Wegener's Granulomatosis Complicated by Pericardial Tamponade and Renal Failure

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Dear Sir,

A 50-year-old woman was admitted to hospital on March 30, 1994, because of coughing and uremia. She had a history of coughing, white mucoid sputum of small volume, weakness, anorexia and pleuritic chest pain. There was nothing in her past and family history. Her physical examination revealed a temperature of 36 °C, pulse 80/min and regular, blood pressure 100/60 mm Hg, respiration 22/min. She was pale. Cardiac examination was normal. The breath sounds were heard less on the basal segments of the right lung. We could not find any organomegaly and LAP.

Laboratory findings were as follows: hemoglobin 8.1 g/dl, WBC 9,600/mm³, platelets 147,000/mm³, ESR 120 mm/h, BUN 145 mg/dl, creatinine 13.6 mg/dl, Na 142 mEq/dl, K 6.1 mEq/dl, Cl 102 mEq/dl, Ca 5.6 mg/dl, P 6.1 mg/dl, serum protein 6.6 g/dl, albumin 2.7 g/dl; blood smear and bone marrow were normal, microscopic analysis of urine revealed 15-20 RBC, fine granular casts and 10-15 leukocytes/high-power field; ANF, RF and anti-DNA were negative, ANCA was not tested. C3 and C4 were normal; daily proteinuria 1 g. HBsAg and anti-HCV were negative; computed tomography of the lung revealed patchy infiltrates and bilateral hilar lymphadenopathy, X-ray examination of paranasal sinuses revealed left maxillar sinusitis. In abdominal ultrasonography, the sizes of the left and right kidneys were 98 × 37 and 97 × 34 mm, respectively. Abdominal computed tomography revealed no further pathology. In renal biopsy,

in the glomerular compartment, there were segmental changes, excessive polymorpho-nuclear leukocyte (PMNL) infiltration, nuclear debris, plasma cells and attachments on Bowman's capsules; in the interstitial compartment, granulomas which revealed necrosis in the inner zones, some fibrosis, atrophy of some tubules, leukocyte and hyaline casts in tubule lumens were seen. In the vessel walls, thickening, necrosis that consisted of PMNL, plasma cells and lymphocytes and in some areas occlusion were seen. Immunohistochemically, IgM was localized in the glomerular tufts and along the capillaries, IgG and IgA were found positive in the glomerulus and vessel walls, locally. Amyloid was negative. Pathologic examination of pericardial biopsy was necrotizing vasculitis that suggested Wegener's granulomatosis (WG). Epidermal hyperkeratosis and atrophy, collagenous degeneration in band form and mononuclear inflammatory cell infiltration around vessels in the upper dermis were observed in a dermal

biopsy. IgM and IgA were localized on the dermal capillaries in a granular form, IgG was found in the dermo-epidermal compartment in a granular form, IgA was detected in the dermoepidermal compartment in a focal, granular form. The lupus band test was positive in the dermal biopsy.

The patient received cyclophosphamide 400 mg/m² every 3 weeks and methylprednisolone, 60 mg/day i.v. In our case, we controlled uremia with dialysis therapy. At the 30th hospital day, we observed pericardial

frôlement and a blood pressure of 60/40 mm Hg. Echocardiography suggested pericardial tamponade. Pericardial drainage was performed. Due to the nature of the disease and a complication of medications, septicemia developed, antibiotic therapy was ineffective and the patient died.

We present a case with WG as a systemic disease consisting of necrotizing granulomatous vasculitis of the upper and lower respiratory tracts, focal necrotizing glomerulonephritis and systemic vasculitis of small arteries and veins [1]. In WG there is involvement of the upper and lower respiratory tracts, lungs, kidneys and eyes. Involvement of the central nervous system, myocardium, pericardium, dermal tissues, muscle and bone is less frequent [2]. In addition to involvement of the lung and kidney, we found necrotizing vasculitis and granuloma formation in biopsies of the skin and pericardium. ANF is positive in 20% of cases, anti-DNA negative, C3 and C4 are normal, RF is positive in 50% of cases [3]. cANCA and pANCA are positive in 88-90% and 3-5% of clinically active cases, respectively [2-4]. In the case of renal involvement, the patient may clinically be asymptomatic or there may be rapidly progressive glomerulonephritis with proteinuria, hematuria and red cell casts in urine analysis [3]. In renal biopsy, necrosis, granuloma formation and vasculitis are observed and no immunocomplex deposition is present [4]. Skin specimens most commonly show small-vessel leukocytoclastic vasculitis and, less commonly, dermal

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necrotizing granulomas or necrotizing vasculitis involving larger dermal arteries or veins [2].

Varying degrees of segmental necrotizing glomerulonephritis are noted in 80%. Vasculitis unrelated to glomerular vessels occurred in 8% of specimens, and vasculitis and granulomatous changes are both present in 3% of specimens [2].

In conclusion, the main points that attract attention are:

(1) in our case, the skin biopsy showed vasculitis in the absence of a skin lesion; (2) granuloma formation in renal biopsy is not frequent in WG; however, we found granuloma in the renal interstitium; (3) the most frequent presentation of WG is respiratory involvement; however, renal failure was the most prominent finding in our patient. In spite of therapy, cardiac tamponade which was due to vasculitis proven by biopsy, developed.

References

Stegeman CA, Tervaert JW, et al: Serum markers of T cell activation in relapses of Wegener's granulomatosis. *Clin Exp Immunol* 1993;91: 415-418.

Hoffman GS, Kerr GS, et al: Wegener granulomatosis: An analysis of 158 patients. *Ann Intern Med* 1992;116:488-492.

Jacobson H, Striker G, et al: Renal disease in Wegener's granulomatosis and vasculitis. *Principles Pract Nephrol* 1991;4:316-320.

Gross WL, Schmitt WH, et al: ANCA and associated disease: Immunodiagnostic and pathogenetic aspects. Clin Exp Immunol 1993;91:1 – 12.

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