

The invincible kidney or a disaster waiting to happen? An atypical case of anti-GBM disease with an isolated pulmonary presentation

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Introduction

The majority (80-90%) of patients with anti-GBM disease present with rapidly progressive glomerulonephritis and 40-60% with concomitant pulmonary haemorrhage (1). However, atypical presentations of anti-GBM disease have been described (2, 3, 4). We report such a case of a young male with isolated pulmonary involvement and weakly positive GBM serology who was found to have typical kidney biopsy appearances.

Case Report

A 21-year-old Caucasian man presented to the respiratory clinic with 4-months of daily haemoptysis (mixture of fresh blood and clots). His past medical history comprised solely of intermittent eczema and his family history was unremarkable. He took no regular medications. He reported smoking 10 cigarettes/day with occasional use of cannabis and cocaine. He worked as a self-employed electrician. On examination, he was afebrile and normotensive with saturations of 96% on room air. His chest was clear. His urine dip showed 2+ blood but no protein.

Initial investigations revealed a microcytic anaemia (Hb 95g/L, MCV 79fL, ferritin 104ng/ml), normal platelets and normal coagulation. His CRP was 18. He had normal renal function (creatinine 62umol/L, eGFR>90ml/min) and unobstructed, average-sized kidneys. His initial chest x-ray demonstrated clear lung fields. However, a CT performed subsequently showed bilateral widespread patchy ground-glass appearances in an alveolar distribution and borderline splenomegaly (15cm). He proceeded to have a bronchoscopy that identified light growth of staph aureus but no fungus/mycobacterium. Tests for TB and respiratory viruses also returned negative. Pulmonary function tests did not reveal a raised transfer factor.

An autoimmune panel returned predominantly unremarkable (anti-MPO negative, anti-PR3 negative, normal complements, connective tissue disease screen negative). However, the identification of weakly positive anti-GBM antibodies (14U/ml) almost a month following presentation prompted admission. He was pulsed with intravenous methylprednisolone (500mg) and he then underwent a kidney biopsy which revealed that 1/52 glomeruli exhibited cellular proliferation within the bowman's space, possibly representing an early crescent. There was no evidence of inflammation, necrosis or vascular abnormalities. Staining with IgG and C3 in a typically linear pattern was found on immunofluorescence.

The patient subsequently underwent 4 consecutive plasma exchange sessions and received a dose of 1g rituximab. By this stage his anti-GBM titre had reduced to 2.7U/L. He was thereby discharged on 40mg prednisolone only to represent days later with haemoptysis and a similar anti-GBM titre. Oral cyclophosphamide (2mg/kg) was started and he received a second dose of rituximab 2 weeks later resulting in complete B cell depletion. As he continued to suffer haemoptysis, oral cyclophosphamide was continued until complete resolution of symptoms 7 weeks later. He remains well without further haemoptysis, undetectable anti-GBM antibodies, normal renal function and does not take any medications.

Discussion

Although rare, similar presentations of pulmonary-isolated anti-GBM disease have been described (2, 3, 4). Whilst these presentations with negative or weakly positive anti-GBM serology may appear to suggest a disease subclass with renal sparing, these cases could also represent early detection before potentially devastating kidney damage (2). It is therefore imperative to suspect anti-GBM disease even in the context of seemingly unscathed kidneys.