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Cholesterol Emboli Co-Existing with Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis in a 76-Year-Old Woman

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Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis injures small vessels and causes severe systemic organ injury. Main target antigens of ANCA are myeloperoxidase and proteinase 3. ANCA strongly associates with the development and progression of the vasculitis. Its manifestations include rapidly progressive glomerulonephritis, interstitial pneumonitis, alveolar hemorrhage, purpura, and neurological disorder. Most patients with ANCA-associated vasculitis in Japan are elderly and have atherosclerotic risk factors. Cholesterol emboli are systemic vascular inflammation triggered by cholesterol crystals. Cholesterol emboli cause kidney dysfunction and ischemia of the intestines, brain, heart, skin, and peripheral nerves. Diabetes mellitus, hypertension, hyperlipidemia, and history of cardiovascular diseases are risk factors of the development of cholesterol emboli. We report a case of ANCA-associated vasculitis coexisting with cholesterol emboli. A 76-year-old woman was diagnosed with ANCA-associated interstitial pneumonitis. She rapidly developed progressive glomerulonephritis, purpura, and peripheral sensory nerve disorder. A kidney biopsy revealed that renal dysfunction was caused by vasculitis of the interlobular arteries and cholesterol emboli. A skin biopsy revealed that purpura was caused by cholesterol emboli. Glucocorticoid and statin therapies were administered. Thereafter, the renal function and other symptoms improved and stabilized. The representative symptoms of ANCA-associated vasculitis and cholesterol emboli are closely similar, and it is difficult to distinguish between these diseases when they coexist. Because the background characteristics of patients with ANCA-associated vasculitis and risk factors of cholesterol emboli overlap, at the time of diagnosing ANCA-associated vasculitis, clinicians should consider the possibility of cholesterol emboli coexistence.

Keywords: anti-neutrophil cytoplasmic antibody; anti-neutrophil cytoplasmic antibody-associated vasculitis; cholesterol emboli; purpura; rapidly progressive glomerulonephritis
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Introduction

Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis is characterized by ANCA positivity (Pagnoux 2016; Jennette and Nachman 2017; Harigai et al. 2019; Harada et al. 2019). Main target antigens of ANCA are myeloperoxidase (MPO) and proteinase 3 (PR3). ANCA strongly associates with the development and progression of the vasculitis (Pagnoux 2016; Jennette and Nachman 2017). ANCA-associated vasculitis often injures small vessels, resulting in severe kidney and lung dysfunction, and also develops nerve or skin lesions (Pagnoux 2016; Jennette and Nachman 2017; Harigai et al. 2019; Harada et al. 2019). ANCA-associated vasculitis includes the following clinical types: microscopic polyangiitis, granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, and renal-limited vasculitis (Pagnoux 2016; Jennette and Nachman 2017). Patients with ANCA-associated vasculitis are treated with glucocorticoid and immunosuppressive agents such as cyclophosphamide and rituximab (Pagnoux 2016; Jennette and Nachman 2017; Harigai et al. 2019). Most patients with ANCA-associated vasculitis, especially in Japan, are elderly. Therefore, infectious complications and cardiovascular diseases often develop during the clinical course of ANCA-associated vasculitis (Pagnoux 2016; Jennette and Nachman 2017; Harigai

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et al. 2019; Harada et al. 2019). Most patients with ANCA-associated vasculitis have atherosclerotic risk factors such as hypertension and diabetes mellitus (Harada et al. 2019). Cholesterol emboli are systemic disorders caused by rupture of atherosclerotic plaques in large arteries. After the rupture of atherosclerotic plaques, disseminated cholesterol crystals cause obstruction of small arteries, resulting in ischemic injuries in various organs such as the kidney, gastrointestinal area, brain, and heart (Fukumoto et al. 2003; Meyrier 2006). Cholesterol crystals also induce inflammation, and platelet aggregation and thrombosis in systemic vessels (Fukumoto et al. 2003; Meyrier 2006; Mulay et al. 2018). Other symptoms include sensory nerve disturbances and skin lesions (Fukumoto et al. 2003; Meyrier 2006). General atherosclerotic factors such as hypertension, diabetes mellitus, and smoking are risk factors for cholesterol emboli (Fukumoto et al. 2003). Although catheterization and anticoagulation are risk factors for cholesterol emboli, spontaneous cholesterol emboli also occur (Fukumoto et al. 2003; Meyrier 2006). A blue toe is one of the representative clinical findings of cholesterol emboli. In addition, biopsy-proven cholesterol crystals are the key to the diagnosis of cholesterol emboli. Cholesterol emboli are generally treated using statins and glucocorticoids (Fukumoto et al. 2003; Meyrier 2006).

Here, we report a patient with rapidly progressive glomerulonephritis, probably caused by ANCA-associated vasculitis and cholesterol emboli. Clinical manifestations of ANCA-associated vasculitis are similar to those of cholesterol emboli, including renal failure, nerve symptoms, and skin lesions. We investigated the clinical features of ANCA-associated vasculitis and cholesterol emboli, and the association between ANCA-associated vasculitis and cholesterol emboli.

Case Report

The patient is a 76-year-old woman with rapidly progressive glomerulonephritis and a history of hypertension. Prior to the present admission, she had undergone a follow-up examination for an interstitial shadow of the lung that was found on computed tomography four years prior. Because interstitial shadow worsened and positivity of myeloperoxidase-ANCA (MPO-ANCA 156 U/mL) was found, she was diagnosed with interstitial pneumonitis associated with ANCA-associated vasculitis one year prior to the present episode. She was treated with oral prednisolone (PSL 30 mg/day); the dose was tapered to 12.5 mg/day for approximately four months.

During her clinical course, she noticed purpura on the bilateral lower limbs and sensory disturbance in lower legs two months prior to admission. Her renal function was within normal range (serum creatinine 0.6 mg/dL) two months before her admission. However, her serum creatinine level rapidly increased to 3.71 mg/dL by the time of admission. In addition, her serum potassium level was high at 7.4 mmol/L. She was admitted to our hospital. Vital signs were notable for blood pressure 143/92 mmHg. Her physical examination was notable for purpura on both lower limbs and a sensory disturbance in her lower legs. A blue toe was not observed. Elevated white blood cell count (15,320 /μL) with normal eosinophil count was observed. The levels of blood urea nitrogen (58.9 mg/dL), serum creatinine (3.48 mg/dL) and C-reactive protein (CRP; 3.37 mg/dL) were elevated. In addition, urinalysis revealed that she had proteinuria (0.69 g/gCr), an elevation of N-acetyl-β-D-glucosaminidase (NAG) (12.2 U/L), and β2-microglobulin (β2-MG) (41,679 μg/L). Hematuria was not detected. Tests for anti-nuclear antibody, and anti-double stranded DNA antibody were negative. Serum complement levels were within normal range. MPO-ANCA was positive with a high titer (244.5 U/mL). The laboratory data are presented in Table 1.

Based on the positivity of MPO-ANCA preceding interstitial pneumonitis, purpura, and sensory disturbance, rapid decline in renal function was clinically diagnosed as a symptom of systemic ANCA-associated vasculitis. We categorized her ANCA-associated vasculitis as microscopic polyangiitis. The Birmingham Vasculitis Activity Score (version 3) was evaluated as 22 (General: 2, Cutaneous: 2, Mucous membranes / eyes: 0, ENT: 0, Chest: 0, Cardiovascular: 0, Abdominal: 0, Renal: 12, Nerve system: 6) (Mukhtyar et al. 2009). In addition, we diagnosed her with rapid progressive glomerulonephritis due to ANCA-associated vasculitis. We therefore evaluated the disease severity based on the Japanese guideline of rapid progressive glomerulonephritis (Yamagata et al. 2012). According to the guideline, the score of the current case was 6 points, and her clinical severity was categorized as grade III (Yamagata et al. 2012). Methylprednisolone pulse and oral glucocorticoid therapies were recommended. Intravenous methylprednisolone pulse therapy was performed and an oral dose of PSL (40 mg/day, approximately 1.0 mg/kg/day) was administered. Because of her hyperkalemia and kidney dysfunction, hemodialysis was performed once. After initiation of glucocorticoid therapy, her renal function and inflammatory reaction gradually improved (Fig. 1). On the 8-day hospital stay, kidney and skin biopsies were performed. The renal specimen revealed that seven of 37 glomeruli exhibited global sclerosis. No crescent formation or fibrinoid necrosis was detected in any of the glomeruli. There was no obvious infiltration of inflammatory cells into the tubulointerstitial area (Fig. 2A-C). There was no mesangial cell proliferation or abnormalities in the glomerular basal membranes, whereas intracapillary proliferation was detected segmentally (Fig. 2D). Hyalinosis and exudative lesions were observed in glomerular vascular pole, indicating vascular endothelial injuries in afferent and efferent arterioles (Fig. 2E). Vascular endothelial cell proliferation and fibrous lesions were detected in the interlobular arteries, resulting in narrowing of interlobular arteries (Fig. 2F). Internal and external elastic laminas of the interlobular arteries were torn. These findings suggested that severe

Table 1. Main clinical data of the current case at hospital admission.

Urinalysis				TC	209	(142-219)	mg/dL
Protein	1+			HDL-C	49	(> 40)	mg/dL
	0.69	(< 0.14)	g/gCr	LDL-C	120	(0-139)	mg/dL
Hematuria	-			TG	111	(< 149)	mg/dL
Urinary RBC	< 1		/HPF	AST	9	(13-30)	U/L
NAG	12.2	(0.3-11.5)	U/L	ALT	15	(7-23)	U/L
β2-MG	41,679	(50-210)	μg/L	LDH	165	(124-222)	U/L
				ALP	185	(106-322)	U/L
Blood analysis				γGT	25	(9-32)	U/L
WBC	15,320	(3,300-8,600)	/μL	T.Bil	0.5	(0.40-1.50)	mg/dL
Neut	96	(41.8-75.0)	%	CK	17	(41-153)	U/L
Lym	2.9	(18.5-48.7)	%	Glu	114	(73-109)	mg/dL
Mono	0.8	(2.2-7.9)	%	CRP	3.47	(0.00-0.14)	mg/dL
Eos	0.1	(0.4-8.7)	%	HbA1c	6.3	(4.9-6.0)	%
Baso	0.2	(0.2-1.5)	%	IgA	165	(93-393)	mg/dL
Hb	11.9	(11.6-14.8)	g/dL	IgM	79	(50-269)	mg/dL
Plt	38.6	(15.8-34.8)	$\times 10^4/\mu$L	IgG	848	(861-1747)	mg/dL
PT-INR	1.00	(0.85-1.15)		RF	283	(0-14)	U/mL
APTT	23.4	(23-38)	sec	C3	109	(73-148)	mg/dL
Fib	500	(180-350)	mg/dL	C4	20.9	(11.0-31.0)	mg/dL
D-dimer	3.2	(0.0-1.0)	μg/mL	CH50	64.6	(30.0-53.0)	U/mL
Total protein	5.8	(8.1-6.6)	g/dL	ANA	(–)		
Albumin	2.8	(5.1-4.1)	g/dL	ds-DNA antibody	4.7	(0.0-12.0)	IU/mL
BUN	58.9	(8.0-20.0)	mg/dL	PR3-ANCA	< 1.0	(< 3.4)	U/mL
Cr	3.48	(0.46-0.79)	mg/dL	MPO-ANCA	244.5	(< 3.4)	U/mL
UA	6.7	(2.6-5.5)	mg/dL	Anti-GBM antibody	< 2.0	(< 2.9)	U/mL
Na	139	(138-145)	mEq/L				
K	7.4	(3.6-4.8)	mEq/L				
Cl	112	(101-108)	mEq/L				
Corrected Ca	9.5	(8.7-9.9)	mg/dL				
P	4.3	(2.7-4.6)	mg/dL				

Reference values are shown in parentheses.
NAG, N-acetyl-β-D-glucosaminidase; β2-MG, β2-microglobulin; WBC, white blood cell count; Hb, hemoglobin; Plt, platelet; PT-INR, prothrombin time international normalized ratio; APTT, activated partial thromboplastin time; Fib, fibrinogen; BUN, blood urea nitrogen; Cr, creatinine; UA, uric acid; Na, sodium; K, potassium; Cl, chloride; Ca, calcium; iP, inorganic phosphorus; TC, total cholesterol; HDL-C, high density lipoprotein cholesterol; LDL-C, low density lipoprotein cholesterol; TG, triglycerides; AST, aspartate aminotransferase; ALT, alanine aminotransferase; LDH, lactate dehydrogenase; ALP, alkaline phosphatase; γGT, gamma-glutamyl transpeptidase; T.Bil, total bilirubin; CK, creatinine phosphokinase, Glu, glucose; CRP, C-reactive protein; IgA, immunoglobulin A, IgM, immunoglobulin M; IgG, immunoglobulin G; RF, rheumatoid factor; ANA, anti-nuclear antibody; C3, complement 3; C4, complement 4, CH50, complement hemolytic activity assay; MPO-ANCA, myeloperoxidase-anti-neutrophil cytoplasmic antibody; PR3-ANCA, proteinase 3-anti-neutrophil cytoplasmic antibody; Anti-GBM antibody, anti-glomerular basement membrane antibody.

vascular inflammation occurred throughout the entire layer of interlobular arteries (Fig. 2G). Needle-shaped cholesterol emboli were observed in the lumen of interlobular arteries (Fig. 2H). Immunofluorescence analysis revealed neither immunoglobulin nor complement deposition. Electron microscopic analysis demonstrated that electron-dense depositions were not observed in the basal membrane or mesangial region. Although crescent formation or fibrinoid necrosis was not detected in the glomeruli, whole-layer vasculitis was detected in relatively thick vessels and interlobular arteries. Therefore, we speculated that ANCA-associated vasculitis would be a major cause of renal dysfunction.

Cholesterol emboli might also cause the decline of renal function in the current case. Although purpura of the bilateral lower limbs and sensory nerve disturbance appeared to be caused by ANCA-associated vasculitis, skin biopsy revealed needle-shaped cholesterol emboli without vasculitis lesions (Fig. 3A, B). There was no granuloma formation or eosinophil infiltration in the kidney and skin

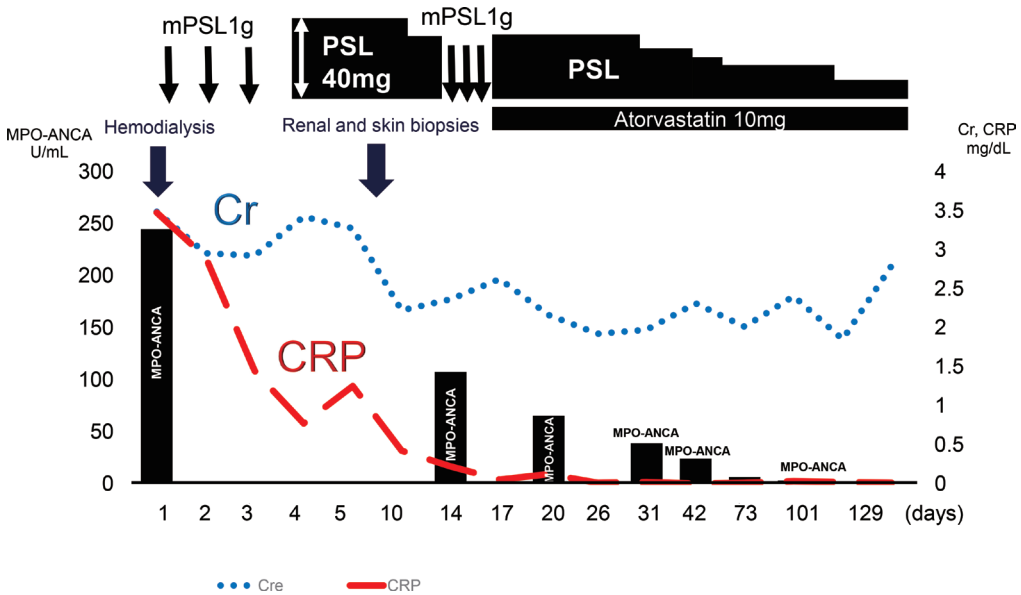

Fig. 1. Clinical course of serum creatinine, C-reactive protein and ANCA titer.
After initiation of glucocorticoid therapy, her renal function and inflammatory reaction gradually improved. The dosage of prednisone was reduced to 30 mg/day after her 10th hospital day, and methylprednisolone pulse therapy was administered again from her 15th hospital day. Atorvastatin was initiated from her 18th hospital day when she was diagnosed with cholesterol emboli. The dosage of prednisone was subsequently tapered gradually at two-to-four-week intervals. During the course of immunosuppressive therapy, Serum creatinine level improved and stabilized (2.0-2.5 mg/dl), and titer of MPO-ANCA decreased and finally turned to be negative at five months after the start of methylprednisolone and consecutive glucocorticoid therapy.
Cr, creatinine; CRP, C-reactive protein; MPO-ANCA, myeloperoxidase-anti-neutrophil cytoplasmic antibody; mPSL, methylprednisolone; PSL, prednisolone.

biopsy specimens. Magnetic resonance imaging of the aorta revealed atherosclerotic and calcified plaques in the thoracoabdominal aorta, suggesting that these lesions had cholesterol crystal sources (Fig. 4).

Based on these findings, the patient was diagnosed with ANCA-associated vasculitis coexisting with cholesterol emboli. The oral dosage of PSL was reduced to 30 mg/day after her 10th hospital day, and intravenous methylprednisolone pulse therapy was performed again at her 15th day after admission. Atorvastatin treatment was initiated from her 18th hospital day because she was diagnosed with cholesterol emboli. The oral dosage of PSL was subsequently tapered gradually over two-to-four-week intervals. During the course of immunosuppressive therapy, her serum creatinine level improved and stabilized (2.0-2.5 mg/dl), and her titer of MPO-ANCA decreased and turned negative after five months from the start of methylprednisolone pulse and consecutive glucocorticoid therapy (Fig. 1).

Consent

Written informed consent was obtained from the patient.

Discussion

Here, we present a case of ANCA-associated vasculitis coexisting with cholesterol emboli. ANCA-associated vasculitis was diagnosed prior to the diagnosis of cholesterol emboli in the current case; thus, the possibility of cholesterol emboli was not considered at the start of initial therapy. However, renal dysfunction, peripheral nerve disorder and purpura of the lower limbs were observed. Although skin lesions were likely caused by cholesterol emboli based on the biopsy results, renal dysfunction and peripheral sensory nerve disorder of the lower limbs could be caused by both ANCA-associated vasculitis and cholesterol emboli. Both ANCA-associated vasculitis and cholesterol emboli can cause rapid progressive kidney dysfunction, and therefore, ANCA-associated vasculitis and cholesterol emboli should be considered as causes of acute kidney injury (Fukumoto et al. 2003; Meyrier 2006; Pagnoux 2016; Jennette and Nachman 2017; Harigai et al. 2019; Harada et al. 2019).

In the current case, urinalysis did not show hematuria, which is atypical in general cases of rapidly progressive glomerulonephritis due to ANCA-associated vasculitis. According to a previous study of urinalyses in ANCA-associated vasculitis patients, 14% of patients had negative of hematuria and/or proteinuria (Hasegawa et al. 2018). Patients with ANCA-associated vasculitis and hematuria present with crescent formation more frequently than patients without hematuria (Hasegawa et al. 2018). In the current case, there was no crescent formation in the kidney biopsy. The lesion of vasculitis was not distributed to the glomerular capillaries but was instead distributed to the interlobular arteries. It is therefore suspected that urinalysis in the current case did not indicate hematuria.

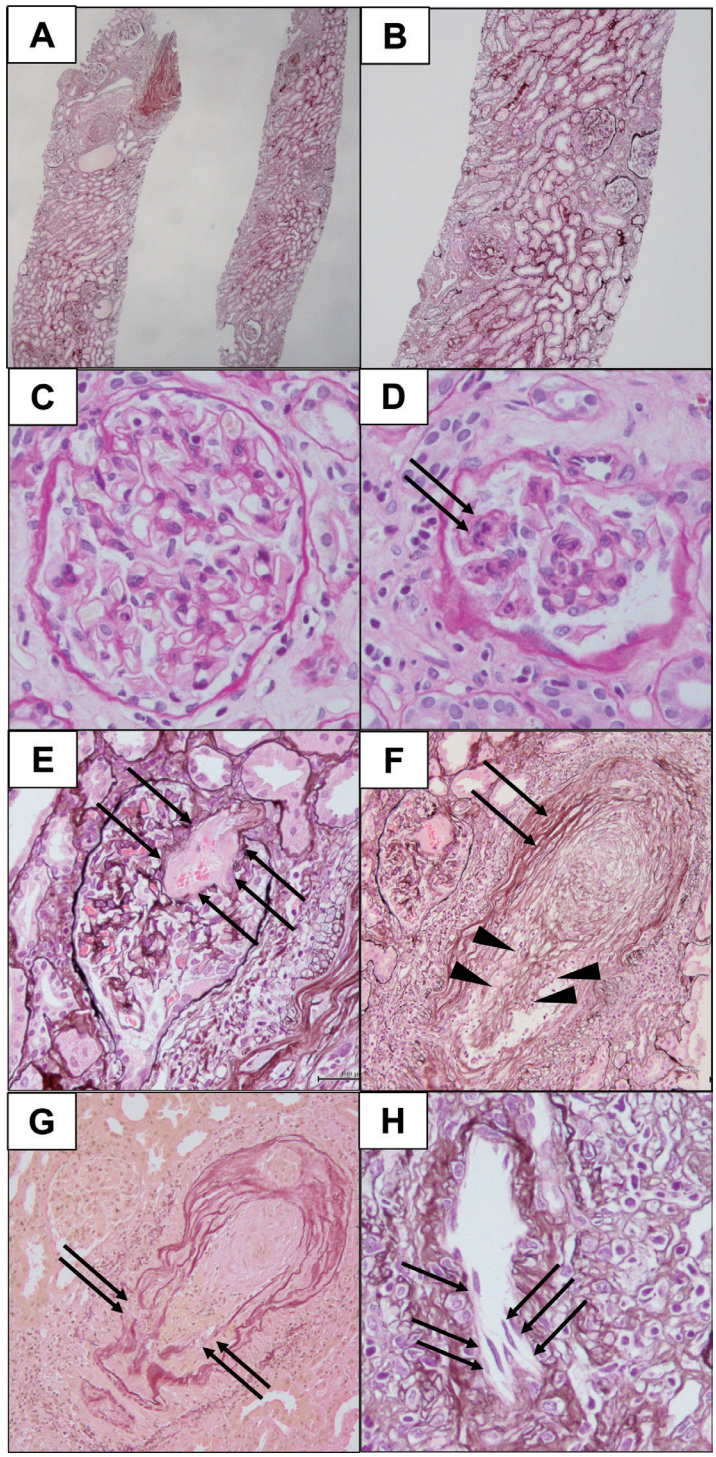

Fig. 2. Pathological findings of kidney biopsy.
The kidney specimen demonstrates that 37 glomeruli, of which seven exhibited global sclerosis. Crescent formation, or fibrinoid necrosis was not detected in the glomeruli. There was no obvious infiltration of inflammatory cells into tubulointerstitial area (Periodic acid-Schiff staining) (A, B, C). Segmental intracapillary proliferation (arrows) is detected (Periodic acid-Schiff staining) (D). Hyalinosis and exudative lesion (arrows) are observed in afferent and efferent arterioles at glomerular vascular pole (Periodic acid-methenamine-silver staining) (E). Vascular endothelial cell proliferation (arrowheads) and fibrous lesion (arrows) are detected in the interlobular arteries (Periodic acid-methenamine-silver staining). As a result, interlobular arteries are narrowed (F). Internal and external elastic laminas of the interlobular arteries are torn (arrows) (Elastica van Gieson staining) (G). Needle-shaped cholesterol emboli (arrows) are observed in the lumen of interlobular artery (Periodic acid-methenamine-silver staining) (H).

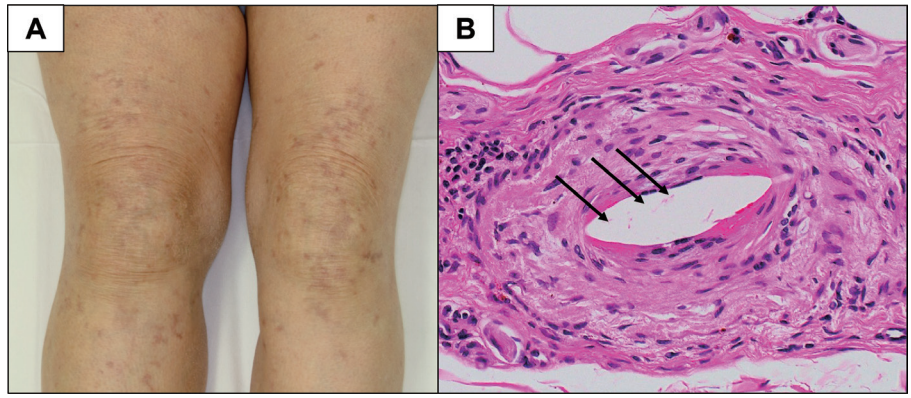

Fig. 3. Findings of skin biopsy.
Purpura on the bilateral lower limbs. (A). Skin biopsy specimen reveals needle-shaped cholesterol emboli (arrows), whereas lesion of leuko-cytoclastic vasculitis is not detected (B).

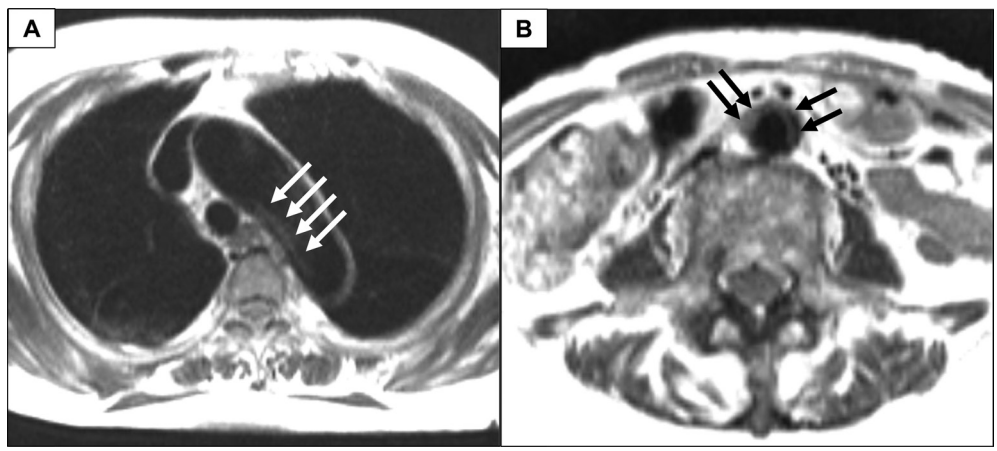

Fig. 4. Findings of magnetic resonance imaging.
Magnetic resonance imaging of the thoracoabdominal aorta. Atherosclerotic and calcified plaque (arrows) are detected in the aortic arch and abdominal aorta (A and B).

To investigate the clinical course and characteristics of ANCA-associated vasculitis coexisting with cholesterol emboli, we reviewed previous case reports on the coexistence of cholesterol emboli with ANCA-associated vasculitis (Kaplan-Pavlovcic et al. 1998; Palmgren et al. 2000; Maeshima et al. 2001; Aviles et al. 2002; de et al. 2002; Sugimoto et al. 2006), as well as coexistence cases of cholesterol emboli and ANCA positivity without ANCA-associated vasculitis manifestation (Peat and Mathieson 1996; Kaplan-Pavlovcic et al. 1998; Delen et al. 2003; Miguélez et al. 2003; Maejima et al. 2010; Zhang et al. 2016) (Table 2). Most of the patients in such studies were elderly males, with the age distribution ranging from 47 to 76 years, and the male to female ratio was 10:3 (Table 2). All patients had a history of hypertension, hyperlipidemia or cardiovascular diseases (Table 2). A prior study reported that the general background characteristics of patients with ANCA-associated vasculitis were predominantly elderly individuals with atherosclerotic risk factors such as hypertension and diabetes (Harada et al. 2019). These characteristics are consistent with those in the current study and overlap with the risk factors for cholesterol emboli. Seven cases were detected as MPO-ANCA, three cases were detected as PR3-ANCA, one case was detected as both MPO and PR3-ANCA, and two cases did not describe the type of ANCA.

Regarding the clinical course (order of the onset of ANCA-associated vasculitis and cholesterol emboli), three patterns were observed in previous reports. In six (46%) cases, cholesterol emboli developed initially, and then ANCA positivity was detected without any manifestation of ANCA-associated vasculitis. In five (39%) cases, cholesterol emboli developed, and then ANCA-associated vasculitis subsequently developed. Not all patients with ANCA positivity exhibited vasculitis symptoms, therefore we defined patients who had ANCA positivity with vasculitis symptoms as ANCA-associated vasculitis, and distinguished them from patients who had ANCA positivity without symptoms of vasculitis. These findings suggest that cholesterol emboli often induced ANCA positivity or ANCA-associated vasculitis. It is suspected that cholesterol crystals can damage the vascular endothelium and

Table 2. Clinical findings of previously reported 12 cases and the current case of the coexisting of cholesterol emboli with ANCA-associated vasculitis or ANCA positivity.

	Age	Sex	ANCA type	History of atherosclerotic diseases	Order of the onset of AAV and CE	ESRD	Death
Peat and Mathieson 1996	73	M	No data	HT, CVD	CE preceded and ANCA positivity without AAV manifestation	+	+
Kaplan-Pavlovcic et al. 1998	63	M	MPO	HT, CVD	AAV preceded CE	+	–
Kaplan-Pavlovcic et al. 1998	69	F	MPO	HT, HL, CVD	CE preceded and ANCA positivity without AAV manifestation	–	–
Palmgren et al. 2000	No data	M	No data	CVD	CE preceded AAV	–	+
Maeshima et al. 2001	50	M	MPO, PR3	CVD	CE preceded AAV	+	–
Aviles et al. 2002	67	M	MPO	CVD	CE preceded AAV	–	–
de et al. 2002	47	M	PR3	HT, HL, CVD	CE preceded AAV	–	–
Delen et al. 2003	65	F	MPO	HT, CVD	CE preceded and ANCA positivity without AAV manifestation	–	–
Miguélez et al. 2003	70	M	PR3	HT, HL, CVD	CE preceded and ANCA positivity without AAV manifestation	+	+
Sugimoto et al. 2006	75	M	MPO	CVD	CE preceded AAV	–	–
Maejima et al. 2010	76	M	MPO	CVD	CE preceded and ANCA positivity without AAV manifestation	–	+
Zhang et al. 2016	69	M	PR3	HT, HL, CVD	CE preceded and ANCA positivity without AAV manifestation	+	–
Current case	76	F	MPO	HT	AAV preceded CE	–	–

ANCA, anti-neutrophil cytoplasmic antibody; AAV, ANCA-associated vasculitis; CE, cholesterol emboli; CVD, cardiovascular diseases; ESRD, end stage renal disease; F, female; HL, hyperlipidemia; HT, hypertension; M, male; MPO, myeloperoxidase; PR3, proteinase 3.

cause inflammation, increased cytokine (release or production), and neutrophil activation, thus resulting in ANCA production and development of ANCA-associated vasculitis (Zhang et al. 2016).

In the present case, MPO-ANCA positivity was detected one year prior to the diagnosis of cholesterol emboli. This clinical course may be relatively rare, but it is possible that ANCA-associated vasculitis can cause cholesterol emboli. A report that described an atypical type of ANCA-associated vasculitis indicated that some cases of ANCA-associated vasculitis could damage not only small vessels, but also large vessels, such as in the aorta (Chirinos et al. 2004). When large vessel vasculitis occurs, vascular inflammation may break down plaques on the aorta and induce cholesterol emboli. Based on the atypical distribution of vasculitis seen in our current study, the presence of interlobular arteritis but not glomerular capillaritis, as well as the detection of obvious atherosclerotic and calcified lesion in the thoracoabdominal aorta, may support our proposed mechanism.

Concerning prognosis, the renal survival and survival rate of the coexisting of cholesterol emboli with ANCA-associated vasculitis or ANCA positivity were 62% (of 13 patients, 5 patients required maintenance hemodialysis) and 69% (of 13 patients, 4 patients were died), respectively (Table 2). The renal survival and survival rate of the patients with ANCA-associated vasculitis at a time interval of 12 months were 78% and 79%, respectively (Yamagata et al. 2012). The renal survival and survival rate of the patients with cholesterol emboli were 84% and 84%, respectively (Fukumoto et al. 2003). Thus, the prognosis of these coexisting cases of cholesterol emboli with ANCA-associated vasculitis or ANCA positivity may be not so different from ANCA-associated vasculitis or cholesterol emboli alone.

In conclusion, we treated a woman with ANCA-associated vasculitis coexisting with cholesterol emboli. The clinical symptoms of ANCA-associated vasculitis and cholesterol emboli are similar, and it can be difficult to distinguish between them. Most patients who presented with ANCA-associated vasculitis coexisting with cholesterol emboli were elderly men with a history of hypertension, hyperlipidemia, and/or cardiovascular diseases. Given that some cases of ANCA-associated vasculitis present with large vessel vasculitis such as in the aorta, ANCA-associated vasculitis might be associated with the development of cholesterol emboli. Further evaluation of similar cases is needed to verify this association. Overlap does exist between the background characteristics of patients with ANCA-associated vasculitis and risk factors of cholesterol emboli; thus, at the time of diagnosing ANCA-associated vasculitis, clinicians should consider the possibility of the coexistence of cholesterol emboli.

Conflict of Interest

The authors declare no conflict of interest.

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