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[Case Report]

HYPERTROPHIC CRANIAL PACHYMENINGITIS IN MPO-ANCA-RELATED VASCULITIS : A CASE REPORT AND LITERATURE REVIEW

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Abstract : A 75-year-old woman presented with rapidly progressive glomerulonephritis with positive results for anti-myeloperoxidase anti-neutrophil cytoplasmic antibody (MPO-ANCA). Corticosteroid therapy was successfully introduced. However, 7 months later, magnetic resonance imaging revealed marked swelling in the falx cerebri and high density regions were apparent on gallium scintigraphy, leading to diagnosis of hypertrophic cranial pachymeningitis (HCP). Symptoms improved with intensified corticosteroid therapy, but radiological examination 9 months later revealed right nasal sinus inflammation accompanied by osteolytic change. Granulomatosis with polyangiitis (Wegener's) was finally diagnosed. HCP is an important complication in MPO-ANCA-related vasculitis, and needs to be considered during the clinical course.

Key words : anti-myeloperoxidase anti-neutrophil cytoplasmic antibody (MPO-ANCA), hypertrophic cranial pachymeningitis (HCP), granulomatosis with polyangiitis (Wegener's)

INTRODUCTION

Hypertrophic cranial pachymeningitis (HCP) is a chronic, fibrosing, inflammatory disorder that involves the dura mater of the brain and can occur in patients of all age groups, although incidence peaks in the sixth decade of life[1]. Several causes have been recognized, including infections (such as syphilis or tuberculosis), connective tissue diseases, neoplasms and vasculitides[1-3].

Several anti-neutrophil cytoplasmic antibody (ANCA)-related cases of HCP have been reported since the mid-1990s, mainly from Japan. Interestingly, most cases have presented as MPO-ANCA-positive, limited-type vasculitis, and are less likely to be accompanied by active nephritis such as rapidly progressive glomerulonephritis (RPGN) or crescentic glomerulonephritis in the clinical course[4-6]. Cases of anti-myeloperoxidase ANCA (MPO-ANCA)-related HCP have included patients with microscopic polyangiitis (MPA) and, notably, patients with granulomatosis with polyangiitis (GPA) (Wegener's)[4-7]. This latter pathology is generally characterized by proteinase 3-ANCA.

Here, we present the case of a patient who developed HCP during successful treatment of MPO-ANCA-related vasculitis and was finally diagnosed with GPA. We have reviewed case reports of MPO-ANCA-related HCP to show the distinctive clinical forms of this pathology.

CASE REPORT

A 75-year-old woman presented with progressive renal dysfunction (serum creatinine (s-Cre), 2.0 mg/dL ; estimated glomerular filtration rate (eGFR), 19.5 mL/min/1.73 m^2) accompanying hematuria and proteinuria, with the presence of MPO-ANCA (196 EU) from 9 months before admission. She was diagnosed with MPO-ANCA-related vasculitis by renal biopsy. Histological findings (Fig. 1) showed vasculitis of small-sized arteries as the main finding, compatible with ANCA-related vasculitis. Upon corticosteroid therapy with oral prednisolone (PSL)

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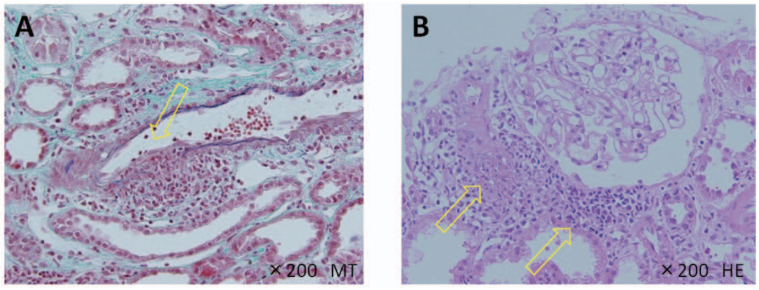

Fig. 1. Renal biopsy finding. Small-sized arteries are the main vessels affected by vasculitis in renal histology. A) Infiltration of inflammatory cells and destroyed vascular structures (yellow arrow). B) Small vessel vasculitis and inflammation spreading to adjacent glomeruli (yellow arrow). One of ten sampled glomeruli is sclerotic, typical crescent formations are absent and some inflammation cells such as neutrophils are present in the glomerular capillary. No granuloma formation is evident.

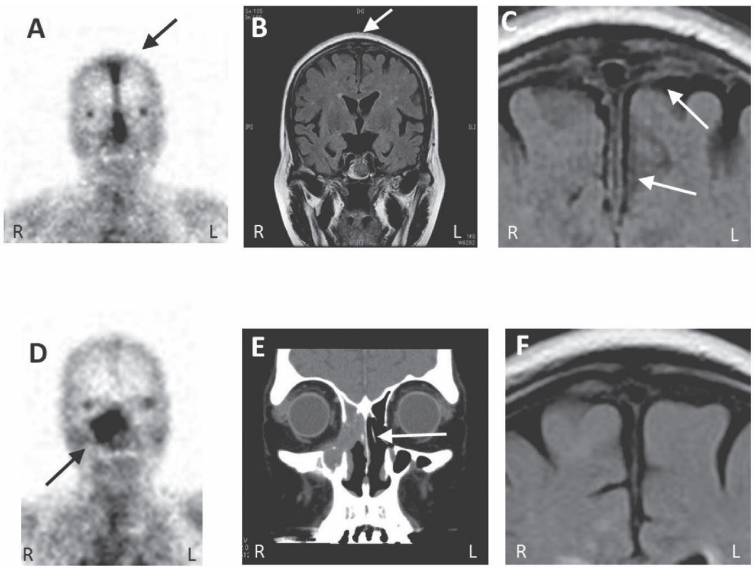

Fig. 2. Imaging findings. A) Gallium scintigraphy showing hypertrophic pachymeningitis (HCP). Dense accumulation is seen at the falx cerebri (black arrow). B) Magnetic resonance imaging (MRI) showing HCP. A slightly swollen falx cerebri is evident (white arrow). C) Enlargement of the thickened region of dura mater is evident (white arrow). D) Ga scintigraphy indicating sinusitis (black arrow). E) Contrast-enhanced computed tomography showing right nasal sinus inflammation with osteolytic change (white arrow). F) Swelling in the falx cerebri is not apparent on MRI at the time of sinusitis.

at an initial dose of 40 mg/day (0.9 mg/kg/day), urine abnormalities disappeared, renal function stabilized (s-Cre, 1.2 mg/dL), and MPO-ANCA titer decreased to within the normal range.

The patient displayed persistent frontal headache, general fatigue and fever (37-38°C) from 2 months before admission with elevated levels of C-reactive protein (CRP). She was admitted to our hospital when symptoms persisted. Physical examination did not reveal any neurological abnormalities such as neck rigidity or disturbance of consciousness. She was in a state of remission from nephritis associated with vasculitis during corticosteroid taper at a PSL dose of 15 mg/day. According to laboratory data, inflammatory findings (CRP, 9.34 mg/dL ; erythrocyte sedimentation rate (ESR), 44 mm/h) and renal function (s-Cre, 1.21 mg/dL ; eGFR, 33.9 mL/min/1.73 m^2) were stable without active urinary sediments. Both MPO-ANCA and PR3-ANCA remained negative. Cerebrospinal fluid (CSF) analysis revealed modest pleocytosis (white blood cell (WBC) count, 20/mm^3 ; 50% lymphocytes), slightly elevated protein and glucose levels, and normal cerebrospinal fluid pressure. Gallium scintigraphy detected distinctive dense accumulation in the falx cerebri (Fig. 2A), and cranial magnetic resonance imaging (MRI) showed swelling in the same area (Fig. 2B, C), suggesting the pres-

ence of HCP. Clinically, headache and fever disappeared within 7 days and follow-up MRI revealed regression of the falx swelling 4 months after initiating corticosteroid therapy. The clinical course of the patient is shown in Figure 2.

Nine months after HCP onset, the patient reported aches and pains in the maxilla, general fatigue and slight fever. Laboratory data showed elevated levels of inflammatory markers (CRP, 11.6 mg/dL ; ESR, 61 mm/h), although renal function remained stable (s-Cre, 1.14 mg/dL ; eGFR, 36.0 mL/min/1.73 m^2). Gallium scintigraphy detected a dense accumulation in the area of the right maxillary sinus (Fig. 2D), and contrast-enhanced computed tomography showed right nasal sinus inflammation with osteolytic change (Fig. 2E). Despite nasal mucosa biopsy, granulomatous formation was not confirmed. Histological findings revealed only chronic active inflammation associated with infiltration of lymphocytes, neutrophils and plasma cells. Finally, she was diagnosed with GPA using diagnostic criteria[8]. These criteria represent an algorithm for the diagnosis of vasculitides published by Watts et al in 2007, and were designed for use in epidemiological research. In terms of GPA, even if organ involvement cannot be confirmed histologically, we can classify GPA according to ANCA and surrogate markers (in the present case, sinusitis with osteolytic change). The patient therefore underwent intensification of corticosteroid therapy. MPO-ANCA and PR3-ANCA both remained negative. Furthermore, both renal involvement and HCP had been in remission since the previous treatment with PSL at 12.5 mg/day and Mizoribine (MZR) at 125 mg/day. Sinusitis improved and maintenance therapy for GPA has since been successfully continued.

DISCUSSION

In the present case, HCP developed during the course of successful treatment for MPO-ANCA-related vasculitis. Although cranial nerve dysfunction such as optic nerve neuropathy, oculomotor disturbance and facial nerve palsy are frequently complications in HCP[9], the patient showed no symptoms suggestive of cranial nerve dysfunction. We then reviewed 29 case reports of MPO-ANCA-related HCP found on PUBMED to confirm details of the clinical course (Table 1)[4-7,9-34]. Twenty-three of the 29 cases (79%) were reported from Japan, 18 (62%) showed HCP developed as an initial target involvement of vasculitis, and 17 (59%) showed limited form. Notably, only three cases (10%) showed active nephritis such as RPGN or crescentic glomerulonephritis in the clinical course[4-6]. In detail, two cases simultaneously developed active GN and HCP, and one patient developed RPGN after the onset of HCP. Furthermore, regarding diagnosis of the type of vasculitis, 5 of 29 cases (17%) were diagnosed with MPA, while 8 cases (29%) were diagnosed with GPA. According to those case reports, active kidney involvement is rarely accompanied. In addition, MPO-ANCA-related HCP is more likely to be diagnosed with GPA than with MPA. The present case is unique in that the patient initially presented with RPGN. On the other hand, from the perspective of GPA, the incidence of central nervous system involvement with GPA has been reported in about 20% of cases, with HCP representing less than 1% of these[9,35]. We could not obtain a complete view of relationships between GPA and HCP, PR3 ANCA and HCP, because cases including full details of the clinical course were scarce.

Imaging findings from modalities such as MRI and scintigraphy were important diagnostic clues to HCP in the present case. In general, gadolinium-enhanced MRI is used as a standard for diagnostic imaging[1]. Since gadolinium enhancement in this case was relatively contraindicated due to the unstable renal function (s-Cre, 1.1-1.4 mg/dL ; eGFR, 29-36 mL/min/1.73 m^2), use of MRI and Ga scintigraphy allowed successful diagnosis[36]. In this case, swelling in the falx cerebri was not detected on MRI at the time of sinusitis (Fig. 2F), and HCP was not a complication at the time of recurrence.

Although most cases appear responsive to corticosteroid therapy, as seen in the present case, some cases recur or progress despite treatment[1]. Combined therapy with corticosteroid and other immunosuppressive drugs, such as cyclophosphamide, azathioprine or methotrexate, has been reported as effective for HCP[2,4,20,37-39]. In the present case of HCP onset, after the introduction of methylprednisolone (m-PSL) pulse therapy, combination therapy with oral PSL and MZR promptly resolved all symptoms. A study on the rationale for MZR treatment is currently underway in Japan[40]. After the diagnosis of GPA, we conducted m-PSL pulse therapy twice and plasma exchange three times. In consideration of the risk of myelosuppression associated with previous treatment and the age of the patient, we selected cyclosporine as the immunosuppressant while continuing trimethoprim-sulfamethoxazole. Sinusitis improved and maintenance therapy for GPA has since been successfully

Table 1. Reported cases of MPO-ANCA-related hypertrophic cranial pachymeningitis.

No.	Year	Age, Sex	Country	MPO-ANCA	CRP (mg/dL) (ESR (mm/h))	Clinical diagnosis	Onset	Treatment	Renal involvement	Outcome	Ref.
1	1994	63, F	Japan	Positive ♣	17.5	HCP	New onset	CS, AZA	ND	Improved	[28]§
2	1995	77, F	Japan	Positive ♣	Negative	HCP	New onset	CS†	None	Improved	[27]§
3	1997	64, M	Japan	Positive*	Positive*	HCP, Horner syndrome	New onset	CS†	ND	Improved	[29]
4	1998	47, F	Japan	518 EU	4.2	HCP	New onset	CS†	None	Improved	[26]
5	1999	75, F	USA	128 U/mL‡	ESR, 113	GPA	New onset	CS†, CY	None	Improved	[30]
6		70, M	Japan	26 / 99 EU	3.1 / 6.0	Limited GPA	Relapse	CS, CY	None	Improved	[31]§
7	2000	72, F	Japan	65 EU	14.8	HCP, RA	New onset	CS	None	Improved	[24]
8		44, M	Japan	Positive*	3.6	HCP	New onset	CS†	ND	Improved	[25]
9		56, F	Japan	321 EU	17.1	MPA	New onset	CS†	Active vasculitis	Improved	[4]
10	2002	66, F	Japan	×220‡	ND	HCP	New onset	CS	ND (on HD)	Improved	[23]§
11	2003	63, F	Canada	14.4 U/mL	ND	Limited GPA	Relapse	CS, CY	None	Improved	[22]
12	2004	60, M	Japan	23 EU	12.8	GPA	Relapse	CS, CY	None	Improved	[20]
13		61, M	Japan	63 EU	14.4	Silicosis	New onset	CS†	None	Improved	[21]
14		67, M	Japan	38 EU	4.1	Silicosis	Relapse	CS	None	Improved	[21]
15		68, M	Japan	467 EU	ESR, 113	MPA	New onset	CS†, CY	Active vasculitis	Improved	[5]
16	2005	51, F	France	Positive ♣	4.7	HCP	New onset	CS†, CY, MTX	None	Improved	[19]
17	2006	54, M	France	Positive*	ESR, 15	GPA	Relapse	CS, CY	ND	Improved	[9]
18	2007	34, F	Japan	122 EU	0.12	HCP, PTU	New onset	CS	None	Improved	[18]
19	2008	77, F	Japan	MPO, 32 EU With PR3 positive	12.4	MPA	New onset	CS†	Active vasculitis	Dead	[6]
20	2009	80, F	Japan	70 EU	14.4	GPA	Relapse	CS, CY	Urinary abnormality only	Improved	[17]§
21	2010	65, M	Japan	Positive*	ND	MPA (probable)	Relapse	CS†, CY	None	Improved	[12]§
22		69, F	Italy	1.7 titer	1.3	HCP	New onset	CS†, AZA	None	Improved	[13]
23		61, M	Japan	ND	ND	GPA	Relapse	CS, CY	None	Improved	[14]§
24		75, M	Japan	Negative¶	0.3	HCP	Relapse	CS†, CY	None	Improved	[15]
25	2011	63, M	Japan	Negative¶	4.0	Limited GPA	Relapse	CS, CY	None	Improved	[10]§
26		54, F	Korea	388 AAU	4.6	HCP	New onset	CS, CY	ND	Improved	[11]
27		76, F	Japan	Negative¶	2.8	MPA	Relapse	CS†	ND	Improved	[7]§
28		61, F	Japan	Positive*	1.4	HCP	New onset	CS†	ND	Improved	[7]§
29	2012	64, F	Japan	MPO, 40 IU/ml with PR3 positive	6.6	HCP	New onset, relapse	CS†, CY	None	Improved	[33]
30		75, F	Japan	Negative¶	9.3	GPA	Relapse	CS†, MZR	Active vasculitis	Improved	Present case

Abbreviations : CRP, C-reactive protein ; ESR, erythrocyte sedimentation rate (mm/h) ; EU, ELISA units ; AAU, an autoantibody titer ; HCP, hypertrophic cranial pachymeningitis (either p- or MPO-ANCA-related HCP) ; ND, not described in the literature ; GPA, granulomatosis with polyangiitis (Wegener's) ; CS, corticosteroid ; CY, cyclophosphamide ; AZA, azathioprine ; MZR, mizoribine ; MTX, methotrexate.
All references are searchable by PUBMED. (§) indicates the article is in Japanese with an abstract in both Japanese and English ; (†) means cases also treated with pulse corticosteroid therapy ; (♣) means p-ANCA-positive status as evaluated by the indirect immunofluorescence method ; (‡) means ANCA was measured as p-ANCA titer, not MPO-ANCA titer ; (*) means ANCA titer or CRP was not described ; (¶) means that ANCA was negative at the onset of hypertrophic cranial pachymeningitis.

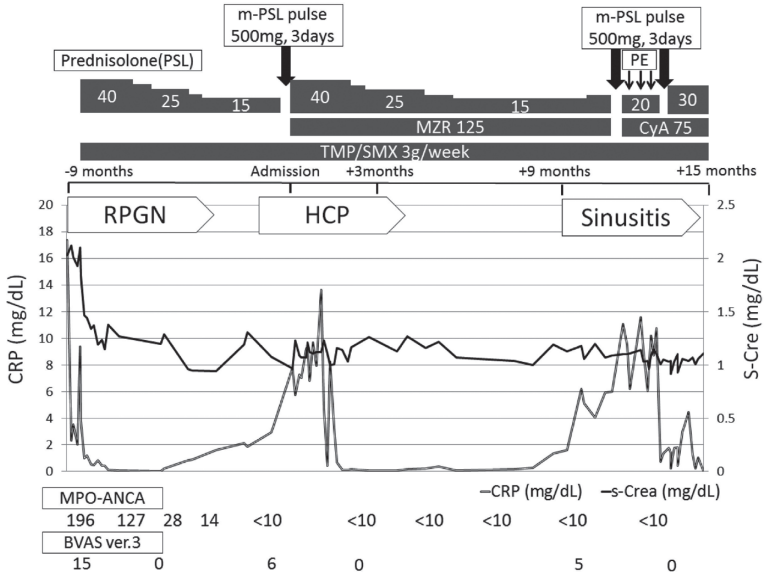

Fig. 3. Clinical course of the patient. m-PSL pulse, methylprednisolone pulse therapy for 3 days ; PE, plasma exchange ; MZR, mizoribine ; CyA, cyclosporine ; TMP/SMX, trimethoprim-sulfamethoxazole ; RPGN, rapidly progressive glomerulonephritis ; HCP, hypetrophic cranial pachymeningitis ; CRP, C-reactive protein ; S-Cre, serum creatinine ; MPO-ANCA, anti-myeloperoxidase anti-neutrophil cytoplasmic antibody ; BVAS ver.3, Birmingham Vasculitis Activity Score (version 3).

continued (Fig. 3).

CONCLUSION

We present a case in which HCP developed during successful treatment of MPO-ANCA-related RPGN. The patient was finally diagnosed with GPA. HCP is an important complication in MPO-ANCA-related vasculitis, and needs to be considered during the clinical course not only by specialists such as rheumatologists and neurologists, but also by other clinicians.

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The authors have no Conflicts of Interest (COI) to declare.

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