CASE REPORT

ANCA and IgA glomerulonephritis all in one: prognosis and complications

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SUMMARY

We present the case of a 75-year-old Hispanic woman with known stage 3 chronic kidney disease, long-standing hypertension and type 2 diabetes mellitus who presented with right-sided abdominal pain and acute kidney injury, nephrotic range proteinuria with positive antimyeloperoxidase antibody. A renal biopsy revealed IgA nephropathy with superimposed pauci-immune antineutrophilic cytoplasmic antibody (ANCA)-associated crescentic glomerulonephritis. The patient was treated with pulse intravenous methylprednisolone, cyclophosphamide and plasmapheresis. One week after her second dose of cyclophosphamide, she was readmitted for infectious complications including influenza A respiratory infection, *Rothia* bacteraemia associated with diarrhoea and herpes zoster of the trunk. In this report, we review the prevalence, treatment and prognosis of coexistent IgA nephropathy and pauci-immune ANCA-associated crescentic glomerulonephritis. We propose that a reduced-dose treatment regimen should be considered in elderly patients due to their higher risk of infectious complications. Current literature suggests that this treatment approach may reduce infectious complications without compromising therapeutic efficacy.

BACKGROUND

Coexistent IgA nephropathy and pauci-immune antineutrophilic cytoplasmic antibody (ANCA)-associated crescentic glomerulonephritis represents the rare concurrence of two common forms of glomerulonephritis. The pathogenesis, treatment and prognosis of this dual glomerulopathy are not well defined. This case illustrates how aggressive therapy of this condition with standard dosing of immunosuppressive agents can place the elderly patient at risk for serious infectious complications. In this report, we review the literature on this dual glomerulonephritis, discuss pathological, clinical and pathogenetic implications and make recommendations for a modified treatment schedule to minimise risks without compromising therapeutic efficacy in the elderly patient population.

CASE PRESENTATION

A 75-year-old Hispanic woman with medical history of long-standing hypertension, non-insulin depend diabetes mellitus (NIDDM) (HgbA1c –7.7), chronic kidney disease stage 3 (estimated glomerular filtration rate (eGFR)

46 mL/min/1.73 m²) and hyperlipidaemia initially presented to the emergency department (ED) for right-sided abdominal pain of 3 days' duration. The patient was in her usual state of health until 1 month prior, when she was treated for pneumonia with azithromycin at an outside hospital. One week later (3 weeks prior to this ED presentation), she noticed malaise, fatigue and loss of appetite. The patient then developed a sharp right-sided, non-radiating abdominal pain, which was associated with non-bloody, non-bilious vomiting. She also noticed the onset of gross haematuria. She denied any history of nephrolithiasis, dysuria, fever or chills. Aside from night sweats for several weeks, she denied cough, shortness of breath or other systemic complaints. There was no family history of kidney disease and no toxic exposures or habits.

On physical examination, the patient exhibited a blood pressure of 155/71 mm Hg, heart rate of 78/min, temperature of 98.8°F and respiratory rate of 18/min. She appeared comfortable and was in no acute distress. Her lungs were clear, with a normal cardiac examination. Her abdominal examination was non-tender and soft. There was right costovertebral angle tenderness. Her lower extremities showed no oedema, and there was no evidence of skin rash. The rest of her examination was within normal limits.

INVESTIGATIONS

The laboratory tests are listed in [table 1](#). Given the acute nature of patient's kidney injury with nephrotic range proteinuria, an investigation into aetiologies of rapidly progressive glomerulonephritis was commenced ([table 1](#)). Her chest X-ray showed a right lower lobe infiltrate, which was attributed to her recent pneumonia. Ultrasonography demonstrated normal-sized kidneys measuring 12.4 cm on the right and 11.1 cm on the left with mild bilateral subcapsular oedema. The patient was admitted to the medical intensive care unit. The patient was empirically given 7 days of ceftriaxone for the treatment of presumed urosepsis, based on the presence of pyuria, leucocytosis, night sweats and acute kidney injury. However, her renal function failed to improve after 4 days and intermittent haemodialysis was initiated via a temporary dialysis catheter. On day 5 of hospitalisation, the high-titre positive antimyeloperoxidase (anti-MPO) serology result was obtained and she was begun on pulse intravenous

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Table 1 Laboratory parameters

	On admission
Urinalysis	Cloudy, large blood, >300 protein, negative leucocyte esterase, negative nitrite, negative glucose, pH 5.5, specific gravity 1.015
Urine microscopy	Many monomorphic red blood cells, few leucocytes clumped together, few bacteria, no casts
Urine protein to creatinine ratio (mg/g)	8242.1
Leucocyte count ($10^9/L$)	20.1 (3.5–11)
Granulocytes (%/##/nL)	74.7/15.01 (48–82) (1.7–9.0)
Lymphocytes (%/##/nL)	18/3.77 (19–48) (1.2–35)
Monocytes (%/##/nL)	5.6/1.13 (4–12) (0–1.1)
Eosinophils (%/##/nL)	0.5/0.10 (0–3%) (0–0.7)
Basophils (%/##)	0.4/0.09 (≤ 2) (≤ 2.0)
Haemoglobin (g/dL)	10.6 (12–16)
Haematocrit (%)	32.9 (36–46)
Platelets (cells/mm ³)	275 (150–440)
Sodium (mEq/L)	144 (135–145)
Potassium (mEq/L)	5.9 (3.5–5)
Chloride (mEq/L)	105 (98–108)
Bicarbonate (mEq/L)	19.4 (24–30)
BUN (mg/dL)	101 (5–26)
Creatinine (mg/dL)	9.6 (0.1–1.5)
PT/INR (s/ratio)	10.9/1.02 (9.5–12.2/0.9–1.2)
aPTT (s)	23.6 (20.1–31.2)
Fibrinogen (mg/dL)	66.8 (200–400)
Albumin (g/dL)	3.2 (3.5–5.5)
AST (U/L)	17 (1–40)
ALT (U/L)	12 (1–40)
Total bilirubin (mg/dL)	0.2 (0.1–1.2)
Direct bilirubin (mg/dL)	0.2 (0.1–0.6)
Lactate (mmol/L)	1.3 (0.3–1.3)
Serum protein electrophoresis	
Total protein (g/dL)	7.1 (6.0–8.5)
Albumin (%)	31.3 (52–62)
Alpha 1 (%)	3.9 (2.5–5.0)
Alpha 2 (%)	21.6 (7.0–13.0)
Beta	13.5 (8.0–14.0)
Gamma	29.7 (12–22)
Comment	Polyclonal increase in the gamma region with increase in alpha 2 region
Urine protein electrophoresis	
Protein Ur (mg/dL)	360 (1–14)
Albumin (%)	49.9 (57.8–65.9)
Alpha 1	6.8
Alpha 2	10.4
Beta	8.3
Gamma	24.6
Comment	All fractions present
Myeloperoxidase Ab (units)	113.9 (normal range ≤ 20.0)
Proteinase-3 (units)	8.2 (normal range ≤ 20.0)
GBM Ab IgG (U)	<0.2 (normal <1.0 negative)
C3 complement (mg/dL)	139 (90–170)
C4 complement (mg/dL)	44 (10–40)
ESR (mm/hour)	100 (0–30)
CRP (mg/L)	78.2 (0–5)
ANA titre	Homogenous, 1:80
SSA/SSA Index (normal range) (AU/mL)	Negative/29 (≤ 99)

Continued

Table 1 Continued

	On admission
SSB /SSB Index (normal range) (AU/mL)	Negative/10 (≤ 99)
Anti-Smith Ab (SM)/SM Index (AU/mL)	Negative/9 (≤ 99)
Anti-ribonucleoprotein (RNP) Ab/RNP Index (AU/mL)	Negative/50 (≤ 99)
SCL-70/SCL-70 Index (AU/mL)	Negative/39 (≤ 99)
Jo 1/Jo 1 Index (AU/mL)	Negative/8 (≤ 99)
dsDNA/dsDNA Index (IU/mL)	Negative/25 (≤ 99)
Centromere/Centromere Index (AU/mL)	Negative/3 (≤ 99)
Histone/Histone Index (AU/mL)	Negative/18 (≤ 99)

Numbers in parentheses present normal range.

ALT, alanine transaminase; ANA, antinuclear antibody; aPTT, activated partial thromboplastin time; AST, aspartate transaminase; CRP, C reactive protein; dsDNA, double-stranded DNA; ESR, erythrocyte sedimentation rate; GBM, glomerular basement membrane; INR, international normalised ratio; PT, prothrombin time; RNP, ribonucleoprotein; SM, smith; SSA, anti-Ro; SSB, anti-La.

methylprednisolone 1 g daily for 3 days, followed by a kidney biopsy.

DIFFERENTIAL DIAGNOSIS

- ▶ Microscopic polyangiitis
- ▶ Granulomatosis with polyangiitis
- ▶ Crescentic IgA nephropathy
- ▶ Acute infection-related glomerulonephritis
- ▶ Minimal change disease with acute tubular injury
- ▶ Collapsing focal segmental glomerulosclerosis
- ▶ Renal pathology

Of the 13 glomeruli sampled for light microscopy, 6 contained cellular crescents (figure 1A), ranging from circumferential to segmental. In the underlying glomerular tuft, there was variable mesangial and segmental endocapillary hypercellularity with infiltrating leucocytes (figure 1B). One glomerulus had segmental fibrinoid necrosis (figure 1C). Five glomeruli were globally sclerotic. Tubular atrophy and interstitial fibrosis occupied approximately 35% of the cortex. There were many red blood cell casts associated with acute tubular injury (figure 1D). Vessels displayed mild intimal sclerosis but no evidence of arteritis.

Among the seven glomeruli sampled for immunofluorescence, five were sclerosing. The remaining two showed dominant 2–3+ positivity for IgA in a granular global mesangial distribution (figure 1E), accompanied by 1+C3, 1+kappa and 2+lambda. There was no glomerular staining for IgG, IgM or C1q. Three glomeruli had 2+ segmental staining for fibrin/fibrinogen, suggesting necrotising lesions. Electron microscopy (three glomeruli sampled) revealed mesangial electron dense deposits and a single subepithelial deposit, accompanied by mild global mesangial hypercellularity. No subendothelial deposits were identified. There was segmental fibrinoid necrosis with glomerular basement membrane rupture, fibrin extravasation into the urinary space and cellular crescent formation.

The renal biopsy diagnosis was crescentic IgA nephropathy associated with perinuclear-ANCA/anti-MPO antibody seropositivity. Because the immune complex load appeared insufficient to account for the severity of the crescentic and necrotising features, the findings suggested IgA nephropathy and superimposed pauci-immune ANCA-associated crescentic glomerulonephritis.

TREATMENT

The patient was treated empirically with three doses of pulse intravenous methylprednisolone at 1000 mg intravenous solusets

Figure 1 (A) A cellular crescent (arrow) compresses the glomerular tuft (Jones methenamine silver (JMS), $\times 400$). (B) A glomerulus displays endocapillary and extracapillary (crescentic) proliferation. There is periglomerular interstitial inflammation (H&E, $\times 400$). (C) A glomerulus shows segmental fibrinoid necrosis with endocapillary fibrin, rupture of glomerular basement membrane and extravasation of fibrin into the urinary space. The adjacent glomerular segments have mild mesangial hypercellularity (JMS, $\times 600$). (D) Numerous red blood cell casts distend the tubular lumina, associated with acute tubular epithelial injury (H&E, $\times 200$). (E) A representative immunofluorescence micrograph shows a glomerulus with intense global mesangial staining for IgA, without staining of the peripheral glomerular capillary walls (immunofluorescence, $\times 600$).

per day, followed by oral prednisone 60 mg per day. Her renal biopsy was performed after the initial intravenous steroid treatment. When the renal biopsy results returned, the patient was given one dose of intravenous cyclophosphamide 700 mg (dose adjusted for eGFR of <15 mL/min) on day 12 of hospitalisation. Plasmapheresis was initiated 4 days after the initial cyclophosphamide dose for a total seven treatments administered every other day, using 4 units of fresh frozen plasma as the replacement fluid.

OUTCOME AND FOLLOW-UP

On hospital day 27, as the patient's renal function still had not recovered, requiring intermittent haemodialysis, she was discharged to follow-up at a dialysis clinic, while still receiving 60 mg of prednisone. In addition, she received her second dose of cyclophosphamide as an outpatient 1 month later. Eight days after her second dose of cyclophosphamide, the patient was readmitted to the medical intensive care unit for generalised malaise and fever with a right-sided vesicular rash in a dermatomal distribution on the trunk, consistent with herpes zoster, which was treated with 10 days of oral acyclovir. Her hospital stay was further complicated by the influenza A infection, bacteraemia secondary to the pansensitive *Rothia* species, and *Clostridium difficile*-negative diarrhoea. She was aggressively treated with antimicrobials given her immunocompromised state: she received dose-adjusted oseltamivir orally for 5 days, vancomycin intravenously for 10 days and empiric flagyl intravenously for 5 days. At last follow-up, the patient continues to require intermittent haemodialysis 7 months after renal biopsy.

DISCUSSION

We present a case of coexisting IgA nephropathy and pauci-immune ANCA-associated crescentic glomerulonephritis. Both entities are prevalent separately, but their coexistence is less common. In the literature, the prevalence of IgA nephropathy and coexisting ANCA-associated glomerulonephritis ranges from 0.2% to 2%.^{1–4} IgA nephropathy, on the other hand, is the most common primary glomerulonephritis throughout most developed countries of the world,⁵ with biopsy prevalence ranging from 3% to 16%.^{6,7} IgA nephropathy is characterised by dominant or codominant glomerular immune deposits of IgA, either alone or with lower-intensity staining for IgG and/or IgM in the glomerular mesangium.⁵ ANCA-associated glomerulonephritis typically manifests as 'pauci-immune' focal segmental necrotising and crescentic glomerulonephritis.⁸ The coexistence of both pathological findings in a biopsy can be considered a dual glomerulopathy. There are fewer than 20 case reports and

4 case series describing the association of IgA nephropathy and ANCA seropositivity.

Haas *et al* reported a series of six patients with severe crescentic glomerulonephritis with mesangial IgA deposits and positive ANCA serum titres.² Four patients had anti-PR3 ANCA, one had anti-MPO and one had both anti-PR3 and anti-MPO ANCA. Of these, five cases were from approximately 2250 total native renal biopsies, which demonstrated a 0.2% prevalence. Interestingly, the glomeruli in these cases tended to exhibit a mild, focal and segmental mesangial and endocapillary hypercellularity, as compared with patients with ANCA-negative IgA nephropathy who typically demonstrate greater mesangial and endocapillary proliferation. Five patients had available follow-up data, three of which had improved kidney function after treatment with cyclophosphamide and corticosteroids. The remaining two required dialysis at time of diagnosis and continued to require dialysis despite therapy.

Bantis *et al* described a case series of 8 patients from a total of 393 patients with IgA nephropathy diagnosed by renal biopsy (2.04% prevalence).¹ Of the eight patients, five had anti-MPO ANCA and three had anti-PR3 ANCA. Compared with their ANCA-negative counterparts, patients who had crescentic IgA nephropathy associated with ANCA seropositivity presented with a clinical syndrome of rapidly progressive glomerulonephritis with greater renal insufficiency. Despite similar intensity of mesangial IgA deposits, once again, mesangial hypercellularity was much greater in ANCA-negative as compared with ANCA-positive patients. All patients (8/8) with crescentic IgA and ANCA seropositivity received aggressive immunosuppressive therapy consisting of cyclophosphamide and corticosteroids, and all experienced subsequent improvement in renal function. In contrast, only a minority of the ANCA-negative group received aggressive therapy (5/26); the remainder received steroids alone, mycophenolate mofetil or angiotensin-converting enzyme inhibition. All of those patients experienced further progression of renal failure over 6 months of follow-up. The authors concluded that the less aggressive treatment offered to the ANCA-negative patients with IgA nephropathy may have been responsible for their less favourable outcome.

Huang *et al* described a case series of 14 Chinese patients with IgA nephropathy and ANCA seropositivity from among 787 biopsied cases of IgA nephropathy, comprising a 2% prevalence.³ Of the 14 patients identified (12 anti-MPO ANCA, 2 anti-PR3 ANCA), only 3 had crescentic glomerulonephritis. Each of the three patients with crescentic lesions demonstrated a different outcome after treatment with cyclophosphamide and steroids: one patient who was dialysis dependent at the time of biopsy remained on dialysis, another died of acute heart failure and the

third had improved renal function at 26 months after biopsy. Of the remaining 11 patients without crescents, 7 were treated with immunosuppressive and supportive therapy, 6 had stable or improved renal function and 1 progressed to ESRD, while 4 were lost to follow-up. The authors thus concluded that positive ANCA serology in the setting of IgA nephropathy is not always pathogenic and that biopsy findings including the prevalence of necrotising and crescentic features should guide the choice of therapy.

Comparing clinical consequences of patients with ANCA-positive IgA nephropathy with patients with ANCA-negative IgA nephropathy and patients with ANCA-associated systemic vasculitis (AASV), Yang *et al* found that the clinical and histopathological features of patients with ANCA-positive IgA nephropathy were comparable to those of patients with AASV and reflected a more severe presentation.⁴ Both groups tended to be older, had lower haemoglobin and higher inflammatory indicator levels at baseline and a higher percentage of constitutional symptoms and pulmonary involvement as compared with patients with ANCA-negative IgA nephropathy. Histologically, there was a significantly higher percentage of glomeruli with fibrinoid necrosis in these two groups as compared with patients with ANCA-negative IgA nephropathy. Contrary to their severe presentations, as in the previous study,¹ renal prognosis in patients with ANCA-positive crescentic IgA nephropathy was better after aggressive immunosuppressive therapy in the short term as compared with ANCA-negative patients: they were more likely to withdraw from dialysis (75% vs 9.1%, $p=0.03$) and not reach ESRD within 6 months (11.1% vs 66.7%, $p=0.01$). The authors suggested that screening for ANCA should be taken into consideration in certain patients with IgA nephropathy: the elderly, those with severe clinical or systemic manifestations and those with fibrinoid necrosis on renal biopsy.

A number of clinical and pathological features can influence prognosis in this setting. Chen *et al* showed in their retrospective study of 215 Chinese patients with ANCA-associated crescentic glomerulonephritis that a serum creatinine greater than 4 mg/dL and greater than 60% globally sclerotic glomeruli on biopsy were independent predictors of progression to ESRD.⁹ Our patient presented with a serum creatinine of 9.6 mg/dL and 38% globally sclerotic glomeruli. Furthermore, according to the Oxford/IgA Classification,^{10 11} our case demonstrated all four features associated with potential progression including diffuse mesangial proliferation (M1), focal endocapillary proliferation (E1), segmental sclerosis (S1) and significant tubular atrophy/interstitial fibrosis (T1). In addition, the presence of over 25% glomeruli with active crescents warranted a designation of (C2) according to new prognostic criteria.¹² Thus, although the literature suggests that patients with ANCA-positive crescentic IgA nephropathy may have a better renal outcome than their ANCA-negative counterparts, the combination of negative prognostic clinical and pathological features in our case likely contributed to the poor renal outcome.

The question remains whether the coexistence of ANCAs and IgA is a mere coincidence or is truly pathogenic. It is possible that patients have pre-existing IgA deposits in the mesangium and then develop necrotising and crescentic lesions following the appearance of ANCAs. This is inferred from the study that showed that 4.8% of autopsy cases harbour mesangial IgA deposits without a clear history of kidney disease.⁷ On the other hand, patients with crescentic IgA could have coincidental ANCA autoantibodies with no additive pathogenic potential. O'Donoghue *et al*, on screening 100 patients with IgA nephropathy, found 2 patients with ANCA positivity without crescents.¹³

Others, however, have found a distinctive clinical and histologic picture of IgA nephropathy and ANCA positivity, suggesting pathogenicity.¹² It is likely that these two scenarios are not mutually exclusive. In some patients with IgA nephropathy, the presence of positive ANCA serology drives the glomerulonephritis to a more necrotising and crescentic phenotype, whereas in others, this serological finding is non-pathogenic. Based on the limited number of cases reported, it is also unclear if the type of ANCA, that is, anti-MPO versus anti-PR3, confers any pathological or clinical distinction. Given that most cases with necroses and crescents present with rapidly deteriorating renal function, often requiring dialysis, these patients are usually offered aggressive immunosuppressive therapy. Lee *et al* demonstrated that even for patients with severe ANCA-associated glomerulonephritis with an initial eGFR <15, over 50% of patients still responded to immunosuppressive therapy and were dialysis free at the end of 4 months.¹⁴

Unfortunately, most immunosuppressive regimens carry life-threatening risks including infection-related complications, especially in the elderly population. In ANCA-associated vasculitis, older age in itself is independently associated with mortality,^{15 16} with 36%–80% of deaths attributed to infections.^{17–19} A study done by Haris *et al*²⁰ showed that treating elderly patients (age >65) with 'reduced-dose' (ie, approximately 20% lower dose) immunosuppressives versus the standard dose given in younger patients (age <65) showed no difference in vasculitis outcomes. There was also no significant difference in severe, non-lethal infections, requiring hospitalisations between the elderly and younger patients (39% vs 50%, respectively; $p=0.29$). There was, however, an increased mortality in the elderly group ($p=0.007$), which was attributed primarily to cardiovascular causes.

Our patient was dialysis dependent at the time of biopsy and had received aggressive treatment with standard dose-adjusted cyclophosphamide and pulse intravenous steroids followed by daily oral steroids. During the follow-up period, the patient continued to develop multiple infectious complications requiring intensive care. The patient has since recovered from all infections, but remains dialysis dependent 7 months postdiagnosis. Her disease course highlights that, in selected elderly patients requiring immunosuppressive therapy, a reduced-dose immunosuppressive regimen is preferable, as it can provide similar efficacy with reduced risk of infection in this susceptible population.

Patient's perspective

► It is unfortunate that I developed this unusual disease, but I am grateful for the care that I am getting. My ongoing dialysis has made me feel better, and I am continuing to improve both physically and emotionally.

Contributors PN researched and prepared the manuscript. VD'A contributed to the pathology assessment and writing of the manuscript. KA took care of the patient and helped to design the manuscript. BJ took care of the patient and helped to design and write the manuscript.

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Patient consent Obtained.

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Learning points

- The prevalence of IgA nephropathy and coexisting antineutrophilic cytoplasmic antibody (ANCA)-associated crescentic glomerulonephritis ranges from 0.2% to 2%.
- The combination of IgA nephropathy and ANCA seropositivity commonly presents with more severe clinical and histological picture but may also be more responsive to treatment when compared with ANCA-negative crescentic IgA nephropathy. However, this prognosis must take into consideration traditional markers for progression such as the Oxford/IgA classification features association with progression. Advanced age should be taken into account when choosing the optimal immunosuppressive therapy for ANCA vasculitis. Reduced-dose immunosuppressive regimens have been shown to have similar efficacy and tolerable treatment-related complications.

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