SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Nephrology-Hemodialysis-Transplantation

ANCA and Anti-GBM Double-Positive Vasculitis: Case Report

Samia Elkarci[1*], Younes Lakziz[1], Mariam Chettati[1], Wafaa Fadili[1], Inass Laouad[1]

[1]Nephrology-Hemodialysis-Transplantation Departement, Arrazi hospital, University hospital Mohammed VI, Marrakech, Morocco

DOI: 10.36347/sasjm.2022.v08i04.019 | **Received:** 02.03.2022 | **Accepted:** 06.04.2022 | **Published:** 23.04.2022

***Corresponding author:** Samia Elkarci
Nephrology-Hemodialysis-Transplantation Departement, Arrazi hospital, University hospital Mohammed VI, Marrakech, Morocco

Abstract **Case Report**

Double-positive vasculitis with anti-polynuclear cytoplasm (ANCA) and anti-glomerular basement membrane (GBM) antibodies is a rare entity of systemic vasculitis defined by the presence of ANCA and anti-GBM antibodies. The gradual accumulation of clinical and therapeutic data shows the usefulness of identifying and differentiating this entity from the two vasculitis respectively associated with the isolated presence of each of these two antibodies. Indeed, the double-positive ANCA and anti-GBM vasculitis appears to associate the characteristics of the demography and the extra-renal and pulmonary involvement of the ANCA-associated vasculitis on the one hand, and of the histological type and severe renal prognosis of the anti-MBG vasculitis on the other hand, with the renal involvement which is the only involvement consistently observed in double-positive vasculitis [1]. The aim of this case is to describe the clinico-biological, histological and prognostic characteristics of this entity, in light of recent literature and ongoing therapeutic changes in the two eponymous vasculitis.

Key words: ANCA-associated vasculitis, anti-GBM vasculitis, double-positive vasculitis.

INTRODUCTION

ANCA vasculitis and Good-Pasture's disease are extremely severe pathologies requiring prompt management. Double-positive vasculitis is a rare entity, associating ANCA and anti-GBM antibodies.

We report here the case of a patient with double-positive vasculitis describing the clinical presentation, the therapeutic management and the prognosis.

CASE PRESENTATION

A 23-year-old woman, with no medical history, who consulted for a generalized edematous syndrome with recurrent epistaxis in a context of anuria and deterioration in general condition, and whose clinical examination revealed edema of the lower limbs, low abundance ascites with a proteinuria cross on the urine dipstick without hematuria.

Biological exams show severe renal insufficiency at 140 mg of serum creatinine (GFR at 3.4 ml/ mn), anemia (Hemoglobin at 5.3 g/dl) and a biological inflammatory syndrome; ANCA positive anti-myeloperoxidase type (anti MPO). Chest X-ray with no abnormalities.

On histology, glomerulonephritis with extra-capillary proliferation, on immunofluorescence, linear deposits of IgG and C3 at the glomerular basement membrane, suggestive of anti-GBM vasculitis.

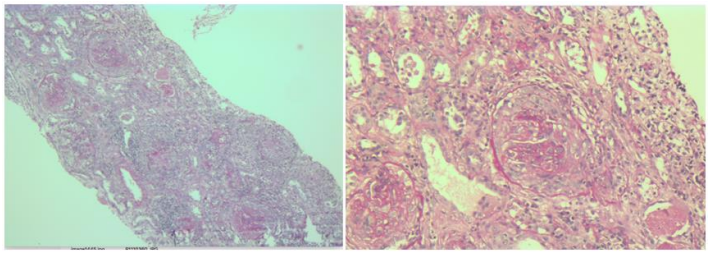

Fig-1 & 2: Optical microscopy: extra-capillary glomerulonephritis

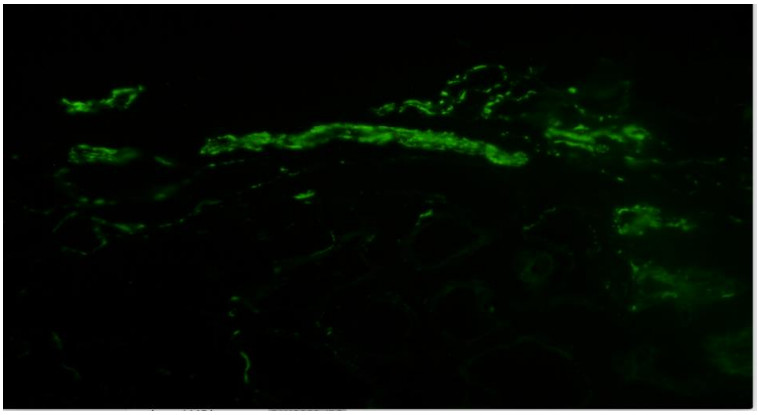

Fig-3: Immunofluorescence: linear deposits of anti-IgG

Regarding therapeutic management, given the low prevalence of this type of vasculitis, no interventional studies have been performed in these patients and no solid therapeutic recommendations can be made [2,3]. Our patient received high-dose of corticosteroids and intravenous bolus of Cyclophosphamide with the need for hemdialysis.

The renal prognosis at one year was severe with end-stage renal failure.

DISCUSSION/CONCLUSION

ANCA and anti-MBG double-positive vasculitis seems to combine the demographic and clinical features of ANCA vasculitis with the histological type and severe renal prognosis of anti-MBG vasculitis [4]. This association is a rare and severe entity, requiring extensive clinical studies detailing therapeutic management.

REFERENCE

1. Philip, R., Dumont, A., Le Mauff, B., Martinet, M., Silva, M., De Boysson, H., ... & Deshayes, S. (2019). ANCA and anti-MBG double-positive vasculitis: An update on the clinical and therapeutic specificities and comparison with the two eponymous vasculitis. *La Revue de Medecine Interne*, *41*(1), 21-26.
2. McAdoo, S. P., Tanna, A., Hrušková, Z., Holm, L., Weiner, M., Arulkumaran, N., ... & Pusey, C. D. (2017). Patients double-seropositive for ANCA and anti-GBM antibodies have varied renal survival, frequency of relapse, and outcomes compared to single-seropositive patients. *Kidney international*, *92*(3), 693-702.
3. Rutgers, A., Slot, M., van Paassen, P., van Breda Vriesman, P., Heeringa, P., & Tervaert, J. W. C. (2005). Coexistence of anti-glomerular basement membrane antibodies and myeloperoxidase-ANCAs in crescentic glomerulonephritis. *American journal of kidney diseases*, *46*(2), 253-262.
4. Kallenberg, C. G. (2015). Pathogenesis and treatment of ANCA-associated vasculitides. *Clin Exp Rheumatol*, *33*(4 Suppl 92), S11-S14.