

and prednisone. *J Am Med Assoc* 1955; 158: 459.

A. Rodríguez García¹ and R. Guerra Rodríguez²

¹Hospital General de Fuerteventura. ²Hospital Universitario Insular de Gran Canaria.

Correspondence: Alejandra Rodríguez García. jairarodriguez@hotmail.com. Hospital General de Fuerteventura. Ctra. Aeropuerto, km 1. 35600 Puerto de Rosario. Las Palmas de Gran Canaria. España.

Genetics and environment: pathogenetic factors of vasculitis?

Nefrología 2008; 28 (5) 568

To the editor: Etiopathogenesis of vasculitis is not fully understood, and environmental factors have been implicated in genetically predisposed individuals.^{1,2} The histological renal expression of systemic vasculitis is a pauci-immune necrotizing glomerulonephritis (PNG).³ Two cases of familial vasculitis in two brothers living in a rural environment are reported here.

CASE 1

A 63-year-old male, a shepherd living in a rural environment. He consulted in 1988 for an impaired general status, and laboratory tests revealed oliguric acute renal failure. Histological analysis of renal biopsy found a necrotizing vasculitis with extracapillary proliferative glomerulonephritis. The patient was treated with corticosteroids and oral cyclophosphamide. No renal function improvement occurred, and renal replacement therapy was required until the patient died in 2003.

CASE 2

A 72-year-old male, a farmer living in a rural environment. In April 2004 he complained of cough, expectoration, fatigue, and anorexia. His family history revealed that the patient reported as case 1 was his brother. His parents had died at an advanced age, and he

had two sisters with type 2 diabetes mellitus, one of them with a history of pulmonary tuberculosis.

Based on his personal history, clinical signs, and renal function impairment, a renal biopsy was performed, which confirmed the presence of PNG in the setting of a systemic vasculitis associated to P-ANCA (positive anti-MPO, titer 442 U/mL).

DISCUSSION

Pauci-immune necrotizing glomerulonephritis with extracapillary proliferation is the renal pathological expression of systemic vasculitis. This group of diseases is characterized by inflammation of small and medium-sized blood vessels, and includes Wegener's granulomatosis (WG), microscopic polyangiitis (MPA), Churg-Strauss syndrome, or vasculitis limited to the kidney.³

Their etiopathogenesis is unknown. Occurrence of these diseases in several members of a same family has suggested that genetic factors could contribute to its occurrence.¹ We report two cases of PNG as a renal manifestation of systemic vasculitis in two brothers. In case 2, the disease started when the first patient had already died. The presence on the same disease in two brothers could support the genetic component in the etiopathogenesis of vasculitis.

Some researchers have attempted to find associations of vasculitis with HLA genes.¹ Recent studies found a positive association with HLA DR1, particularly in patients with WG, and negative associations with HLA DR3, particularly in Churg-Strauss granulomatosis and polyarteritis nodosa.^{1,4} Our patient had the haplotype A1, B8 B35 Cw4 Cw7 DR3 and DR5 DQ2. Case 1 haplotype is unknown because the patient died before the second patient experienced the disease.

It has also been suggested that environmental factors could contribute to disease development in genetically predisposed individuals.² Patients reported here lived in a rural environment, and some environmental component may possibly have contributed to occurrence of the same disease in both patients.

In conclusion, these two cases of PNG as an expression of systemic vasculitis in two brothers living in a similar environment could support the suggested hypothesis of an influence of environmental factors on the etiopathogenesis of vasculitis in genetically predisposed individuals.

1. Griffith ME, Pushey CD. HLA genes in ANCA-associated vasculitides. *Exp Clin Immunogenet* 1997; 14: 196-205.
2. Huang DR, Zhou Y, Hoffman GS. Systemic vasculitis. Pathogenesis: immunogenetic factors. *Best Pract Res Clin Rheumatol* 2001; 15: 239-258.
3. Jennette JC, Falk RJ. Small-vessel vasculitis. *N Engl J Med* 1997; 337: 1512-1523.
4. Boki KA, Dafni U, Karpouzas GA, Papasteriades C, Drosos AA, Moutsopoulos HM. Necrotizing vasculitis in Greece: clinical, immunological and immunogenetic aspects. A study of 66 patients. *Br J Rheumatol* 1997; 36: 1059-1066.

M. Heras Benito, M. J. Fernández-Reyes Luis, R. Sánchez Hernández and A. Saiz*

*Servicio de Nefrología. Hospital General Segovia. *Servicio de Anatomía Patológica. Hospital Ramón y Cajal. Madrid.*

Correspondence: Manuel Heras Benito. mheras@hge.sacyl.es. Hospital General de Segovia. Ctra. de Ávila, s/n. 40002 Segovia. España.

Intracranial hypertension as presentation of neurobrucellosis in a patient on hemodialysis

Nefrología 2008; 28 (5) 568-569

To the editor: Brucellosis is a zoonosis with a high incidence rate in Spain, particularly in rural areas. While neurological involvement is uncommon, it has clinical significance due to the associated morbidity.

We report the case of a 29-year-old male born in Senegal with an unremarkable epidemiological history. His clinical history included arterial hypertension and chronic kidney disease (CKD) from an unknown cause on chronic hemodialysis for one year. Patient reported low grade fever and fatigue for the past 15 days. During hemodialysis, he expe-