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# A 12-year-old boy with renal failure and haemoptysis

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## Case

A 12-year-old boy, previously well, presented with increasing pallor, lethargy and intermittent gross haematuria for 1 month. He also had a history of haemoptysis for the previous 5 days. His blood pressure was normal and he had pedal oedema. There was no hepatosplenomegaly, joint swelling or skin rash. Laboratory investigations (Table 1) revealed anaemia, hypoalbuminaemia and elevated serum creatinine (estimated glomerular filtration rate by Schwartz formula: 13 mL/min/1.73 $m^2$). His white cell count, platelet count and clotting profile were normal. His urine analysis showed 2+ protein, 3+ blood and coarse granular casts. He had nephrotic-range proteinuria. His chest X-ray showed bilateral nodular infiltrates (Figure 1A) and a CT of the chest revealed bilateral diffuse ground glass opacities consistent with alveolar haemorrhage (Figure 1B). A kidney biopsy was performed for further evaluation.

## Question

What is the diagnosis?

## Answer

Microscopic polyangiitis

## Discussion

The ANCA (anti nuclear antibody)-associated vasculitis (AAV) includes granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), eosinophilic GPA (EGPA or Churg-Strauss syndrome) and renal-limited vasculitis [1]. A positive ANCA with a cytoplasmic pattern (c-ANCA; anti-proteinase 3 specificity) is highly associated with GPA, whereas MPA has a perinuclear pattern ANCA (p-ANCA; usually with antimyeloperoxidase specificity) [2]. The clinical manifestations of AAV range from non-specific constitutional symptoms to multi-organ involvement and life-threatening vasculitis. The kidney is the most commonly involved organ, clinical manifestations of which include haematuria, proteinuria and elevated serum creatinine [3]. Pulmonary involvement can present as pulmonary renal syndrome in the setting of renal failure. Patients mostly present as intra-alveolar haemorrhage with consequent cough, dyspnoea and haemoptysis [2]. Our patients had strongly positive p-ANCA (75 U/mL, normal: 2 U/mL), negative c-ANCA and no upper airway symptoms, thus pointing towards the diagnosis of microscopic polyangiitis. MPA is rare in children and the median age of onset is 50 years with male preponderance [4]. In contrast to adult data, female preponderance is reported in the paediatric literature with a peak age of onset of 11–12 years. Renal involvement has been reported in 100% of children and pulmonary renal syndrome in 30–50% of children [4].

The renal biopsy in all types of AAV typically shows pauci-immune necrotizing glomerulonephritis [4]. The kidney biopsy of our patient showed focal sclerosing and proliferative glomerulonephritis with fibrocellular to fibrous crescents in 6 out of 14 glomeruli (Figure 2). The tubulointerstitial compartment showed tubular atrophy, interstitial fibrosis and infiltrates of neutrophils, lymphocytes and plasma cells. The immunoflourescence was negative.

The management of AAV generally follows the principle of initial aggressive induction therapy with potent immunosuppressive agents with an aim to control disease activity [5]. This is followed by a prolonged maintenance phase to prevent disease flares. Plasma exchange is usually reserved for severe kidney involvement (serum creatinine >500 μmol/L), concurrent anti-GBM antibody positivity and active pulmonary haemorrhage [5]. There is limited data on long-term outcome of patients with MPA. Patient survival is reported as 77–100% at 1 year, 46–80% at 5 years and 60–80% at 10 years [6]. Progression to end-stage renal disease is seen in a third of the patients. The initial severity of renal disease, chronicity on renal biopsy and delay in diagnosis are important prognostic determinants of long-term renal outcome [6]. Our patient was managed with intravenous pulse

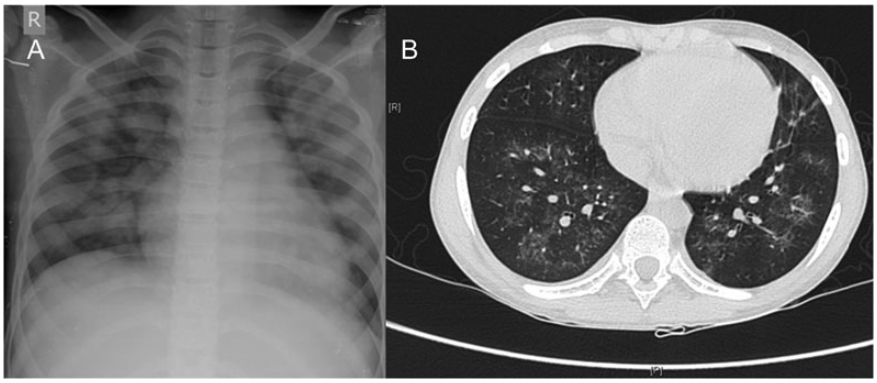


**Fig. 1.** (**A**) Chest X-ray showing bilateral nodular infiltrates and (**B**) high-resolution CT chest (transverse view) showing diffuse patchy ground glass opacities bilaterally resulting from alveolar haemorrhage.

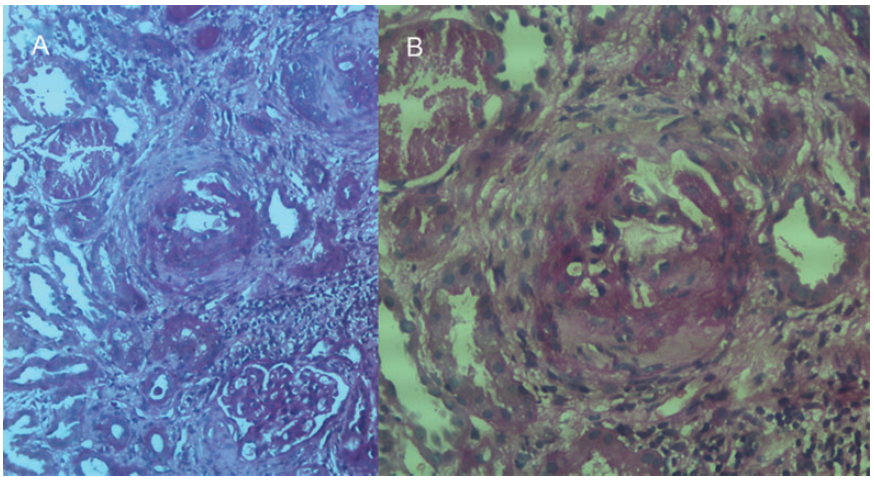


**Fig. 2.** (**A**) Light microscopy showing proliferative and sclerosing glomerulonephritis (PAS 20×) and (**B**) a glomerulus with fibrocellular to fibrous crescent (PAS 40×).

**Table 1.** Laboratory results of our patient

| Investigation | Result |
|---|---|
| Haemogobin | 6.8 g/dL |
| Reticulocyte count | 1.3% |
| Serum creatinine | 4.7 mg/dL (415.5 μmol/L) |
| Serum albumin | 3.1 g/dL |
| Spot early morning urine protein–creatinine ratio | 5.5 mg/mg |
| C3; C4 | C3 101 mg/dL (90–180); C4 50 mg/dL (10–40) |
| Spot early morning urine protein–creatinine ratio | 5.5 mg/mg |
| Anti-GBM antibody | Negative |
| ANA | Negative |
| p-ANCA (normal: 2 U/mL) | 75 U/mL |
| c-ANCA | <2 U/mL |

ANA, anti-nuclear antibody; anti-GBM, anti-glomerular basement membrane; p-perinuclear c-cytoplasmic ANCA, anti-neutrophilic cytoplasmic antibody.

methylprednisolone for 5 days followed by oral prednisolone 2 mg/kg/day. In addition, he received haemodialysis and a total of 14 sessions of double-volume plasma exchange. Oral cyclophosphamide therapy was also started. His haemoptysis subsided after 14 days but there was no improvement in his renal function. The child was eventually started on chronic peritoneal dialysis and discharged in stable condition.

## Conclusions

Rapid diagnosis and management is imperative in a child with renal failure and ongoing haemoptysis (pulmonary renal syndrome) and AAV should be considered in the differential diagnosis. Early recognition and aggressive management may improve outcome.

*Conflict of interest statement.* None declared.

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