


An Autopsy Case of Renal-Limited Granulomatosis With Polyangiitis Presenting With Acute Renal Failure and Initial Delirium

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Abstract

Granulomatosis with polyangiitis (GPA) has three clinicopathological features, namely, necrotizing granulomatosis of the upper respiratory tract and lungs, focal segmental necrotizing glomerulonephritis of the kidney, and necrotizing vasculitis of small vessels throughout the body. A 92-year-old man with clinically diagnosed probable Alzheimer's disease (AD) exhibited subacute deterioration in cognitive function. On admission, he was diagnosed with acute renal failure with an elevated creatinine level (5.48 mg/dL) as well as severe disturbance of consciousness. Antineutrophil cytoplasmic antibodies (ANCA) directed against proteinase 3 (PR3-ANCA) were highly positive with ≥ 350 U/mL. The patient was diagnosed with GPA and was managed with steroid pulse therapy. However, he died without any improvement in renal function. As a result of the autopsy, the patient was diagnosed with definite AD, and his impaired consciousness was found not to be caused by central nervous system involvement due to GPA. As necrotizing crescentic glomerulonephritis was observed, the cause of the acute progressive renal failure was found to be PR3-ANCA-positive GPA. The autopsy revealed no GPA-related lesions in other parts of the body aside from the kidneys. It is rare to encounter cases of PR3-ANCA-positive GPA with renal-limited vasculitis and acute renal failure as the initial manifestation, as in the present case. Making an accurate clinical diagnosis of older patients suffering from various diseases in multiple organs is challenging. Although autopsy has the limitation of a terminal image, it is extremely useful in elucidating the pathophysiology of the older patient in this case.

Keywords: Alzheimer's disease; Delirium; Granulomatosis with polyangiitis; Renal-limited; PR3-ANCA

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Introduction

Granulomatosis with polyangiitis (GPA) is a type of vasculitis syndrome in which inflammation occurs in the walls of small vessels. It is classified as an antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis. This disease was first reported by the German pathologist Friedrich Wegener in 1938, hence the former name Wegener's granulomatosis. However, at the 2012 Chapel Hill Consensus Conference, the disease was renamed GPA due to the new international classification criteria based on pathogenesis [1]. GPA is a refractory vasculitis with three clinicopathological features: 1) necrotizing granulomatosis of the upper respiratory tract (nose, pharynx, and sinuses) and lungs, 2) focal segmental necrotizing glomerulonephritis of the kidney, and 3) necrotizing vasculitis of small vessels throughout the body. ANCA directed against proteinase 3 (PR3-ANCA) is a disease-labeling antibody for ANCA-associated vasculitis. PR3-ANCA is present in the majority of patients with GPA and appears to be extremely specific [2, 3]. The titer of PR3-ANCA easily parallels GPA severity and is a predictor of relapse and disease severity [2]. The involvement of the central nervous system (CNS) due to systemic secondary CNS vasculitis, such as cognitive impairment, consciousness disturbance, headache, and visual disturbance, may not be a major symptom of ANCA-associated vasculitis [4, 5]. However, the frequency of CNS involvement in all cases of ANCA-associated vasculitis is reportedly 11 times higher than that in the general population [6].

We have recently experienced an autopsy case of an older patient with Alzheimer's disease (AD) who initially presented with cognitive impairment and later developed rapidly progressive glomerulonephritis with highly elevated PR3-ANCA. Thus, we herein report the clinical course and autopsy results of this patient.

Case Report

Approximately 2 months before admission, a 92-year-old man with a history of chronic heart failure and hypertension presented to our hospital due to a gradual progressive deterioration of cognitive dysfunction, such as recent memory impairment and disorientation. There was no history of smoking or

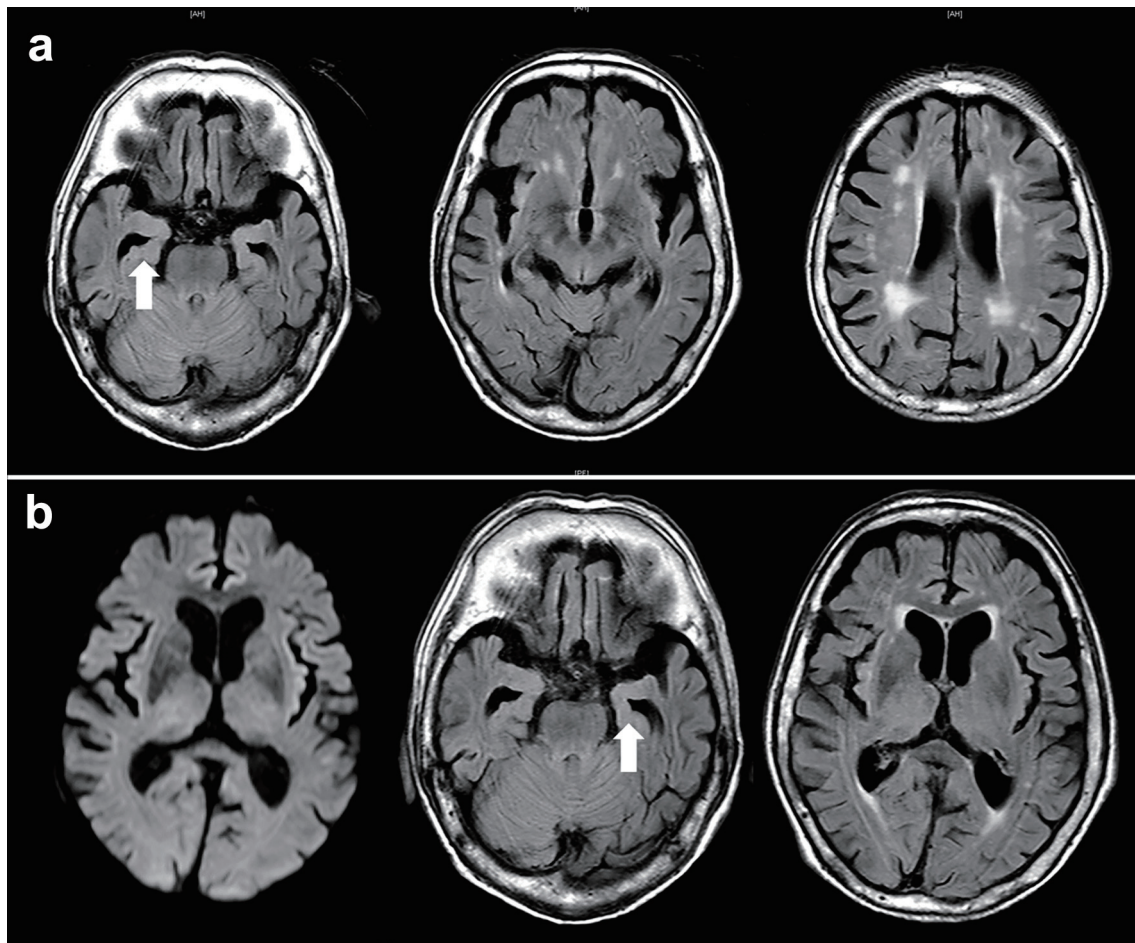


Figure 1. Axial view of brain magnetic resonance imaging (MRI). (a) Approximately 2 months before admission, fluid-attenuated inversion recovery images show atrophy of the brain cortex with significant hippocampal atrophy in the medial temporal lobe, indicating Alzheimer's disease. Mild periventricular white matter lesions are observed. (b) MRI image of the brain on admission. Diffusion-weighted imaging shows no apparent abnormalities. FLAIR images also show findings similar to the previous ones. No lesions in the brain are observed that would cause impaired consciousness. The arrows indicate hippocampal atrophy.

alcohol consumption, and no notable family history of hereditary diseases. Comprehensive neuropsychological tests were conducted to assess his cognitive function. A decline of 17/30 points was observed in the mini-mental state examination. The clinical dementia rating was 2.0, and the functional assessment staging of AD was stage 5. On the fluid-attenuated inversion recovery (FLAIR) images of brain magnetic resonance imaging (MRI), significant brain atrophy was detected predominantly in the hippocampus and temporal lobes, suggesting AD (Fig. 1a). The patient's blood thyroid hormone and vitamin B levels were within the normal range. He was diagnosed with probable AD, meeting the clinical diagnostic criteria, and denying any organic disease that would accompany other dementias. His laboratory results are shown in Table 1. Blood examination at this time revealed a serum creatinine (Cr) level of 1.34 mg/dL, suggesting mild renal dysfunction, but hematuria was not observed.

Approximately 2 weeks before admission, he began showing symptoms consistent with delirium, including incoherence, hallucination, and disorientation. These symptoms also

seemed to indicate behavioral and psychological symptoms of dementia (BPSD). The patient stopped eating. Eventually, he developed a worsening consciousness disorder and was urgently admitted to our hospital. His level of consciousness was E1V1M2 on the Glasgow coma scale, indicating severe consciousness impairment. His initial vital signs were as follows: heart rate, 63 beats/min; respiratory rate, 24 breaths/min; blood pressure, 156/77 mm Hg; body temperature, 36.7 °C; and oxygen saturation, 97% with room air. He had a height of 147.3 cm, a body weight of 45.6 kg, and a body mass index of 21.0 kg/m². Furthermore, gross hematuria and anemia in the palpebral conjunctiva were observed. Diffusion-weighted and FLAIR images of MRI depicted no apparent abnormalities (Fig. 1b). Blood counts showed a decrease in hemoglobin to 6.8 g/dL, possibly due to hematuria. Renal dysfunction was detected with elevated levels of serum Cr (5.48 mg/dL) and blood urea nitrogen (BUN, 73.8 mg/dL) (Table 1). In electrolyte concentrations, serum potassium was elevated at 5.5 mmol/L. Based on the presence of renal dysfunction and heart failure (N-terminal pro-B-type natriuretic peptide 1,052 pg/

Table 1. Laboratory Data

Variable (normal range)	Results		
	2 months before admission	On admission	1 week after admission
Blood examination			
WBC ($3.3 - 8.6 \times 10^3/\mu\text{L}$)	$7.3 \times 10^3/\mu\text{L}$	$5.9 \times 10^3/\mu\text{L}$	$3.5 \times 10^3/\mu\text{L}$
Hemoglobin (11.2 - 15.2 g/dL)	9.6 g/dL	6.8 g/dL	5.1 g/dL
Platelets ($158 - 348 \times 10^3/\mu\text{L}$)	$100 \times 10^3/\mu\text{L}$	$52 \times 10^3/\mu\text{L}$	$23 \times 10^3/\mu\text{L}$
Albumin (4.1 - 5.1 g/dL)	2.7 g/dL	2.1 g/dL	1.7 g/dL
CPK (59 - 248 U/L)		78 U/L	284 U/L
BUN (8 - 20 mg/dL)	24.2 mg/dL	73.8 mg/dL	126.0 mg/dL
Cr (0.65 - 1.70 mg/dL)	1.34 mg/dL	5.48 mg/dL	9.82 mg/dL
Na (138 - 145 mmol/L)	135 mmol/L	137 mmol/L	147 mmol/L
K (3.6 - 4.8 mmol/L)	4.0 mmol/L	5.5 mmol/L	4.8 mmol/L
Cl (101 - 108 mmol/L)	106 mmol/L	103 mmol/L	111 mmol/L
Ca (8.8 - 10.1 mg/dL)		7.8 mg/dL	7.9 mg/dL
Glucose (73 - 109 mg/dL)	138 mg/dL	103 mg/dL	161 mg/dL
CRP (< 0.14 mg/dL)	0.39 mg/dL	5.89 mg/dL	7.55 mg/dL
Troponin T (< 14.0 pg/dL)		5.89 pg/dL	161.0 pg/mL
NT-proBNP (< 125.0 pg/dL)	1,329 pg/mL	1,052 pg/mL	46,835 pg/mL
Hepatitis B surface antigen		Negative	
Hepatitis C virus antibody		Negative	
Blood CMV pp65 antigenemia		Negative	
Antinuclear antibody titers (< 1:40)		1:40 (cytoplasmic)	
Anti-PR3 antibody (ELISA) (< 3.5 U/mL)			≥ 350 U/mL
Anti-MPO antibody (ELISA) (< 3.5 U/mL)			< 0.1 U/mL
Urine examination			
Clarity	Straw	Red	
pH (4.8 - 7.5)	6.0	8.0	
Urine specific gravity (1.008 - 1.034)	1.020	1.009	
Protein (-)	-	2+	
WBC (1 - 4/HPF)	1 - 4/HPF	1 - 4/HPF	
RBC (5 - 9/HPF)	5 - 9/HPF	≥ 100 /HPF	
Squamous epithelium (< 1/HPF)	1 - 4/HPF	1 - 4/HPF	
Hyaline casts (< 1/WF)		1/WF	

BUN: blood urea nitrogen; CMV: cytomegalovirus; CPK: creatine phosphokinase; Cr: creatinine; CRP: C-reactive protein; ELISA: enzyme-linked immunosorbent assay; HPF: high power field; MPO: myeloperoxidase; NT-proBNP: N-terminal pro-B-type natriuretic peptide; RBC: red blood cell; WBC: white blood cell; WF: whole field.

mL) detected through laboratory investigations on admission, a thoracic abdominal computed tomography (CT) scan was performed (Fig. 2). A chest CT scan revealed no sign of lesions in the lung, but an enlarged left atrium and congestion were found in the heart (Fig. 2a, b). On abdominal CT scan, no obvious abnormal lesions in the kidneys or urinary system organs causing postrenal failure were observed (Fig. 2c, d). Initially, the patient was managed with fluid therapy, considering the possibility of prerenal renal failure due to dehydration. However, his condition did not improve. On laboratory inves-

tigations 1 week after admission, serological tests revealed elevated PR3-ANCA levels (≥ 350 U/mL; normal, < 3.5 U/mL) with negative myeloperoxidase (MPO)-ANCA (Table 1). Based on these findings, the patient was diagnosed with probable GPA. However, his renal function acutely deteriorated within 1 week, with Cr of 9.82 mg/dL and BUN of 126.0 mg/dL (Table 1). Subsequently, the treatment was commenced for possible GPA with a high-dose intravenous methylprednisolone (1,000 mg/day) for 3 days as steroid pulse therapy. As aforementioned, the patient was extremely old and suffering

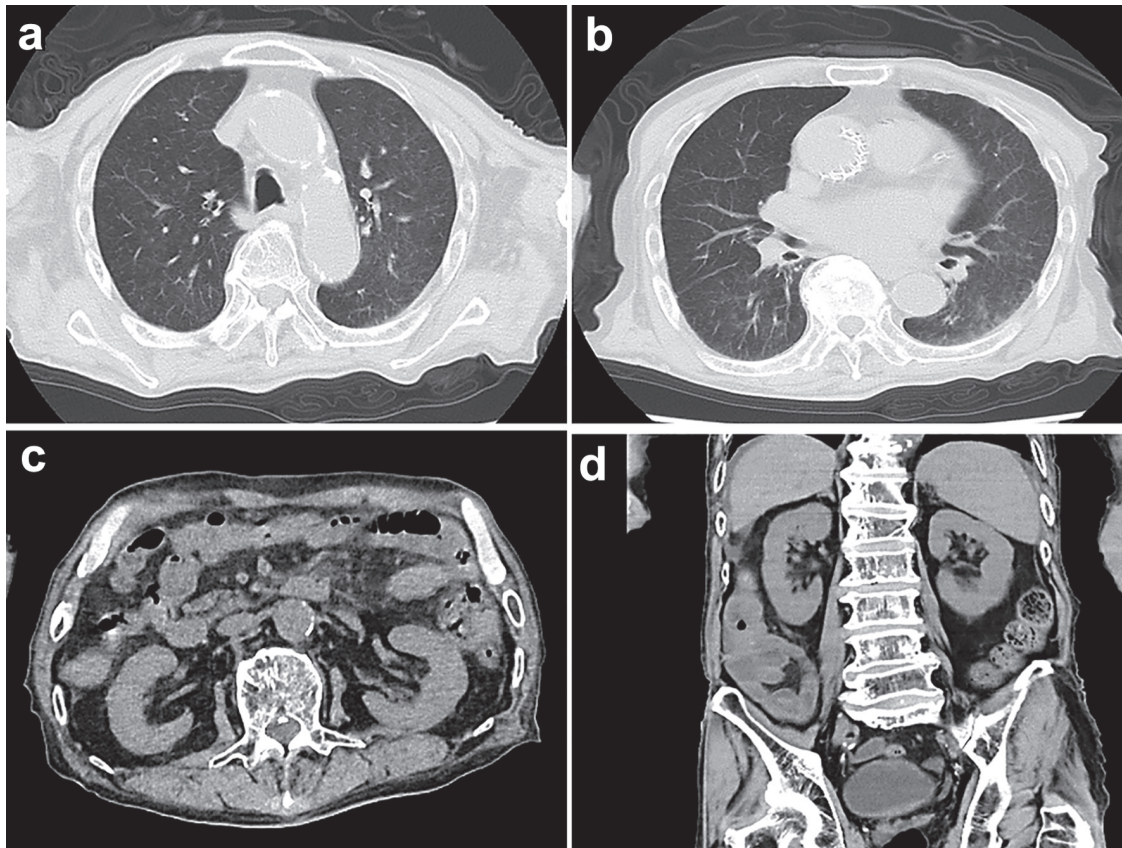


Figure 2. Computed tomography (CT) scan images of the thorax and abdomen on admission. (a, b) Chest CT scan shows no signs of lesion in both lungs, but an enlarged left atrium and congestion in the heart are observed. (c, d) CT scan of the abdomen shows no obvious abnormal findings such as mass lesions or ascites.

from moderate AD. Thus, hemodialysis therapy could not be performed. The family also did not wish for further treatment and decided to follow up with the best support care. Approximately 10 days after admission, the patient became anuric, went into cardiac arrest, and died. The timeline of this case is shown in Figure 3.

Autopsy findings

The questionable points in the clinical course of this case are as follows. 1) Was the patient's cognitive impairment caused by AD? 2) Did the CNS involvement by GPA affect consciousness disturbance and decline in cognitive function? 3) Was GPA the cause of the acute renal failure with strong PR3-ANCA positivity? 4) Assuming that it is GPA, are there histological findings in other organs aside from the kidneys? Considering the aforementioned points, an autopsy was performed with the consent of the family.

Brain

Upon gross autopsy examination, the fresh brain weighed 1,015 g. Mild atrophy was observed in the temporal lobe. His-

tologically, senile plaques were found in the parietal lobe and hippocampus; neurofibrillary tangles in the putamen, parietal lobe, and hippocampus; and neuropil threads in the frontal lobe (Fig. 4). These findings are consistent with the histopathological features of AD. There was no direct expansion of granulomatous processes, vasculitis of cerebral small vessels, and direct formation of granulomas within the brain parenchyma, suggesting GPA.

Kidney

Both kidneys were turbid and swollen. Approximately 15% of the glomeruli had crescents (cellular and fibrous) (Fig. 5). A moderate inflammatory cell infiltrate, mainly lymphocytes, was found in the submucosa of the renal pelvis and in the renal interstitium. These histopathological findings supported GPA.

Lung

Both lungs were fibrinously adherent to the chest wall. Grossly, predominant lower lobe congestion was observed, and a hemosiderin-laden macrophage was prominent in the left lung. There were no findings indicating GPA, such as granuloma-

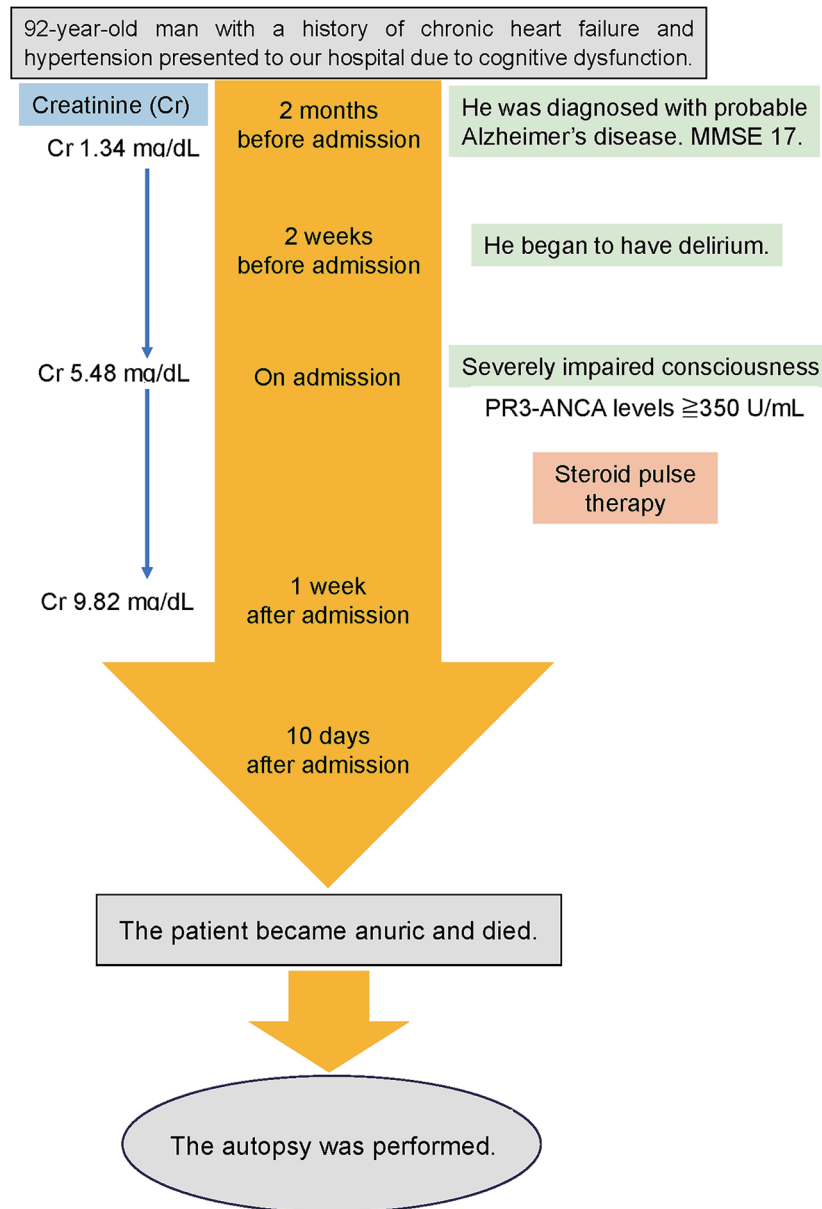


Figure 3. Timeline of this case report. MMSE: mini-mental state examination.

tous inflammation, vasculitis, or mixed cellular infiltration of neutrophils, lymphocytes, or plasma cells.

Heart

The patient had a mild image of acute-stage infarction showing fresh myocardial necrosis in the left ventricular anterior wall and septum.

Others

There were no histological findings indicating granulomatous

lesions or vasculitis of small-sized vessels in other organs, including the upper respiratory tract.

Discussion

In this case, we confirmed the absence of systemic disease or other brain disease that could cause progressive cognitive impairment. According to the diagnostic criteria of the National Institute of Neurological and Communicative Disorders and Stroke AD and Related Disorders Association (NINCDS-ADRDA), a clinical diagnosis of probable AD was made [7]. Based on the NINCDS-ADRDA, the definitive diagnosis of AD was based on neuropathological evidence from a biopsy or au-

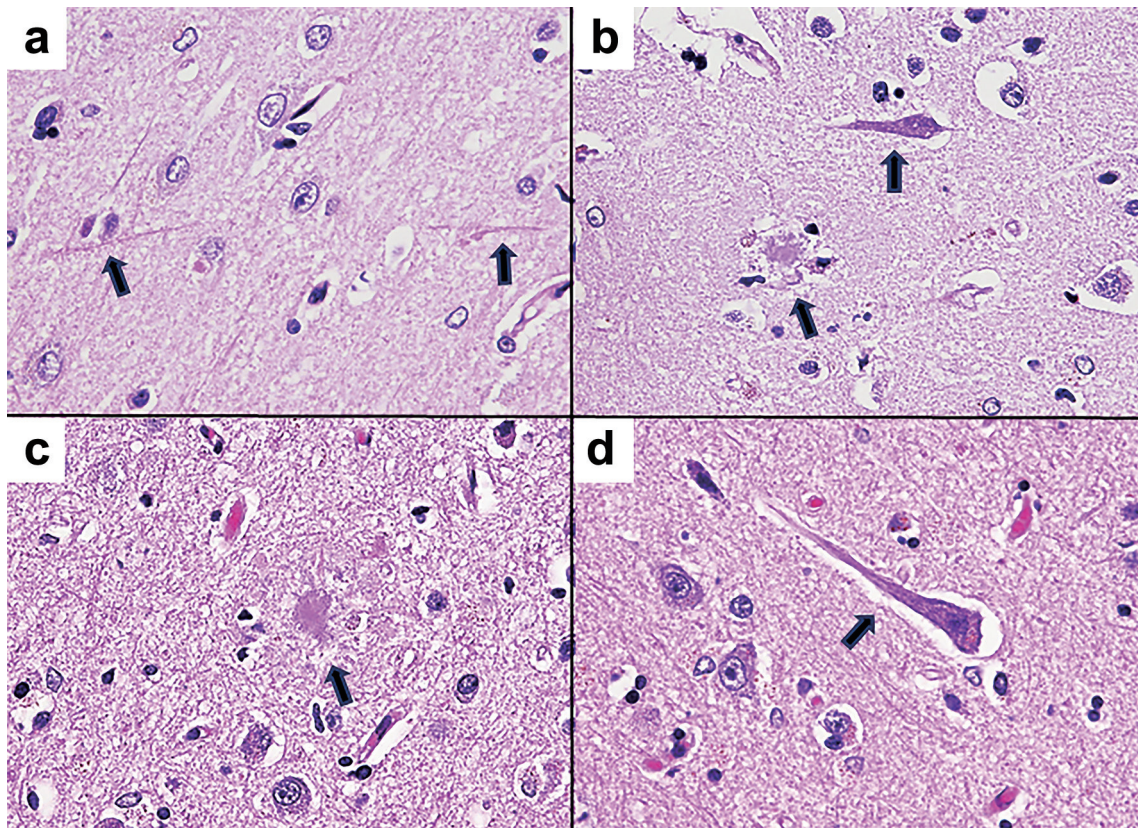


Figure 4. Pathological microscopic images of the brain tissue (hematoxylin and eosin staining, $\times 400$). (a) Neuropil threads are observed in the cortical tissue of the frontal lobe. The arrows indicate neuropil threads. (b) In the cortical tissue of the hippocampus, senile plaques and neurofibrillary tangles are observed. The senile plaque appears to be a neuritic plaque with surrounding neurites. The arrows indicate senile plaque and neurofibrillary tangle. (c) Senile plaque of the cortical tissue of the parietal lobe. The arrow indicates senile plaque. (d) Neurofibrillary tangle of the cortical tissue in the parietal lobe. The arrow indicates neurofibrillary tangle.

topsy. Thus, in actual clinical practice, extremely few patients are definitively diagnosed with AD. The autopsy revealed neuropathological findings that suggest AD (Fig. 4). Thus, the patient was diagnosed with definite AD. The coexistence of moderate AD may have influenced the early detection and diagnosis of acute renal failure. He exhibited subacute deterioration in cognitive function such as the appearance of BPSD or delirium. The autopsy revealed that the aforementioned neurological findings were not related to CNS involvement by GPA. Therefore, the deterioration of the patient's general condition due to dehydration and uremia due to acute renal failure was considered to have caused delirium and eventually severe consciousness disturbance. CNS involvement in GPA is more common in the severe form and reportedly occurs in about 10% of patients [6, 8, 9]. However, in the present case, although PR3-ANCA was abnormally high and GPA was severe, the patient's neurological symptoms were not associated with GPA.

The patient had a strongly positive PR3-ANCA and acute progressive renal failure, but histopathological findings were necessary to make a definitive diagnosis of GPA [10]. At autopsy, necrotizing crescentic glomerulonephritis and peritubular inflammatory cell infiltration were observed in the kidney,

which led to a definitive diagnosis of GPA (Fig. 5). This case also met the criteria for GPA according to the 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology, which requires a total score ≥ 5 [11]. GPA begins in the upper respiratory tract and often progresses to the lower respiratory tract (lung) and kidney. The presence of all three lesions indicates a systemic type, whereas the presence of only one or two lesions indicates a limited type. More than 90% of patients with GPA have been reported to have upper airway involvement [12]. Lung lesions are reported to be present in almost 50% of patients with GPA initially, and lung disease eventually develops in 85-90% of patients in the course of the disease [13]. However, in the autopsy of this case, there were no pathological findings indicating GPA in other parts of the body (e.g., skin, eye, ear, or nervous system), including these major lesions, aside from the kidney. To the best of our knowledge, there are a few studies on PR3-ANCA-positive GPA presenting as a solitary renal mass accompanying glomerulonephritis like this case, although there was no renal mass in our patient [14-16]. Thus, the case of PR3-ANCA-positive GPA with renal-limited vasculitis and acute renal failure as the initial manifestation is rare, as in the present case. Furthermore, renal involvement is clinically evident in only 11%

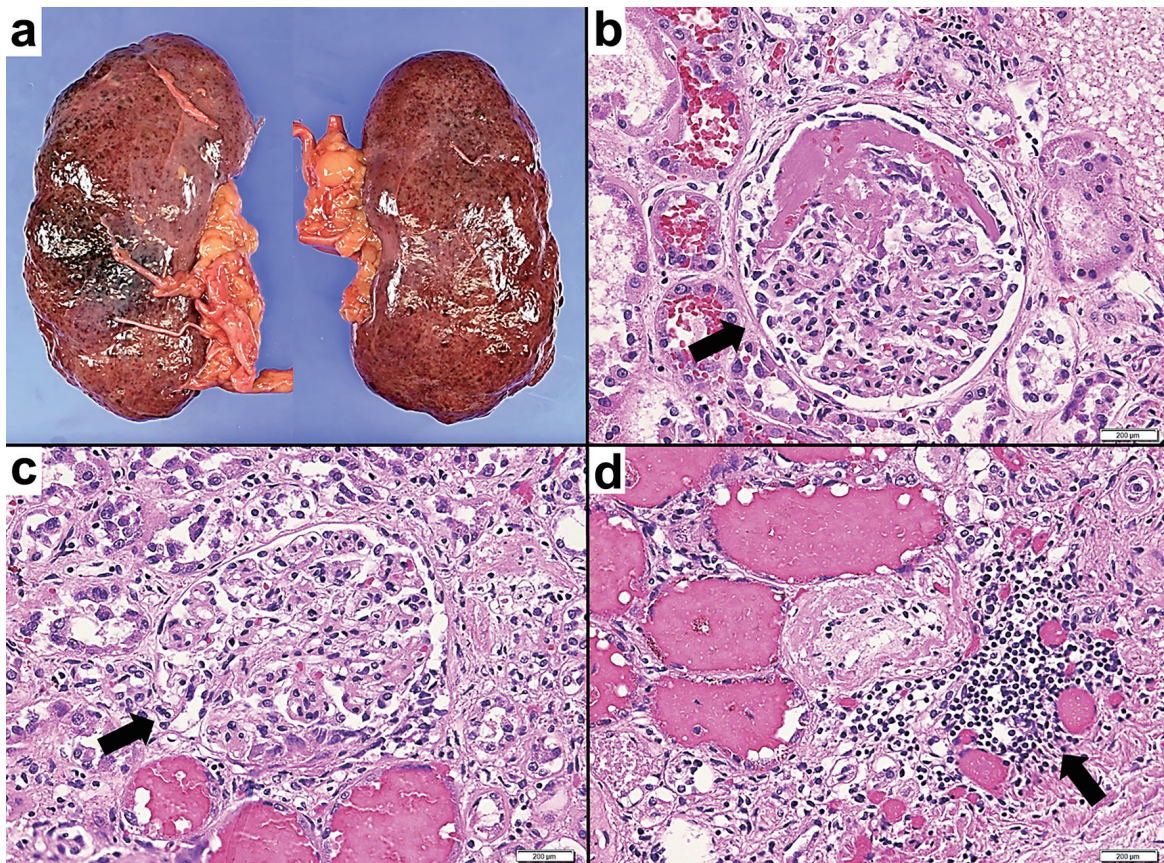


Figure 5. Pathological macroscopic and microscopic images of the kidney tissue. (a) The fresh both kidneys exhibit turbid coloration and swelling. (b, c) Hematoxylin and eosin staining, $\times 400$. (b) Glomerular lesions exhibiting fibrous crescent formation are observed. The arrow indicates fibrous crescentic glomerulonephritis. (c) Glomerular lesions exhibiting cellular crescent formation are observed. Inflammatory cells including neutrophils accumulate in areas demonstrating fibrin precipitation and loop rupture. The arrow indicates cellular crescentic glomerulonephritis. (d) Hematoxylin and eosin staining, $\times 200$. Inflammatory cells such as lymphocytes and neutrophils accumulate in the tubulointerstitium. The arrow indicates peritubular inflammatory cell infiltration.

to 20% of cases at onset, but 77% to 85% of patients with GPA eventually develop glomerulonephritis within 2 years of GPA onset [10]. Additionally, necrotizing glomerulonephritis is present in approximately 70% to 80% of patients with generalized GPA, together with pulmonary involvement [17]. GPA supposedly begins with limited organ involvement and may change to a more systemic form in which the nose, lungs, and kidneys are involved [10]. In the present case, if the patient had not died of acute renal failure, other lesions would have appeared in the upper airway, lungs, etc., and the GPA would have progressed to a systemic form. In the diagnosis of renal-limited GPA, microscopic polyangiitis (MPA) is the most difficult disease to differentiate when the observation period is short, as in this case. Because the development of necrotizing crescentic glomerulonephritis is much more common in patients with MPA than in those with GPA, a certain period of observation will be necessary to improve the diagnostic accuracy of renal-limited GPA and MPA [2]. Moreover, PR3-ANCA is detected not only in about 65% to 90% of patients with GPA but also in approximately 10-20% of patients with MPA [2, 17, 18]. Conversely, in a small number of patients with GPA, MPO-ANCA may be detected or ANCA may not be detected, and most patients with

MPA (65-90%) have MPO-ANCA [2, 17, 18]. Therefore, PR3-ANCA positivity alone should not be used to diagnose GPA or determine the treatment. However, the frequency of ANCA being detected as a nonpathogenic autoantibody in patients aged 70 to 90 years is extremely low, less than 1%, and PR3-ANCA is supposed to be correlated with GPA severity and relapse potential, making it highly disease-specific (specificity $> 90\%$) [2, 3]. Based on these backgrounds, although there are certain exceptions, the patient was diagnosed with renal-limited GPA based on the combination of the strong positivity of PR3-ANCA (≥ 350 U/mL), negative MPO-ANCA, onset of acute kidney failure, and consistent renal pathology.

Conclusion

The patient clinically diagnosed with probable AD was confirmed to have definite AD by autopsy. The subacute cognitive deterioration and impaired consciousness observed in the patient on admission were found to be caused by delirium resulting from severe dehydration or uremia, not by CNS involvement due to GPA. Necrotizing crescentic glomerulonephritis

and peritubular inflammatory cell infiltration were also found. Therefore, the cause of the acute progressive renal failure in this case was determined to be PR3-ANCA-positive GPA, and the autopsy revealed no GPA-related lesions in other parts of the body aside from the kidney. It is rare to encounter cases of PR3-ANCA-positive GPA with renal-limited vasculitis and acute renal failure as the initial manifestation, as in the present case. Making an accurate clinical diagnosis of older patients suffering from various diseases in multiple organs is extremely challenging. Although an autopsy has the limitation of a terminal image, it is extremely useful in elucidating the pathophysiology of older patients as it enables observation over a wide area, unlike biopsy. We report this case to utilize the results of this autopsy for the development of clinical medicine.

Acknowledgments

None to declare.

Financial Disclosure

No funding was received.

Conflict of Interest

The authors declare that there is no conflict of interest relevant to this article.

Informed Consent

Written informed consent was obtained from the patient's family for publication of this case report.

Author Contributions

S. Tetsuka drafted the original manuscript. Y. Dobashi performed the autopsy and described the histopathological findings. All authors reviewed and revised the manuscript draft and approved the final version for submission.

Data Availability

The data used to support the findings of this study are available from the corresponding author upon request.

Abbreviations

GPA: granulomatosis with polyangiitis; ANCA: antineutrophil cytoplasmic antibody; PR3-ANCA: ANCA directed against proteinase 3; CNS: central nervous system; AD: Alzheimer's

disease; FLAIR: fluid-attenuated inversion recovery; MRI: magnetic resonance imaging; Cr: creatinine; BUN: blood urea nitrogen; BPSD: behavioral and psychological symptoms of dementia; CT: computed tomography; MPA: microscopic polyangiitis; MPO: myeloperoxidase; NINCDS-ADRDA: National Institute of Neurological and Communicative Disorders and Stroke AD and Related Disorders Association

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