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Case Report
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Anti-Neutrophil Cytoplasmic Antibody (ANCA) - Negative Small Vessel Vasculitis: A Rare Case Study of Hydronephrosis and Pulmonary Infiltrates

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Abstract

Hydronephrosis and pulmonary infiltrates are rare manifestations of ANCA - negative small vessel vasculitis. We report the case of a 66-year-old woman with progressive renal failure in which unilateral hydronephrosis and pulmonary infiltrates were the first manifestation of the disease. Tissue biopsy of kidney confirms the diagnosis of ANCA-negative pauci-immune Crescentic Glomerulonephritis (CrGN). Repeated ANCA tests were always negative, and other immune markers were normal. The hydronephrosis and pulmonary infiltrates rapidly improved under immunosuppressive therapy with high-dose corticosteroids and mycophenolate mofetil oral QD. The signs and symptoms of ANCA negative vasculitis are varied, which can lead to misdiagnosis. In clinical practice, patients with progressive renal failure and hydronephrosis but ANCA negative need to be screened for vessel vasculitis also.

Keywords: ANCA-negative vasculitis; ANCA-negative pauci-immune CrGN; Hydronephrosis; Pulmonary infiltrates

Introduction

ANCA-Associated Vasculitis (AAV) is a group of diseases characterized by inflammation and destruction of small-and medium-sized blood vessels and the presence of circulating ANCA [1]. ANCA is pathogenic significance in AAV, which is involved in the occurrence, development and prognosis of the disease [2]. In most patients, pauci-immune CrGN is a manifestation of ANCA - associated vasculitis [3]. However, a subgroup of patients (10% to 30%) with pauci-immune CrGN was short of Anti-Neutrophil Cytoplasmic Antibodies (ANCAs) [3]. Limited information on ANCA-negative pauci-immune CrGN is existed. Patients with ANCA-negative had fewer respiratory symptoms than those with ANCA positive [4]. Chen et al. [3] found that patients who were ANCA negative had fewer systemic manifestation and less extra renal involvement than patients who were ANCA positive [3].

Here, we report a case of ANCA-negative pauci-immune CrGN presented with right kidney hydronephrosis and pulmonary infiltrates.

Case Presentation

A 66-year-old female with unilateral hydronephrosis, cough, occasionally hemoptysis, renal insufficiency of unclear etiology was admitted at our institution. Patient experience rapid loss of renal function, accompanied by proteinuria, dysmorphic hematuria (40% to 50% at the beginning) and hydronephrosis. Chest CT studies of our hospital showed large dense shadows in the middle and lower lobes of the right lung. Nodular density shadows are scattered in both lungs accompanying pleural effusion (Figure 1A). Urinary ultrasound showed extent of hydronephrosis about 11 mm (Figure 2A). Antinuclear Antibody (ANA) was <1:20, and the remainder of serum immune index was essentially normal (Table 1). The laboratory test was suggestive of an acute-on-chronic renal failure, hemodialysis had to be initiated for worsening renal failure, and a renal biopsy was executed. Renal histopathological picture of our patient (Figure 3) was performed showing cellular crescent involved little or no glomerular immunoglobulin deposition.

Immediately, she received the treatment of Intravenous Methylprednisolone Pulse (IVMP) 500 mg/d for 3 days, followed by methylprednisolone 40 mg Intravenous injection (IV) QD. She received

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Table 1: Summary of the pertinent laboratory values during first admission.

Lab	Results	Lab	Results	Lab	Results
WBC	9.71 × 10^9/L	D-Dimer	0.50 mg/L FEU	Scr	440 μmol/L
Hgb	83 g/L	GBM Ab	Negative	GFR	8.6 ml/min
Hct	24.20%	SS-A/Ro	Negative	TSH	2.6005 mIU/L
PLT	291 × 10^9/L	SS-B/La	Negative	PCT	0.11 ng/ml
HS-CRP	42.53 mg/L	ACA	Negative	Alb	24.5 g/L
ESR	122 mm/H	24 h PRO	1.64 g	BUN	12.59 mmol/L
ANA	<1:20	UA	417 μmol/L	HAV-IgM	Negative
RF	<20 IU/ml	p-ANCA	Negative	HBsAg	Negative
Anti-CCP	Negative	c-ANCA	Negative	HCV-IgG	Negative
Anti-ds-DNA	Negative	IgG	1270 mg/dl	HIV (1/2)	Negative
Anti-Smith Ab	Negative	IgM	81 mg/dl	C3	105 mg/dl
Scl-70	Negative	IgG4	252 mg/dl	C4	25 mg/dl

WBC: White Blood Cell; Hgb: Hemoglobin; Hct: Hematocrit; PLT: Platelets; HS-CRP: High-Sensitivity C-Reactive Protein; ESR: Erythrocyte Sedimentation Rate; ANA: Anti-Neutrophil Antibody; RF: Rheumatoid Factor; CCP: Cyclic Citrullinated Peptide; ds-DNA: Double-Stranded DNA; Scl: Scleroderma; GBM: Glomerular Basement Membrane; Ab: Antibody; SS-A: Sjögren-Syndrome-related-antigen A; SS-B: Sjögren-Syndrome-related-antigen B; ACA: Anti Cardiolipin Ab; 24 h PRO: 24 h-Urine Protein; UA: Uric Acid; ANCA: Anti-Neutrophil Cytoplasmic Antibody; Ig: Immunoglobulin; Scr: Serum Creatinine; GFR: Glomerular Filtration Rate; TSH: Thyroid Stimulating Hormone; PCT: Procalcitonin; Alb: Albumin; BUN: Blood Urea Nitrogen; HAV: Hepatitis A Virus; HBsAg: Hepatitis B Surface Antigen; HCV: Hepatitis C Virus; HIV: Human Immunodeficiency Virus; C3: Complement 3; C4: Complement 4

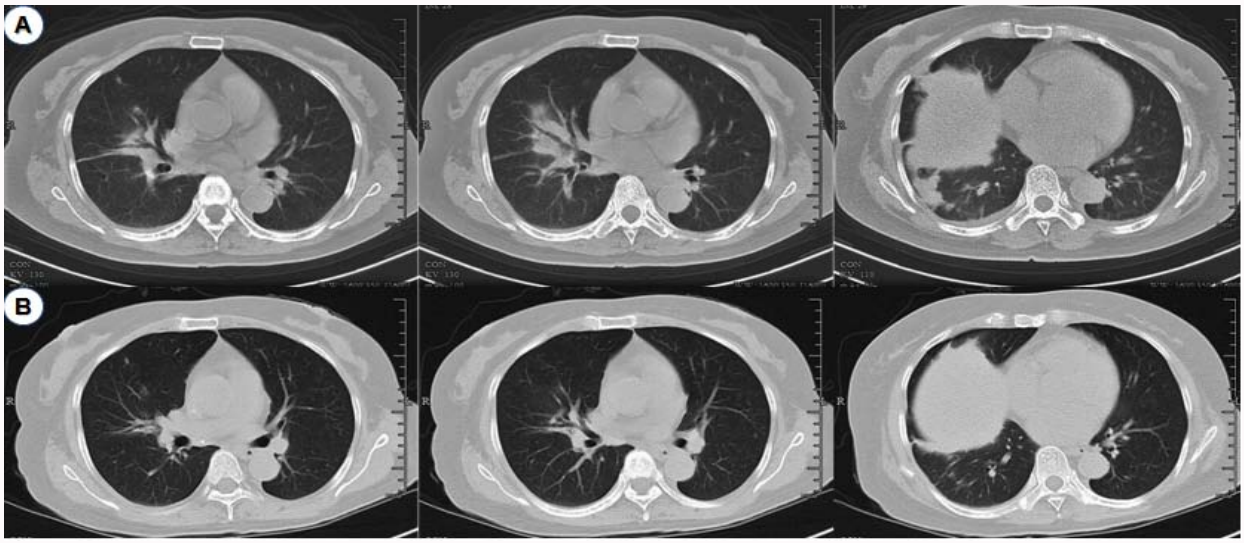

Figure 1: A computed tomography scan of the chest. (A) Hyper-density masses in the middle and inferior lobes of the right lung and scattered nodular soft tissue density shadow on both sides. (B) The multiple lesions were significantly improved compared with the film on admission.

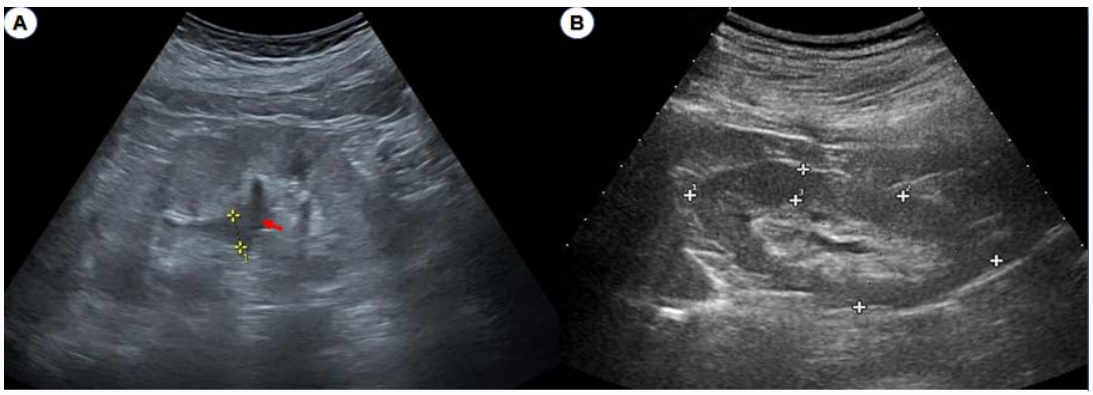

Figure 2: Ultrasonography has an important role in evaluation of hydronephrosis. (A) Hydronephrosis in right kidney before treatment. (B) Hydronephrosis is relieved on the 40th day.

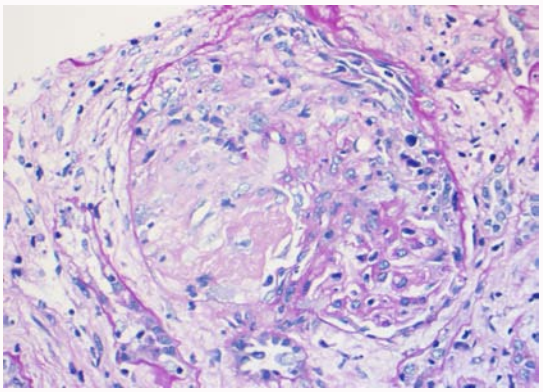

Figure 3: The renal biopsy showed 25 glomeruli, 13 of the glomeruli show presence of cellular crescent, the others with an ischemic appearance. Glomerular lesions included proliferation of mesangial matrix and mesangial cell, with segmental endothelial cell pairing. Renal interstitial fibrosis with mixed inflammatory cell infiltration.

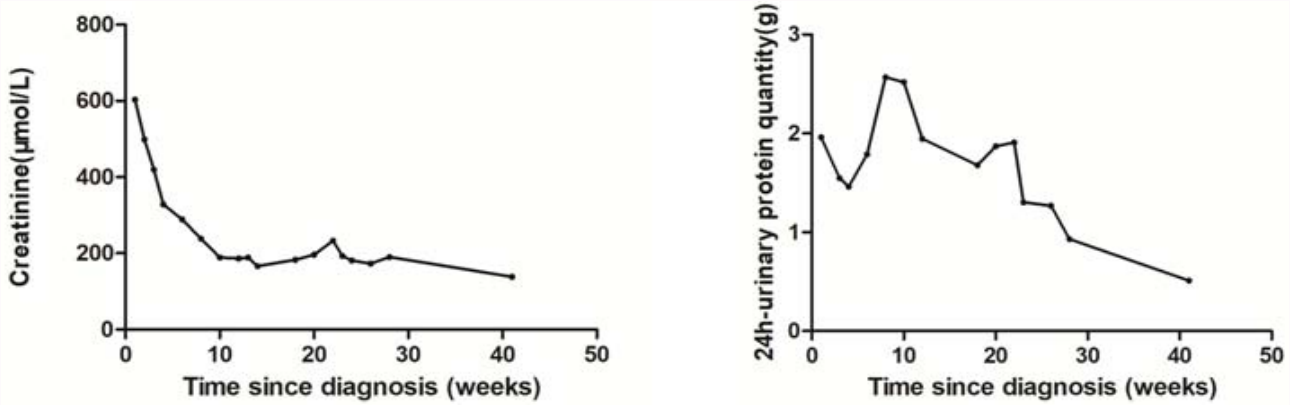

Figure 4: Creatinine and 24 h urinary protein quantity throughout the follow-up period as indicated.

the treatment of IVMP 500 mg/d for 3 days again after 2 weeks, followed by a slowly reduced of oral glucocorticoids from 40 mg/d maintain. Ultrasonography rechecked shows extent of hydronephrosis about 7 mm on the 40th day of treatment (Figure2B). A chest computed tomography showed high density shadows of the lung parenchyma was significantly improved compared with the previous film (Figure 1B). Prednisolone cure was initiated with favorable response, but given the severity of the biopsy findings, cyclophosphamide was added (0.4 g for each time). However, gastrointestinal reaction was obvious with cyclophosphamide, so mycophenolate mofetil 0.75 g/d was added to replace cyclophosphamide. Methylprednisolone was changed to oral prednisolone on the morning of discharge.

Patient adhere to regular follow-up in outpatient department after discharge, a continuous systemic function improvement was evident (Figure 4).

Discussion

The general extra-renal manifestations of ANCA-negative vasculitis presenting fatigue, night sweats, weight loss, fever, myalgia and arthralgia [5]. Pulmonary infiltrates are rare in ANCA-negative crescentic GN as extra-renal involvement [6]. Due to the long time from the onset of the disease to the treatment, long-term pulmonary vasculitis can cause pleural effusion, which is also consistent with this case.

We reviewed the literature and found only two ANCA-negative cases reported by Kamar et al. [7] had unilateral hydronephrosis with granulomatous inflammation as the sole manifestation of the disease [7]. These patients have acute or chronic kidney disease, most of which are improved by glucocorticoid or immunosuppressive therapy, and some patients need surgery to remove the obstruction. As far as we know, we are reporting on the first patient with unilateral hydronephrosis as the presenting manifestation of ANCA-negative vasculitis.

There is study demonstrates the ANCAs play an important role in small-vessel vasculitis. In ANCA-negative pauci-immune CrGN, cell-mediated immunity may play an important pathogenic role [8], and the good clinical efficacy was observed by used high dose steroids and MMF. This report provided clinical experience and improved the understanding of multiple organ injury in ANCA negative primary small vasculitis. The immunity index of ANCA-negative makes the diagnosis difficult and kidney biopsy was needed of our patient to make a correct diagnosis. Most importantly, it is necessary to integrate clinical examination, pathological findings, serologic markers to avoid misdiagnosis and misjudgment of the condition.

For ANCA negative vasculitis with multiple organ injury, it is necessary to conduct further research to explore different treatment methods and improve the prognosis of patients.

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