Case Report

Anti-GBM antibody disease sans crescents with thrombotic microangiopathy

Swarnalata Gowrishankar¹, Anil Patro² and Sanjay Maitra²

¹Department of Anatomic Pathology and Cytology and ²Department of Nephrology, Apollo Hospitals, Jubilee Hills, Hyderabad, Andhra Pradesh, India

Correspondence and offprint requests to: Swarnalata Gowrishankar; E-mail: swarnalata@gmail.com

Abstract

We report a case of a 38-year-old male with acute renal failure, elevated anti-glomerular basement membrane (anti-GBM) antibody titres, bilateral nodular lung opacities and hypertension. In the renal biopsy examination, whereas direct immunofluorescence revealed significant peripheral linear deposits of IgG typical of anti-GBM antibody disease (Goodpasture’s disease), eosin–haemotoxylin staining showed glomerular and vascular changes typical of thrombotic microangiopathy (TMA) and without crescents. We postulate that the TMA was responsible for the acute renal failure and that antibodies, though demonstrable, were not adequate at the site of the glomerular basement membrane to elicit a crescentic response, because of occlusion of the vascular lumina by the thrombotic process. The patient remained dialysis dependent at a 3-month follow-up.

Keywords: anti-GBM antibody disease; Goodpasture’s disease; thrombotic microangiopathy

Introduction

Anti-glomerular basement membrane (anti-GBM) antibody disease with or without lung involvement is the prototype of a crescentic glomerulonephritis, with varying crescents in the renal biopsy and with peripheral linear deposits of IgG on immune study. Thrombotic microangiopathy (TMA) characterized by thrombotic events in the microvasculature is also an uncommon cause of a rapidly progressive renal failure and has been reported in conjunction with conditions such as membranous glomerulonephritis, ANCA-associated vasculitides and SLE. TMA in association with anti-GBM antibody disease is distinctly rare. We report one such case of Goodpasture’s disease, where the renal biopsy had findings of TMA sans crescents.
Anti-GBM antibody disease sans crescents with thrombotic microangiopathy

Fig. 1. Interlobular artery showing prominent intimal myxoid thickening, with an adjoining arteriole (arrowhead) showing fibrin thrombus occluding lumen. The glomerulus shows mild ischaemic changes with no crescent (HE100 ×).

Fig. 2. Peripheral linear deposits of IgG. No crescent (FITC-labelled anti-IgG, 200 ×).

The patient was continued on a further nine courses of total plasma exchange and methyl prednisolone, at the end of which the anti-GBM titre was only weakly positive. At 3 months of follow up, he continues to remain dialysis dependent.

Discussion

Crescentic glomerulonephritis is the hallmark of anti-GBM antibody disease and is seen in up to 97% of these patients with >50% crescents seen in 85% of them. TMA in association with anti-GBM antibody disease is rare, and the reported cases have essentially been crescentic glomerulonephritis with superimposed TMA [1–4]. It has been suggested that the TMA may contribute to the anaemia and renal failure in these patients.

However, there are no reports of a case similar to ours with acute renal failure, lung involvement, positive anti-GBM antibody titres and the biopsy showing significant linear deposits of IgG without any crescents and with features of TMA. We postulate that the cause for renal failure in this young patient was thrombotic microangiopathy probably secondary to accelerated hypertension. He had an underlying Goodpasture’s disease as evidenced by the high titre of anti-GBM antibodies, lung opacities and peripheral linear deposits of IgG in the glomeruli. However, occlusion of the microvasculature secondary to TMA prevented adequate antibodies from reaching the glomerular basement membrane and eliciting the crescentic response. This case taken together with the earlier reports [1–4] also suggests the existence of a possible etiopathogenetic association between TMA and Goodpasture’s disease that needs to be further explored.

Acknowledgement. We acknowledge Prof. Jan Weening for his insights into the pathogenesis.

Conflict of interest statement. None declared.
References

1. Stave GM, Croker BP. Thrombotic microangiopathy in anti-glomerular basement membrane glomerulonephritis. *Arch Pathol Lab Med* 1984; 108: 747–751

2. Stallworthy E, Yehla M. Thrombotic microangiopathy in a patient with anti-glomerular basement membrane antibody disease. *Nephrology* 2006; 11: 375–376

3. Terryn W, Benoit D, Van Loo A et al. Goodposture’s syndrome associated with autoimmune thrombotic thrombocytopenic purpura—an unusual case. *NDT* 2007; 22: 3672

4. Fu KL, Kai CT, Man FL et al. Incidence and outcome of antiglomerular basement membrane disease in Chinese. *Nephrology* 2004; 9: 100–104

*Received for publication: 01.4.09; Accepted in revised form: 21.4.09*