OMCR

Oxford Medical Case Reports, 2019;2, 59–60

doi: 10.1093/omcr/omy124
Clinical Image

CLINICAL IMAGE

A case report of double negative anti-GBM disease

Babikir Kheiri[1,*], Mohammed Osman[1], Farah Elounais[1], Yanal Alnimer[1], Ali K Owda[2], Imad Modawi[2] and Basim Towfiq[1]

[1]Internal Medicine Department, Hurley Medical Center/Michigan State University, Flint, MI 48503, USA, and [2]Department of Nephrology, Hurley Medical Center/Michigan State University, Flint, MI 48503, USA

*Correspondence address. Hurley Medical Center/Michigan State University, Internal Medicine Department, Two Hurley Plaza, Ste 212, Flint, MI 48503, USA. Tel: +1 (810) 262-9080; Fax: +1 (810) 262-7245; E-mail: bkheiri1@hurleymc.com

Abstract

Goodpasture's disease is a life-threatening autoimmune disease that can lead to end stage renal disease and death. We report a case of 61-year-old female who presented with deteriorating renal function. Initial laboratory investigations were negative for autoimmune antibodies including negative anti-neutrophilic cytoplasm antibodies and anti-glomerular basement membrane (anti-GBM) antibodies using both enzyme-linked immunosorbent assay and indirect immunofluorescence. However, renal biopsy was positive for linear IgG staining. Despite starting plasmapheresis and corticosteroids treatment, her renal functions continued to deteriorate and she was started on regular hemodialysis. This case highlights the challenging presentation and diagnosis of anti-GBM disease, which requires a high clinical suspicion necessarily for early diagnosis and treatment to improve survival rates.

CASE REPORT

A 61-year-old female presented with complaints of fatigue, exertional shortness of breath, hemoptysis and intermittent hematuria for 1 month. Vitals signs were unremarkable. Chest examination revealed decreased air entry with basal crackles and abdomen was distended and positive for ascites with 1+ pitting edema. Investigations showed elevated creatinine at 6.7 (Ref: 0.5–1.1 mg/dl), blood urea nitrogen of 61 (Ref: 6–20 mg/dl), normal liver function tests with albumin of 3.4 (Ref: 3.4–4.8 g/dl) and negative hepatitis panel (A–C). Urinalysis showed 4+ protein, 3+ blood with no nitrite/leukocyte esterase. Bence Jones Protein was negative by electrophoresis. She also had negative antinuclear antibodies, ant-neutrophilic cytoplasmic antibody (p- and c-ANCA), anti-double stranded DNA and normal total complement (C3 and C4), which were reported within 2 days of admission. Her anti-GBM was negative using both enzyme-linked immunosorbent assay and indirect immunofluorescence and was performed twice, 2 weeks apart (results were available on days 3 and 18 from admission, respectively). She underwent renal biopsy on day 4 of admission and the result was positive for crescentic glomerulonephritis with segmental scars and fibrous crescents (reported on day 19; Fig. 1a). Immunofluorescence was positive for linear IgG staining, which is consistent with anti-GBM glomerulonephritis (Fig. 1b). She was started on plasmapheresis and corticosteroids on day 21 of admission. However, no significant improvement of her symptoms, serum creatinine or urine output was achieved after six cycles of plasmapheresis and, therefore, she was started on hemodialysis.

DISCUSSION

Goodpasture's disease, first described by Ernest Goodpasture in 1919 [1], is rare autoimmune disease characterized by autoantibodies mostly directed against the alpha3 chain of type IV non-collagenous domain in the lung and kidneys [2]. It typically presents as rapidly progressive crescentic glomerulonephritis and/or pulmonary hemorrhage [3]. Although more than 90% of

Received: May 15, 2018. **Revised:** October 20, 2018. **Accepted:** November 14, 2018

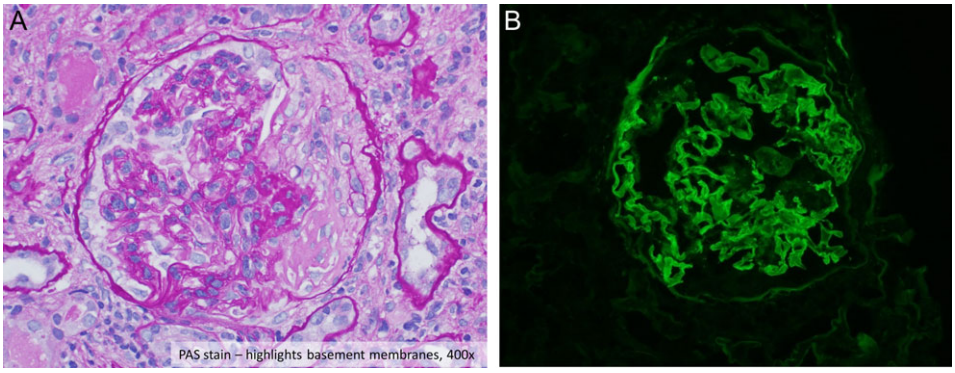

Figure 1: Glomerulus with a fibrocellular cresent (A) and immunofluorescence showing linear pattern IgG (B).

patients have detectable anti-GBM antibodies, there are few cases reported of negative circulating anti-GBM with negative ANCA [4]. Therefore, the diagnosis requires the demonstration of linear immunofluorescent deposits along the glomerular and/or alveolar basement membranes [3]. Successful management is largely dependent on the clinical presentation, biochemical level and histopathological features. Nevertheless, the rationale behind medical therapy is to eliminate the circulating antibodies (plasmapheresis) and to suppress the production of the anti-GBM antibodies (cyclophosphamide and corticosteroids) [5].

ACKNOWLEDGMENTS

We would like to thank Katherine Negele, editorial assistant, research department, Hurley Medical Center, for assistance with manuscript editing.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

None

ETHICAL APPROVAL

Not required.

CONSENT

Consent has been obtained from the patient.

GUARANTOR

Babikir Kheiri and Mohammed Osman.

REFERENCES

1. Goodpasture E. The significance of certain pulmonary lesions in relation to the etiology of influenza. *Am J Med Sci* 1919;**158**:863–70.
2. Hudson BG, Tryggvason K, Sundaramoorthy M, Neilson EG. Alport's syndrome, Goodpasture's syndrome, and type IV collagen. *N Engl J Med* 2003;**348**:2543–56.
3. Greco A, Rizzo MI, De Virgilio A, Gallo A, Fusconi M, Pagliuca G, *et al.* Goodpasture's syndrome: a clinical update. *Autoimmun Rev* 2015;**14**:246–53.
4. Salama AD, Dougan T, Levy JB, Cook HT, Morgan SH, Naudeer S, *et al.* Goodpasture's disease in the absence of circulating anti-glomerular basement membrane antibodies as detected by standard techniques. *Am J Kidney Dis* 2002;**39**:1162–7.
5. McAdoo SP, Pusey CD. Anti-glomerular basement membrane disease. *Clin J Am Soc Nephrol* 2017;**12**:1162–72.