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Atypical Presentation of Anti GBM Disease with Co Existent Membranous Nephropathy – Case Report

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Background: Anti Glomerular Basement Membrane (GBM) disease is characterised by auto-antibodies against an antigen intrinsic to the basement membrane of the glomeruli and alveoli. Presentation is typified by a rapidly progressive glomerulonephritis and 40-60% of cases will have concurrent pulmonary haemorrhage. Indolent presentation with haemo- proteinuria and minimal renal dysfunction is not well reported.

Case Report: A 17 year old male, an active smoker, presented with a prodrome of fever, fatigue and weight loss of one month duration. He had two episodes of macroscopic haematuria initially; No haemoptysis. Clinical examination was unremarkable except for cervical lymphadenopathy. Investigations showed slightly elevated creatinine at 105 umol/L, mild transaminitis and reactive lymphocytosis. Urinalysis showed 3+ blood and protein with urinary albumin:creatinine ratio (ACR) of 127 mg/mmol. Monospot test was positive. Hence Infectious Mononucleosis was diagnosed. Further tests showed anti GBM titre of 11 iu/ml (0-7 iu/ml) with Anti Streptolysis O Titre of 400 unit/ml, normal C3 and C4 levels. ANCA and ANA were negative. He subsequently underwent renal biopsy - From a total of 27 glomeruli, segmental necrosis seen in one glomerulus, fibrocellular segmental lesion in two, and fibrocellular crescents in two. Another glomerulus showed a fibrous crescent. Fifteen glomeruli were hypercellular with segmental areas of mesangial and/or endocapillary proliferation. Immunohistochemistry showed linear staining of GBM with IgG4, IgG and C1q. IgA and C3c were negative. These features were in keeping with anti-GBM disease. Electron microscopy showed subepithelial deposits consistent with co-existent membranous glomerulopathy (MN). When biopsy result was available, the anti-GBM titre was at 6 iu/ml and creatinine had normalised to 85 umol/L. However, he continued to have haemo-proteinuria with urinary ACR of 95 mg/mmol. Anti-PLA2R antibodies were negative. The patient was treated with oral Prednisolone and started on an increasing dose of Mycophenolate Mofetil to 1g twice daily. Subsequent anti GMB titre was 1iu/ml.

Discussion: In this case, fever and constitutional symptoms were likely due to Infectious Mononucleosis. The main clinical differentials for the renal abnormalities were IgA nephropathy vs post infectious glomerulonephritis. However, the raised anti-GBM titre, confirmed twice with a peak of 11 IU/ml, was of great concern given the aggressive nature of anti-GBM disease. Renal pathology appearances were of anti-GBM disease although showing atypical features with focal glomerular lesions and co existent MN. The production of anti-GBM antibodies is thought to be in response to an unknown inciting stimulus and may precede the onset of clinical signs and symptoms by many months. In our case, Infectious Mononucleosis possibly triggered formation of anti-GBM antibodies. The decision to initiate immunosuppress here was based on crescentic glomerular disease, in the context of anti-GBM antibodies with the potential to cause a rapidly progressive glomerulonephritis.

Conclusion: Our case showed co-existent anti-GBM disease and MN in renal biopsy in the context of Infectious Mononucleosis which to our knowledge is not previously reported in literature. Although the presentation in this case was indolent, biopsy confirmed the onset of crescentic glomerular disease warranting immunosuppression. The outcome was excellent, with disappearance of the pathogenic anti-GBM antibodies.