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ARTICLE ORIGINAL

Vasculitis with antibody to the glomerular basement membrane about 25 cases

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KEYWORDS

Anti MBG

Vasculitis

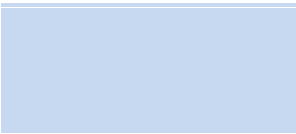
Abstract

Introduction : anti-MBG vasculitis is a rare autoimmune disease of small vessels, characterized by autoantibodies targeting the glomerular and alveolar basement membrane, thus defining renal-pulmonary syndrome.

Methods : We conducted a descriptive, retrospective and monocentric study of anti-MBG vasculitis with renal involvement, diagnosed, treated and followed in nephrology at the CHU Béni-Messous between 2015 and 2025. Demographic, clinical-biological, histological, therapeutic and evolutionary data were collected from medical records.

Results : We retrospectively studied 25 patients with anti-MBG vasculitis over 10 years, with a sex ratio of 1 and a mean age of 55.2 years (16-100 years). Twenty-two percent were masons. Forty percent had a history of High Blood Pressure. Half consulted for general signs (asthenia and deterioration of the general condition), and 3 for macroscopic hematuria. Typical pulmonary involvement (alveolar hemorrhage) was present in 40% of patients. At the time of diagnosis, 90% of patients had severe renal failure requiring dialysis. Anti-MBG antibodies were positive in 77.3% of cases, associated with anti-MPO ANCA positivity in 27.3% of cases; 22.7% were negative and diagnosed by renal biopsy. A histological analysis, performed in 77% of patients, revealed extracapillary circumferential necrosis increasing the lesions. Fifty percent of the patients were already at a terminal stage. From a therapeutic point of view, all received corticosteroids (bolus then oral relay), 22/25 cyclophosphamide and 10 plasmapheresis. Three patients died of intra-alveolar hemorrhage before the start of immunosuppressive therapy. The renal prognosis **was** unfavorable, with dialysis in 21 patients, chronic kidney failure in 1 patient and death in 2 patients after 1 year of progression.

Conclusion : anti-MBG vasculitis is a serious, potentially fatal and functional disease. A rapid diagnosis, based on the search for anti-MBG



antibodies and, if necessary, a renal biopsy (in case of negative antibodies), is crucial to initiate emergency treatment.

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Introduction :

Glomerular Basement Membrane Antibody (anti-MBG) disease, also known as Goodpasture syndrome, is a rare small vessel vasculitis. This autoimmune disease is characterized by the presence of autoantibodies targeting the basement membrane of renal glomeruli and pulmonary alveoli, which leads to a pneumo-renal syndrome. It manifests as a rapidly progressive glomerulonephritis and sometimes alveolar hemorrhages. In 20 to 60% of cases, anti-neutrophil cytoplasmic antibodies (ANCA) are also observed. The therapeutic management must be rapid and combines plasma exchanges, systemic corticosteroids and an immunosuppressant (cyclophosphamide). The objective of our work is to describe the demographic, clinico-biological and histological characteristics of our population, as well as the therapeutic and evolutionary management, and to assess the overall prognosis of the disease in our population.

Materials and methods

We conducted a descriptive, retrospective, monocentric study in the nephrology department of Beni-Messous university hospital, covering the period from January 2015 to December 2024. Patients with an initial diagnosis of glomerular basal membrane disease (anti-MBG) based on the presence of circulating antibodies were included.MBG and/or by renal biopsy showing linear IgG deposits along the glomerular basal membranes.

Medical records were consulted to gather demographic, clinical (age, sex, history of smoking or occupational or toxic exposure, pulmonary and/or renal presentation), biological (creatinine, glomerular filtration clearance, haematuria, proteinuria, anti-MBG antibody titre, ANCA status) and anatomopathological (percentage of crescent glomeruli, interstitial fibrosis). Patients without complete records were excluded. The primary objective was to assess renal survival (defined as stopping dialysis or maintaining native kidney function at 12 months) and overall survival.

Results

Over a ten-year period (2015-2025), we included 25 patients (sex ratio 1) in our study. The mean age at diagnosis was 55.2 years, with extremes ranging from 16 to 100 years. Among these patients, 22% were in a manual profession (masons) and 40% had a history of high blood pressure. Half of the patients (50%) had presented for general symptoms (asthenia and deterioration of general condition), while 3 patients (12%) had consulted for gross hematuria.

Pulmonary involvement of alveolar hemorrhage type was observed in 40% of patients, and isolated renal involvement in 30%. At the renal level, all patients presented with severe insufficiency: creatinine exceeded 600µmol/l and 80% of them were anuric at the time of diagnosis, requiring hemodialysis. Proteinuria and hematuria were present in all cases. All patients (n = 25) also had a biological inflammatory syndrome with an increase in CRP, sedimentation rate, and normochromic normocytic anemia

The search for anti-MBG antibodies (glomerular basement membrane) was positive in 77.3% of cases, and among them, 27.7% showed ANCA positivity with anti-MPO specificity.

Renal biopsy, performed in 20 patients, revealed extracapillary glomerulonephritis with linear IgG deposition in 24 patients (note: even though direct immunofluorescence did not show linear IgG depositions, serum antibodies were present). Half of the patients (50%) were already at the scar stage at the time of diagnosis.

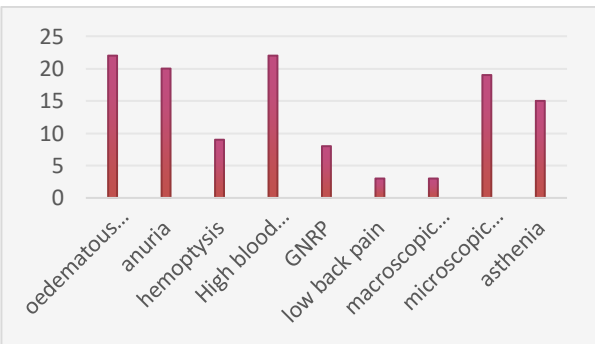


Figure 1. Clinical manifestations

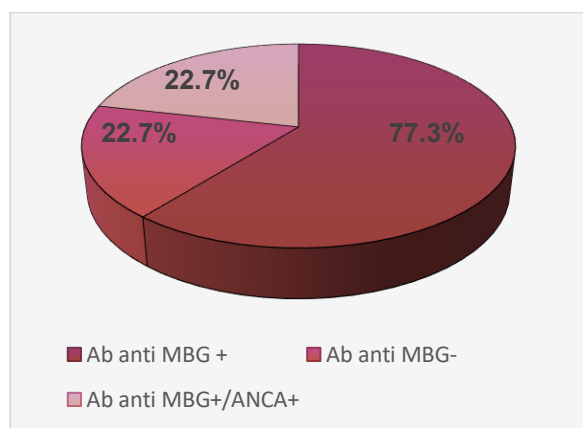
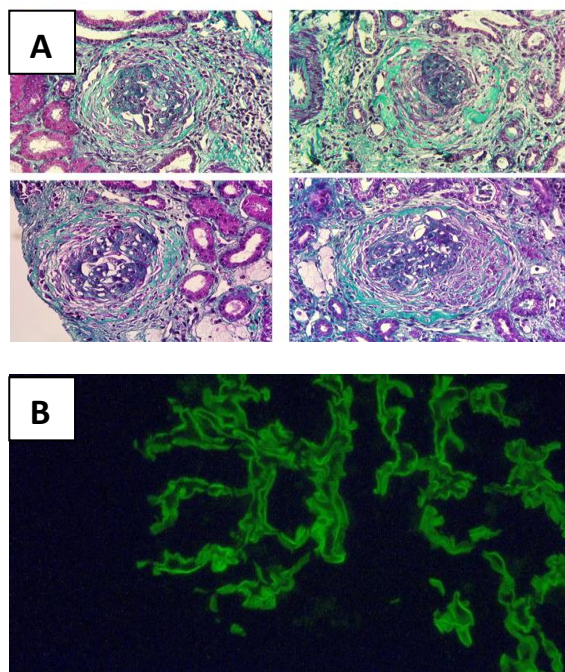


Figure 2. Results of the immunological assessment



Renal biopsy of a 15 year old girl, macroscopic hematuria, Ab anti MBG negatf.

A: MO: Cortico-medullary 10 to 20 Glomeruli, of which 19 G are undergoing transformation into PAC, showing fairly homogeneous lesions, almost of the same age, sclerous Floccus retracted at the center, surrounded by fibrocellular circumferential crescents, rather fibrous, without fibrinoid necrosis.

B: IF: 3 glomeruli: Intense positivity in discontinuous and linear traits along the capillary loops with C3 and IgG.

Therapeutically, corticosteroids were introduced in all patients : 3 bolus of 10 mg/kg/day, then a per os relay at 1 mg/kg/day. Cyclophosphamide induction was used in 22 patients (88%). The other three died of an intra-alveolar hemorrhage before they even received cyclophosphamide. Ten patients (40%) underwent plasmapheresis. After receiving histological results, immunosuppressive therapy was discontinued in 55% of patients who

had renal impairment only and advanced stage, and maintained (with plasmapheresis) in 7 patients (28%) presenting with pulmonary involvement despite a poor renal prognosis. Azathioprine (Imurel®) was used as a maintenance treatment in a patient who remained immunologically positive.

The renal prognosis was unfavorable: 21 of the 22 patients followed lost kidney function, only one partially recovered the function. Three patients died after one year of progression: two from intra-alveolar hemorrhage and one from septic shock .

Discussion

We noted in our study that the average age of diagnosis is 55.2 years with extremes of 16 - 100 years. This suggests that the disease with anti-glomerular basal membrane antibody (anti-MBG) can affect both young and elderly subjects, which echoes what is described in the literature on the bimodal distribution of the disease. Sex ratio 1 indicates a nearly equal male/female distribution in our series. This is in line with some studies that do not show very clear sexual bias, especially at a later age [1]

The role of environmental factors in the onset of disease is suspected [2], 22% of our patients were engaged in a manual profession (masons), this may evoke a role of environmental or professional exposure (e.g., dust, solvents) in the onset of anti-MBG disease. The classic presentation of the disease characterized by a pneumo-renal syndrome concerns 60 to 80% of patients, while 20 to 40% of patients have isolated rapidly progressive glomerulonephritis [2], pulmonary involvement (alveolar hemorrhage) was observed in 40% and the isolated renal involvement in 30% shows that the typical presentation «lungs + kidneys» is present but not systematic. On the renal level, all patients had severe insufficiency (creatinine > 600 µmol/L) and 80% were anuric at diagnosis. This indicates that in our series, patients were already at a very advanced stage of the disease at the time of diagnosis. These conditions (very high creatinine, anuria) are of poor prognosis according to the literature.

The diagnosis of the disease is based on the search for an IgG1 isotype against the basal membrane of the circulating glomerular. Their research is a key diagnostic examination and must be carried out within 24 hours of admission to limit any diagnostic delay [3], The anti-MBG antibodies returned positive in 77.3%, of which 27.7% was associated with a positivity of MPO-specific ANCA, which shows that our series covers both the "classic" forms and the so-called "double positive" forms (anti-GBM + ANCA). The coexistence of anti-MBG antibodies and ANCA, or "double positivity", is more common than assumed by chance, with rates ranging from 21 to 60% (37,38). ANCA with anti-myeloperoxidase specificity (MPO) predominate, characterized in 66 to 81% of

double-positive patients. The "double positives" (anti-GBM + ANCA) are described as having a more complex profile, sometimes better renal survival if early management, but also increased risk of recurrence [4].

The renal biopsy performed in 20/25 showed extracapillary glomerulonephritis with linear IgG deposits in 19 patients (1 patient the direct immunofluorescence did not show linear IgG deposits, serum antibodies were present) : this emphasizes that some cases may be atypical or that the technique can be limiting. Half of the patients were already at the scar stage at the time of diagnosis. All patients received corticosteroid therapy, the majority (88%) received cyclophosphamide induction treatment, and 40% benefited from plasmapheresis. The treatment was in accordance with the classical recommendations of the anti-GBM disease [5]. The fact that in 55% of patients, immunosuppressive treatment was stopped (in cases of renal disease only and advanced stage) while in 28% it was maintained (pulmonary disease despite poor renal prognosis) reflects a pragmatic strategy taking into account the very bleak prognosis in case of major kidney injury. This confirms the delay in diagnosis and undoubtedly explains the poor recovery of kidney function.

Renal prognosis was very poor: 21 of the 22 patients followed lost kidney function, only one partially recovered. In addition, three deaths (two from intra-alveolar hemorrhage and one from septic shock) after a year of evolution. This rate of kidney function loss is extremely high but consistent with the fact that the majority of patients had a very poor renal condition at diagnosis (creatinine > 600 $\mu\text{mol/l}$, anuria). The literature indicates that kidney function at diagnosis is the best predictor of recovery. In a review of 79 Swedish patients with anti-GBM, at 6 months 34% were deceased, 41% on dialysis, only 25% alive with functional kidney [6]. The best predictor of renal survival was renal function at diagnosis. Your series goes in the same direction, but with even more severe results, probably because the population was already in a very advanced stage.

Conclusion

Our study highlights an often-dreaded clinical reality: despite the implementation of recognized treatments, when anti-MBG disease is diagnosed late, with severe renal failure and anuria, the renal

prognosis remains very unfavorable. This reinforces the importance of clinical vigilance, early diagnosis, and possibly increased screening in at-risk situations. Moreover, the significant proportion of forms associated with ANCA or seronegative invites a nuanced look at this autoimmune pathology.

Conflict of interest statement

No conflicts of interest to be disclosed.

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