patients with renal vasculitis and glomerulonephritis; 108 had FSNGN alone, 33 FSNGN and small-artery vasculitis and 29 FSNGN and medium-sized artery vasculitis. There was considerable overlap of clinical, laboratory, pathologic and evolutive findings among the three groups, probably representing the

continuum of inflammatory changes involving the vascular tree: FSNGN is really renal vasculitis.

Giant cell arteritis and renal vasculitis in our patient could represent a single diffuse vascular inflammatory process [1] or the chance association of two different vasculitides. The presence of urinary abnormalities in many patients with TA [10] is suggestive of a common association. Banks et al. [14] have shown that the majority of inflammatory cells in TA are of the helper T cell subset. We

studied with antilymphocyte monoclonal antibodies temporal artery and renal biopsies from our patient, showing a helper T lymphocytes predominance in both tissues. The poor response to steroid therapy of renal disease in our patient, with TA easily controlled with low-dose steroids, pleads against a common pathogenetic mechanism between both features and underscores the need of more aggressive immunosuppressive therapy. Only one reported patient has survived with functioning kidneys and was treated with cyclophosphamide [2]. Our patient is the first case with TA and renal vasculitis leading to end-stage renal disease and further need of maintenance hemodialysis. The effect of cyclophosphamide was thought to be excessively noxious in an elderly patient.

Careful evaluation of urinary abnormalities and renal biopsy in patients with TA could provide new insights about this association and allow prompt diagnosis and treatment.

References

- Small P, Brisson ML: Wegener's granuloma-tosis presenting as temporal arteritis. *Arthritis Rheum* 1991;34:220-223.
- Canton CG, Bernis C, Paraiso V et al: Renal failure in temporal arteritis. *Nephron* 1992; 12: 380-383.
- Wagener HP, Hollenhorst RW: The ocular lesions of temporal arteritis. *Am J Ophthalmol* 1958;45:617-630.
- Fauchald P, Rygvold O, Oystese B: Temporal arteritis and polymyalgia rheumatica. Clinical and biopsy findings. *Ann Intern Med* 1972;77: 845-852.
- O'Neill WM, Hammar SP, Bloomer HA: Giant cell arteritis with visceral angitis. *Arch Intern Med* 1976;136:1157-1160.
- Droz D, Noel LH, Leibowitch M, Barbanel C: Glomerulonephritis and necrotizing angitis. *Adv Nephrol* 1979;8:343-368.
- Elling H, Kristensen IB: Fatal renal failure in polymyalgia rheumatica caused by disseminated giant cell arteritis. *Scand J Rheumatol* 1980;9:206-208.
- Truong L, Kopelman RG, Williams GS, Pirani CL: Temporal arteritis and renal disease. Case report and review of the literature. *Am J Med* 1985;78:171-175.
- Klein RG, Hunder GG, Stanson AW, Sheps SG: Large artery involvement in giant cell (temporal) arteritis. *Ann Intern Med* 1975;66:77-86.
- 10 Hauser WA, Ferguson RH, Holley KE, Kurland LT: Temporal arteritis in Rochester, Minnesota, 1951-1967. *Mayo Clin Proc* 1971;46:597-602.
- Crosby RC, Wadsworth RC: Temporal arteritis: Review of the literature and report of five additional cases. *Arch Intern Med* 1948;81:431-464.
- Ballow JE: Renal vasculitis. *Kidney Int* 1985; 27:954-964.

Wilkowski MJ, Velosa JA, Holley KE, et al: Risk factors in idiopathic renal vasculitis and glomerulonephritis. *Kidney Int* 1989;36:1133-1141.

Banks PM, Cohen MD, Ginsburg WW, Hunder GG: Immunohistologic and cytochemical studies of temporal arteritis. *Arthritis Rheum* 1983;26:1201-1207.