



Title	Hypertrophic cranial pachymeningitis in mpo-anca-related vasculitis: a case report and literature review
Author(s)	Watanabe, Kimio; Tani, Yoshihiro; Kimura, Hiroshi; Asai, Jun; Tanaka, Kenichi; Hayashi, Yoshimitsu; Asahi, Koichi; Nakayama, Masaaki; Watanabe, Tsuyoshi
Citation	Fukushima Journal of Medical Science. 59(1): 56-62
Issue Date	2013
URL	http://ir.fmu.ac.jp/dspace/handle/123456789/366
Rights	© 2013 The Fukushima Society of Medical Science
DOI	10.5387/fms.59.56
Text Version	publisher

This document is downloaded at: 2022-08-23T12:57:26Z

[Case Report]

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

KIMIO WATANABE, YOSHIHIRO TANI, HIROSHI KIMURA, JUN ASAI, KENICHI TANAKA, YOSHIMITSU HAYASHI, KOICHI ASAHI, MASAOKI NAKAYAMA and TSUYOSHI WATANABE

Department of Internal Medicine, Division of Nephrology, Hypertension, Endocrinology, and Diabetology/Metabolism, Fukushima Medical University School of Medicine, Fukushima, Japan

(Received November 26, 2012, accepted February 18, 2013)

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 life¹⁾. Several causes have been recognized, including infections (such as syphilis or tuberculosis), connective tissue diseases, neoplasms and vasculitides¹⁻³⁾.

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 course⁴⁻⁶⁾. 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 m²) 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)

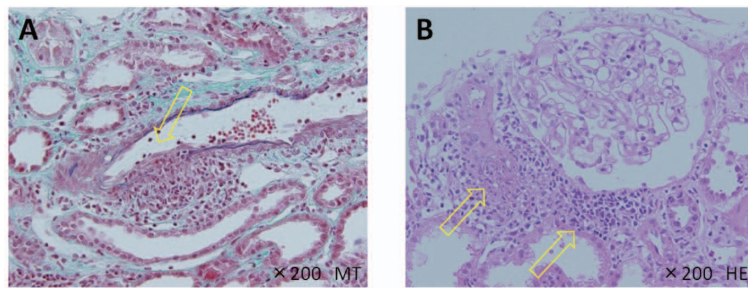


Fig. 1. Renal biopsy finding. Small-sized arteries are the main vessels affected by vasculitis in renal histology. A) Infiltration of inflammatory cells and destroyed vascular structures (yellow arrow). B) Small vessel vasculitis and inflammation spreading to adjacent glomeruli (yellow arrow). One of 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.

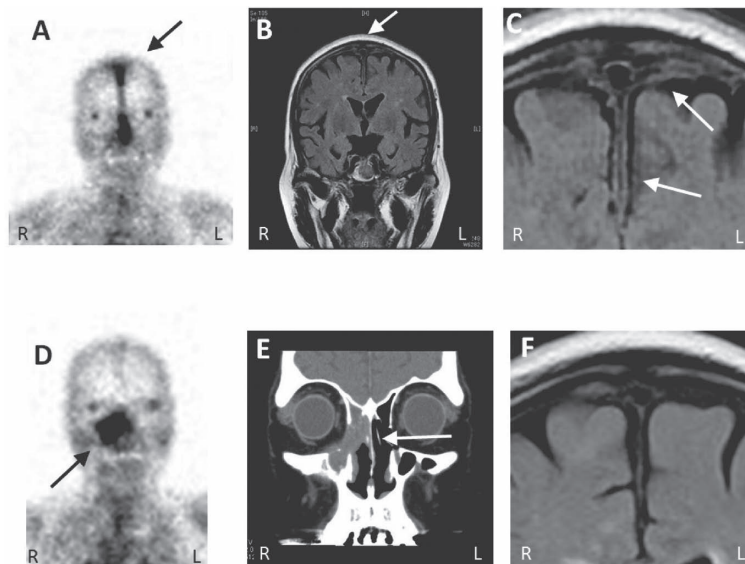


Fig. 2. Imaging findings. A) Gallium scintigraphy showing hypertrophic pachymeningitis (HCP). Dense accumulation is seen at the falx cerebri (black arrow). B) Magnetic resonance imaging (MRI) showing HCP. A slightly swollen falx cerebri is evident (white arrow). C) Enlargement of the thickened region of the 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. Accord-

ing 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 m²) 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/mm³; 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 m²). 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 criteria⁸. 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 HCP⁹, 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 course⁴⁻⁶. 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 imaging¹. 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 m²), use of MRI and Ga scintigraphy allowed successful diagnosis³⁶. 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 treatment¹. 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 Japan⁴⁰. 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	2000	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†	CS†	ND	Improved	[25]
9	56, F	Japan	321 EU	17.1	MPA	New onset	CS†	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	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 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.

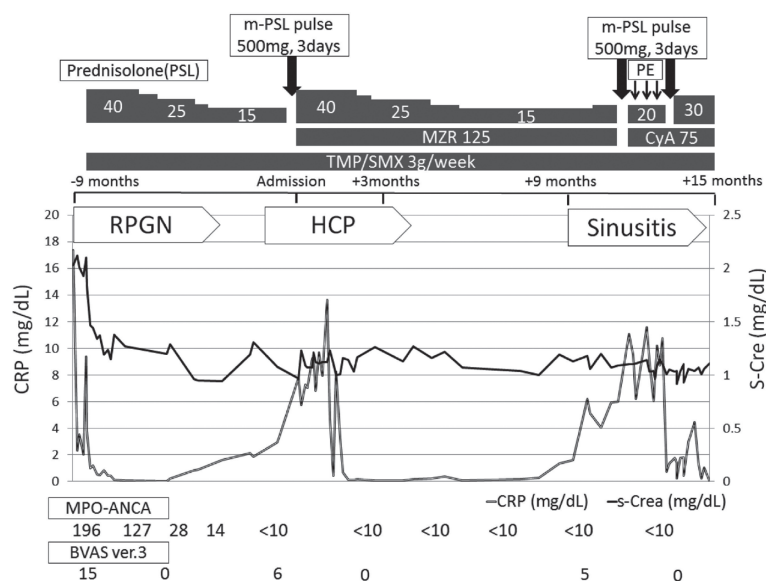


Fig. 3. Clinical course of the patient. m-PSL pulse, methylprednisolone pulse therapy for 3 days; PE, plasma exchange; MZR, mizoribine; CyA, cyclosporine; TMP/SMX, trimethoprim-sulfamethoxazole; RPGN, rapidly progressive glomerulonephritis; HCP, hypertrophic cranial pachymeningitis; CRP, C-reactive protein; S-Crea, 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.

ACKNOWLEDGMENTS

We would like to thank Yasushi Hasegawa, MD, Department of Radiology, Fukushima Medical University for analysis of image findings.

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

REFERENCES

- Mamelak AN, Kelly WM, Davis RL, Rosenblum ML. Idiopathic hypertrophic cranial pachymeningitis. Report of three cases. *J Neurosurg*, **79**: 270-276, 1993.
- Masson C, Henin D, Hauw JJ, Rey A, Raverdy P, Masson M. Cranial pachymeningitis of unknown origin: a study of seven cases. *Neurology*, **43**: 1329-1334, 1993.
- Nakamura T, Hirakawa K, Higashi S, *et al.* CD8+ T lymphocytes infiltrate predominantly in the inflammatory foci of MPO-ANCA-positive thoracic hypertrophic pachymeningitis in a patient with HLA-A24. *Mod Rheumatol*, **17**: 75-80, 2007.
- Kono H, Inokuma S, Nakayama H, Yamazaki J. Pachymeningitis in microscopic polyangiitis (MPA): a case report and a review of central nervous system involvement in MPA. *Clin Exp Rheumatol*, **18**: 397-400, 2000.
- Furukawa Y, Matsumoto Y, Yamada M. Hypertrophic pachymeningitis as an initial and cardinal manifestation of microscopic polyangiitis. *Neurology*, **63**: 1722-1724, 2004.
- Hayashi Y, Sugawara H, Otsuka M, Yamada S, Tabei K, Ueki A. Fatal hemoperitoneum preceded by cranial hypertrophic pachymeningitis in a patient with ANCA-positive microscopic polyangiitis. *Intern Med*, **47**: 1061-1063, 2008.
- Takahashi R, Oyamada T, Ozaki H, Hayashi H, Sakata N, Uchio E. Three cases of optic neuropathy associated with hypertrophic pachymeningitis. (in Japanese). *Nihon Ganka Gakkai Zasshi*, **115**: 602-610, 2011.
- Watts R, Lane S, Hanslik T, *et al.* Development and validation of a consensus methodology for the classification of the ANCA-associated vasculitides and polyarteritis nodosa for epidemiological studies. *Ann Rheum Dis*, **66**: 222-227, 2007.

9. Seror R, Mahr A, Ramanoelina J, Pagnoux C, Cohen P, Guillevin L. Central nervous system involvement in Wegener granulomatosis. *Medicine (Baltimore)*, **85** : 54-65, 2006.
10. Kurihara Y, Oku K, Suzuki A, Ohson Y, Handa M, Okano Y. A case of slowly progressive type 1 diabetes mellitus developing myeloperoxidase-specific anti-neutrophil cytoplasmic antibody-associated vasculitis with hypertrophic pachymeningitis manifesting as multiple cranial nerve palsy. *Nihon Rinsho Meneki Gakkai Kaishi (in Japanese)*, **34** : 510-515, 2011.
11. Lim EJ, Kim SH, Lee SH, Lee KY, Choi JH, Nam EJ. Reversible Sensorineural Hearing Loss due to Pachymeningitis Associated with Elevated Serum MPO-ANCA. *Clin Exp Otorhinolaryngol*, **4** : 155-158, 2011.
12. Miyazaki K, Fukuzumi M, Aizawa T, Shibata M, Sakai T, Tojima H. Patient with MPO-ANCA-associated disease with interstitial pneumonia and lower cranial nerves palsy who was previously exposed to asbestos. *Nihon Kokyuki Gakkai Zasshi (in Japanese)*, **48** : 876-882, 2010.
13. Salvi F, Mascacchi M, Pasini E, *et al.* p-ANCA pachymeningitis presenting with isolated "optic neuropathy". *Neurol Sci*, **31** : 639-641, 2010.
14. Horai Y, Miyamura T, Takahama S, *et al.* Refractory antineutrophil cytoplasmic antibody-associated vasculitis successfully treated with rituximab : a case report. *Nihon Rinsho Meneki Gakkai Kaishi*, **33** : 105-110, 2010.
15. Horino T, Takao T, Taniguchi Y, Terada Y. Hypertrophic pachymeningitis with MPO-ANCA-positive vasculitis. *Clin Rheumatol*, **29** : 111-113, 2010.
16. Yamazaki C, Arai S, Tamura Y, *et al.* Case of rapidly progressive glomerulonephritis with anti-glomerular basement membrane antibody in the course of MPO-ANCA-associated pachymeningitis. *Nihon Jinzo Gakkai Shi (in Japanese)*, **51** : 490-495, 2009.
17. Ogasawara T, Kasamatsu N, Haga T, Kobayashi T, Shibata M, Hashizume I. A case of Wegener's granulomatosis with orbital inflammatory pseudotumor. *Nihon Kokyuki Gakkai Zasshi (in Japanese)*, **47** : 308-313, 2009.
18. Abe T, Nogawa S, Tanahashi N, Shiraishi J, Ikeda E, Suzuki N. Cerebral pachyloptomeningitis associated with MPO-ANCA induced by PTU therapy. *Intern Med*, **46** : 247-250, 2007.
19. Jacobi D, Maillot F, Hommet C, *et al.* P-ANCA cranial pachymeningitis : a case report. *Clin Rheumatol*, **24** : 174-177, 2005.
20. Akahoshi M, Yoshimoto G, Nakashima H, *et al.* MPO-ANCA-positive Wegener's granulomatosis presenting with hypertrophic cranial pachymeningitis : case report and review of the literature. *Mod Rheumatol*, **14** : 179-183, 2004.
21. Saeki T, Fujita N, Kourakata H, Yamazaki H, Miyamura S. Two cases of hypertrophic pachymeningitis associated with myeloperoxidase anti-neutrophil cytoplasmic autoantibody (MPO-ANCA)-positive pulmonary silicosis in tunnel workers. *Clin Rheumatol*, **23** : 76-80, 2004.
22. Fam AG, Lavine E, Lee L, Perez-Ordóñez B, Goyal M. Cranial pachymeningitis : an unusual manifestation of Wegener's granulomatosis. *J Rheumatol*, **30** : 2070-2074, 2003.
23. Tajima Y, Miyazaki Y, Sudoh K, Matumoto A. Intracranial hypertrophic pachymeningitis with high perinuclear anti-neutrophil cytoplasmic antibody (p-ANCA) occurred in a patient on hemodialysis. *Rinsho Shinkeigaku (in Japanese)*, **42** : 243-246, 2002.
24. Funachi M, Yoo BS, Sugiyama M, *et al.* A case of rheumatoid meningitis positive for perinuclear antineutrophil cytoplasmic antibody. *Ann Rheum Dis*, **59** : 1001-1002, 2000.
25. Tamai H, Tamai K, Yuasa H. Pachymeningitis with pseudo-Foster Kennedy syndrome. *Am J Ophthalmol*, **130** : 535-537, 2000.
26. Takahashi K, Kobayashi S, Okada K, Yamaguchi S. Pachymeningitis with a perinuclear antineutrophil cytoplasmic antibody : response to pulse steroid. *Neurology*, **50** : 1190-1191, 1998.
27. Sasaki R, Taniguchi A, Narita Y, Naito Y, Kuzuhara S. A case of optic neuropathy, recurrent transverse myelopathy and hypertrophic pachymeningitis associated with perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA). *Rinsho Shinkeigaku (in Japanese)*, **35** : 513-515, 1995.
28. Ishikura T, Horikiri H, Umehara F, *et al.* Case of hypertrophic meningitis with vasculitis syndrome. *Nihon Naika Gakkai Zasshi (in Japanese)*, **83** : 1817-1819, 1994.
29. Shindo K, Nitta K, Nagasaka T, Shiozawa Z. Idiopathic hypertrophic cranial pachymeningitis associated with Horner's syndrome. A case report. *Rinsho Shinkeigaku (in Japanese)*, **37** : 300-303, 1997.
30. Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 9-1999. A 74-year-old woman with hydrocephalus and pleocytosis. *N Engl J Med*, **340** : 945-953, 1999.
31. Sugiyama Y, Shimizu M, Hoshi A, *et al.* An old man presenting with fluctuating bilateral multiple cranial nerve palsies and positive test for perinuclear antineutrophil cytoplasmic antibody. *No To Shinkei (in Japanese)*, **51** : 825-832, 1999.
32. Yoritaka A, Tsukamoto T, Ohta K, Kishida S. A clinical study of pachymeningitis. *No To Shinkei*

- (in Japanese), **54** : 235-240, 2002.
33. Iguchi A, Wada Y, Kobayashi D, *et al.* A case of MPO- and PR3-ANCA-positive hypertrophic cranial pachymeningitis with elevated serum IgG4. *Mod Rheumatol*, 2012 [In press].
 34. Nagashima T, Maguchi S, Terayama Y, *et al.* P-ANCA-positive Wegener's granulomatosis presenting with hypertrophic pachymeningitis and multiple cranial neuropathies : case report and review of literature. *Neuropathology*, **20** : 23-30, 2000.
 35. Xue J, Wang H, Wu H, Jin Q. Wegener's granulomatosis complicated by central diabetes insipidus and peripheral neutrophily with normal pituitary in a patient. *Rheumatol Int*, **29** : 1213-1217, 2009.
 36. Chrysochou C, Power A, Shurrah AE, *et al.* Low risk for nephrogenic systemic fibrosis in nondialysis patients who have chronic kidney disease and are investigated with gadolinium-enhanced magnetic resonance imaging. *Clin J Am Soc Nephrol*, **5** : 484-489, 2010.
 37. Rudnik A, Larysz D, Gamrot J, Skorupa A, Bierzynska-Macyszyn G, Bazowski P. Idiopathic hypertrophic pachymeningitis - case report and literature review. *Folia Neuropathol*, **45** : 36-42, 2007.
 38. Bosman T, Simonin C, Launay D, Caron S, Destee A, Defebvre L. Idiopathic hypertrophic cranial pachymeningitis treated by oral methotrexate : a case report and review of literature. *Rheumatol Int*, **28** : 713-718, 2008.
 39. Takuma H, Shimada H, Inoue Y, *et al.* Hypertrophic pachymeningitis with anti-neutrophil cytoplasmic antibody (p-ANCA), and diabetes insipidus. *Acta Neurol Scand*, **104** : 397-401, 2001.
 40. Guidelines for the treatment of rapidly progressive glomerulonephritis, second version. *Nihon Jinzo Gakkai Shi* (in Japanese), **53** : 509-555, 2011.