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Distinguishing magnetic resonance imaging features between idiopathic hypertrophic pachymeningitis and secondary hypertrophic pachymeningitis

Saksith Warittikoon, Ketsuda Jakchairoongruang*

Abstract

Background: Hypertrophic pachymeningitis (HP) is a rare chronic inflammatory disorder characterized by marked fibrous thickening of the cerebral and/or spinal dura mater. This condition is caused by infection, inflammation, autoimmune disorder, neoplasms, or idiopathic. Magnetic resonance imaging (MRI) may play an important role in differentiating idiopathic HP from secondary HP, may avoid unnecessarily invasive dural biopsy, and prompt specific treatment.
Objective: To determine the specific MRI findings for differentiation between idiopathic HP and secondary HP.
Method: A total of 34 patients underwent MRI of the brain and cervical spine from January 2003 to December 2015. In all, 23 patients were diagnosed idiopathic HP and 11 patients were secondary HP. Demographic data and imaging findings reveal the following: configuration, thickness, signal intensity on T1-weighted image (T1WI), T2-weighted image (T2WI), and enhancement pattern of the lesions. The data were analyzed by T-test and Fisher's exact test.
Result: Secondary HP were significantly located at anterior and middle cranial fossa ($P = 0.033$). There is no significant difference of lesions in configurations, T1 and T2 signal intensity and patterns of enhancement. There was significant and exclusive difference in T2 hypointense/dark intensity and homogeneous enhancement in idiopathic HP (75%, $P = 0.044$).
Conclusions: MRI may play a complimentarily important role in distinguishing idiopathic HP from secondary HP. Idiopathic HP is probably preferred diagnosis in the lesions with T2-rim pattern and T2 hypointense/dark intensity with homogeneous enhancement.

Keywords: dural inflammation; dural thickening; thickened dura; hypertrophic pachymeningitis; idiopathic hypertrophic pachymeningitis

Hypertrophic pachymeningitis (HP) is a rare chronic inflammatory disorder characterized by marked fibrous thickening of the cerebral and/or spinal dura mater. Neurological symptoms are non-specific. Patients commonly present with longstanding headache and/or progressive neurologic deficits, multiple cranial nerve palsies, or blindness [1]. Several etiologies have been recognized including infections (such as syphilis and tuberculosis), autoimmune disorders (such as IgG4-related disease, rheumatoid arthritis), vasculitis (such as Wegener's granulomatosis), granulomatous inflammation (such as

***Correspondence to: Ketsuda Jakchairoongruang,** Department of Radiology, Faculty of Medicine, Chulalongkorn University, King Chulalongkorn Memorial Hospital, The Thai Red Cross Society, Bangkok 10330, Thailand, e-mail: ketsuda.ja@chula.ac.th
Department of Radiology, Faculty of Medicine, Chulalongkorn University, King Chulalongkorn Memorial Hospital, The Thai Red Cross Society, Bangkok 10330, Thailand

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sarcoidosis), and neoplasms (such as lymphoma or metastasis) [2–5]. Blood serology, CSF examination and cytology, and/or dural biopsy for histopathology are required for determining the etiology. If there is no identifiable cause, the condition is referred to "idiopathic HP."

Idiopathic HP is a diagnosis of exclusion. Dural biopsy is definite diagnosis of idiopathic HP. Pathological findings of idiopathic HP are non-specific chronic inflammation, showing infiltration of lymphocytes, plasma cells, and fibrotic dura [6]. Corticosteroid therapy is an effective treatment in alleviating the symptoms and arresting the progression of idiopathic HP.

To date, there are a few studies regarding magnetic resonance imaging (MRI) findings of idiopathic HP, secondary HP, and comparison between those two entities. This study aims to describe the imaging findings on MRI and to determine the distinguished MRI features of idiopathic HP and secondary HP, which could be a complimentary tool to differentiate these two entities and eventually reduce invasive dural biopsy for diagnosis.

Materials and Methods

Patients

In total, 34 patients were retrospectively collected using searching tools in the picture archiving and communication system (PACS) and hospital information system (HIS) of King Chulalongkorn Memorial Hospital, The Thai Red Cross Society from January 1, 2003 to December 31, 2015. Searching terms included pachymeningitis, dural thickening, thickened dura, and hypertrophic pachymeningitis. Patients who did not have definite diagnosis by laboratory or histopathology, and patients with probable diagnosis by therapeutic diagnosis, who did not have follow-up MRI, were excluded. Patients with dural thickening caused by other entities including intracranial hypotension, post-surgical changes, or dural-based bulky mass were excluded. Our study was approved by the King Chulalongkorn Memorial Hospital Institutional Ethics Committee (certificate of approval no. 754/2015). Informed consent was waived due to retrospective nature of the study.

Imaging analysis

We collected the image data from MRI Signa Excite 1.5T (General Electric Healthcare, USA), MAGNETOM Aera 1.5T (Siemens Healthcare, Germany), Ingenia 1.5T (Philips Healthcare, Netherlands) and Achieva 3.0T (Philips Healthcare, Netherlands). Imaging comparison included T1-weighted image (T1WI), T2-weighted image (T2WI), diffusion-weighted image (DWI), and gadolinium-enhanced T1WI in all available planes for the data collection and analysis. DWI was not acquired in 11 of 23 idiopathic HP patients and 1 of 11 secondary HP patients.

Dural thickening was defined as maximal dural thickness more than 3 mm of any plane. We collected imaging characteristics of dural thickening: locations (supratentorial, infratentorial, diffuse, and spinal canal; **Table 1**), configuration (linear/smooth and nodular/undulated), signal intensity comparing to gray matter (hyperintensity, iso-intensity, hypointensity, and dark intensity (equal or less than CSF on T1WI, and equal or less than cortical bone intensity on T2WI), presence of rim pattern (defined as different intensity of the central part and the periphery of the lesions), presence of restricted diffusion, and pattern of enhancement (homogeneous, heterogeneous, and rim).

MR images were individually and blindly reviewed by one diagnostic radiology resident (SW) and one 2-year experienced neuroradiologists (KJ). Disagreement of imaging interpretation was discussed and consensus.

Statistical analysis

Demographic data, clinical presentation, and each imaging characteristics of each group were described using mean, range, and percentage. Maximal thickness of lesions between two groups was analyzed using T-test. Imaging characteristics between two groups were analyzed using Fisher's exact test. $P < 0.05$ was considered statistically significant. Inter-observer reliability was determined using Kappa statistics. Statistical analysis was performed using SPSS 17.0 for Windows (SPSS, Chicago, IL, USA).

Table 1. Location of hypertrophic pachymeningitis

Supratentorial	Cavernous sinus
	Orbital apex
	Superior orbital fissure
	Sphenoid wing
	Cerebral convexity
	Tentorium cerebelli
	Falx cerebri
	Anterior and middle fossae
Infratentorial	Posterior fossa
	Foramen magnum
Diffuse	Throughout the calvarium
Spinal canal	Spinal canal

Results

Study population

Of 34 patients, 23 patients (68%) were idiopathic HP (8 were men, 15 were women with mean age of 46 years (range: 20–68 years). Eight patients had histopathological diagnosis. In all, 15 patients were diagnosed by therapeutic diagnosis. Eleven of 34 patients (32% were classified as secondary HP (4 were men, 7 were women with mean age of 55 years (range: 8–9 years). Secondary HP group included patients who were diagnosed IgG4-related disease, dural metastasis (from prostate cancer), primary dural lymphoma, acute promyelocytic leukemia, nasopharyngeal carcinoma with dural involvement, HIV infection with cryptococcal meningitis, HIV infection with TB meningitis, cavernous sinus thrombosis secondary to orbital cellulitis, meningioma, mastoiditis, and chronic sinusitis. Six of 11 patients had histopathological diagnosis, and 5 of 11 patients were diagnosed by therapeutic diagnosis.

There was no significant difference of age and sex between two groups (**Table 2**). Headache was present in 15 of 23 (65%) idiopathic HP patients and 2 of 11 (18%) secondary HP patients ($P = 0.026$). There was no significant difference between duration of headache and presence of neurological deficit. Neurologic deficits were presented in 20 idiopathic HP patients (87%), in 9 secondary HP patients (82%), and were mainly cranial nerve(s) palsy. Optic nerve involvement was the most common neurological deficit in idiopathic HP patients, whereas oculomotor nerve palsy was the most common neurological deficit in secondary HP patients (**Table 2**). Other neurological symptoms were both arms weakness and left hemiparesis.

Table 2. Demographic data and clinical presentations of idiopathic HP and secondary HP patients

	IHP n (%)	Secondary HP n (%)	*P*
Number of patients	23 (68)	11 (32)	
Age (mean, range) (years)	46 (20–68)	55 (8–79)	0.235
Male	8 (35)	4 (36)	1.00
Female	15 (65)	7 (64)	
Neurological symptoms			
Headache	15 (65)	2(18)	*0.026**
			1.00
Duration (mean, range) (months)	6.6 (0.1–120)	0.5 (1.5–4.0)	
Neurological deficits	20 (87)	9 (82)	
- CN I	0	1 (9)	
- CN II	9 (39)	2 (18)	
- CN III	8 (35)	3 (27)	
- CN IV	5 (22)	1 (9)	
- CN V	6 (26)	1 (9)	
- CN VI	6 (26)	0	
- CN VII	1 (4)	3 (27)	
- Others (both arms weakness and left hemiparesis)	1 (4)	2 (18)	

**P* by Fisher's exact test

MRI findings

In idiopathic HP patients, there were 12 patients (52%) with linear/smooth dural thickening configuration (**Figure 1**) and 11 patients (48%) with nodular/undulated dural thickening. In secondary HP patients, there were 6 patients (55%) with linear/smooth dural thickening and five patients (45%) with nodular/undulated dural thickening (**Figure 2**). There was no significant difference in configurations of dural thickening between 2 groups.

Most of dural thickening involved supratentorium in both groups (96% in idiopathic HP group, and all patients in secondary HP group). Secondary HP group significantly had dural thickening at anterior and middle cranial fossae (55%) (**Figure 3**) as compared to idiopathic HP patients (13%), $P = 0.033$. There was no secondary HP patient having posterior fossa dural thickening (**Table 3**).

There was no difference of mean value of maximal dural thickness between idiopathic HP group (5.9 mm) and secondary HP group (5.8 mm) with $P = 0.928$. There was no significant difference of signal intensity on T1WI, T2WI, and enhancement pattern, and presence of restricted diffusion between two groups.

Three of 23 idiopathic HP patients (13%) showed central hyperintensity and dark intensity rim on T2WI, which was defined as T2-rim pattern (**Figure 4**). The T2-rim pattern was not found in secondary HP group. Details of MRI findings are in **Table 4**.

There is significantly found T2 hypointensity and homogeneous enhancement in idiopathic HP (80%) (**Figures 1 and 4**). None of secondary HP patients had these findings, $P = 0.014$.

The inter-observer reliability was almost perfect for lesions configuration (k = 1.00), T1 signal intensity (k = 0.879), T2 signal intensity (k = 0.810), and enhancement pattern (k = 0.836).

Discussion

Idiopathic HP can be presented with diffuse and localized dural thickening [7–9]. In our study, we found idiopathic HP

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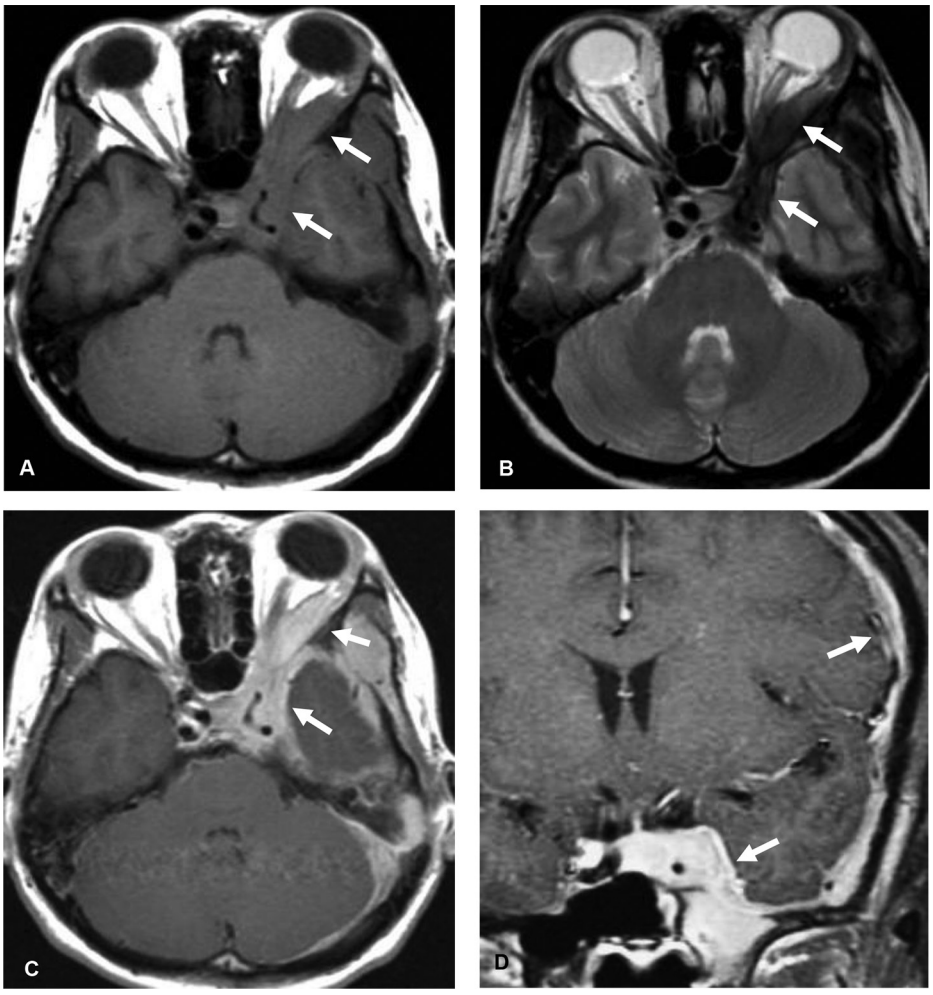

Figure 1. Idiopathic hypertrophic pachymeningitis (IHP). Axial T1-weighted image (**A**), axial T2-weighted image (**B**) shows an infiltrative T1 iso-intense, T2 hypointense lesion (arrow) involving left cavernous sinus with extension through left superior orbital fissure to left orbit, along left sphenoid ridge, left middle and posterior cranial fossae. Homogeneous enhancement is shown in axial (**C**) and coronal (**D**) gadolinium-enhanced T1-weighted images.

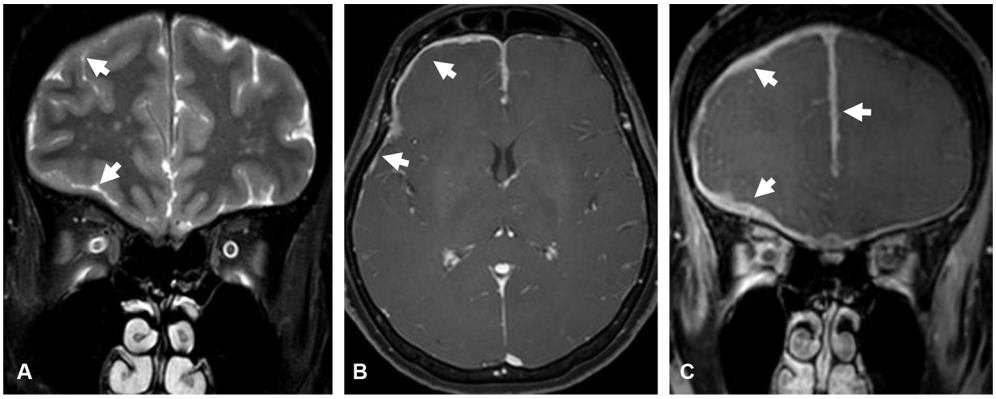

Figure 2. Primary dural lymphoma. Coronal T2-weighted image (**A**) shows hyperintense undulated thickened dura along the right cerebral convexity, anterior cranial fossa, and anterior falx cerebri (arrow). Axial (**B**) and coronal (**C**) gadolinium-enhanced T1-weighted images show homogeneously enhanced dural thickening.

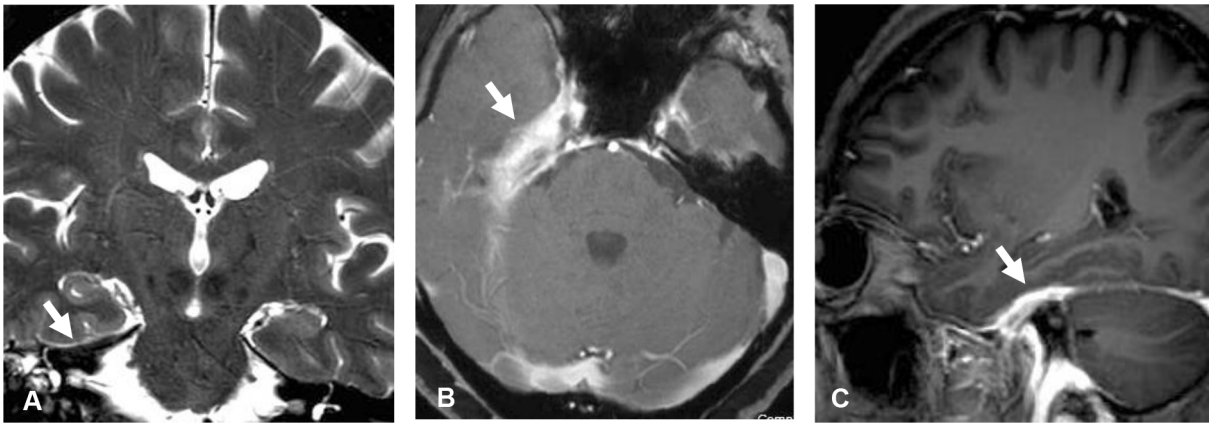

Figure 3. IgG4-related pachymeningitis. Coronal T2-weighted image (**A**) shows dark intensity dural thickening involving right tentorium cerebelli (arrow). Axial (**B**) and sagittal (**C**) gadolinium-enhanced T1-weighted images demonstrate mildly heterogeneous enhancement of the thickened dura involving right medial temporal region, right Meckel's cave, and along anterior part of right tentorium cerebelli (arrow).

Table 3. Configurations and locations of dural thickening in idiopathic HP and secondary HP groups

Characteristics	Idiopathic HP (N = 23) n (%)	Secondary HP (N = 11) n (%)	*P*
Configurations			
Linear/smooth	12 (52)	6 (55)	1.00
Nodular/undulated	11 (48)	5 (45)	1.00
Locations			
Supratentorial lesions	22 (96)	11 (100)	1.00
Cavernous sinus	13 (57)	6 (55)	1.00
Orbital apex	4 (17)	4 (36)	0.388
Supraorbital foramen	1 (4)	0	1.00
Sphenoid wing	2 (9)	2 (18)	0.58
Cerebral convexity	8 (35)	7 (64)	0.151
Tentorium cerebelli	9 (39)	5 (46)	1.00
Falx cerebri	4 (17)	2 (18)	1.00
Anterior and middle cranial fossae	3 (13)	6 (55)	*0.033**
Infratentorial lesions	6 (26)	0	0.145
Posterior fossa	6 (26)	0	0.145
Foramen magnum	0	0	
Diffuse	6 (26)	0	0.145
Spinal canal	1 (4)	0	1.00

Table 4. Signal intensity of dural thickening in idiopathic HP and secondary HP groups

Signal intensity	IHP (N = 23) n (%)	Secondary HP (N = 11) n (%)	*P*
T1-weighted image	15 (65)	9 (82)	0.784
Iso-intensity	7 (30)	2 (18)	
Hypo-intensity and dark intensity	1 (4)	0	
Hyperintensity			
T2-weighted image	5 (22)	3 (27)	0.148
Iso-intensity	12 (52)	3 (27)	
Hypo-intensity and dark intensity	3 (13)	5 (46)	
Hyperintensity	3 (13)	0	
T2-rim pattern			
Diffusion-weighted image	12	10	0.455
Restricted diffusion	0	1 (10)	
No restricted diffusion	12 (100)	9 (90)	
Enhancement pattern	16 (70)	5 (46)	0.344
Homogeneous	5 (22)	5 (46)	
Heterogeneous	2 (9)	1 (9)	
Rim			
T2 hypo-/dark intensity with	12	3	*0.014**
Homogeneous enhancement	9 (75)	0	
Heterogeneous enhancement	2 (17)	3 (100)	
Rim enhancement	1 (8)	0	

**P* by Fisher's exact test

in both diffuse and localized forms. The predominant pattern was the localized dural thickening (74%).

Our study found that idiopathic HP mostly showed variable signal intensity on T1WIs, which were iso-intense in 65% and hypointense in 30% of patients, which were described in the previous study [7].

Prior studies postulated characteristic imaging findings of idiopathic HP including marked enhancement of the dural edges on gadolinium-enhanced T1WIs [10], hypointense area with thin hyperintense edges on T2WIs [10, 11], markedly hypointense on T2-weighted [9]. Dense cellularity and collagenous material or fibrosis are associated with hypointensity on T2WI. Hypervascularity at the dural margins may result in thin hyperintensity at the dural edges [9–11].

Our study found T2 hypo-/dark intensity in majority of idiopathic HP (52%) and in minority of secondary HP

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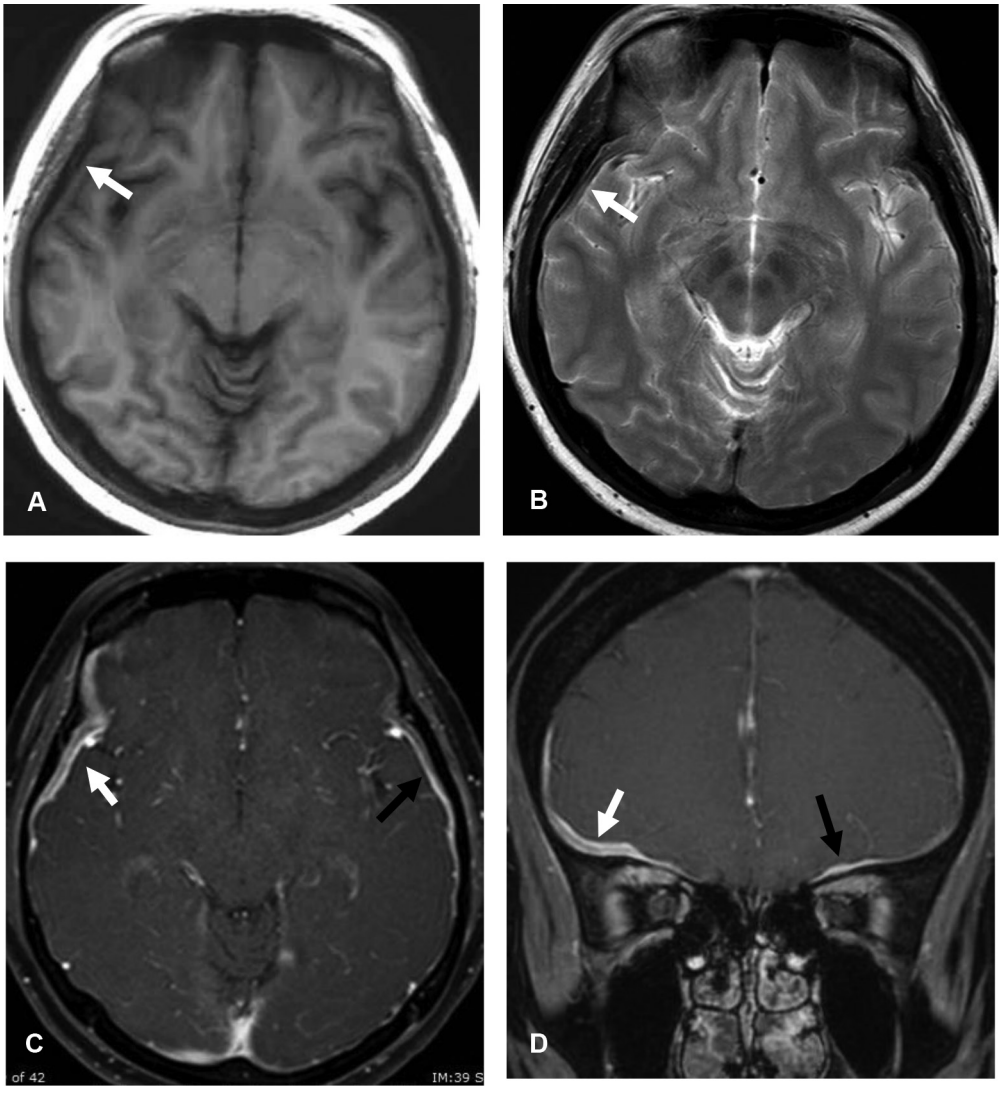

Figure 4. Idiopathic hypertrophic pachymeningitis (IHP) with T2-rim pattern. Axial T1-weighted image (**A**) and T2-weighted image (**B**) show T1 isointense and central T2 hyperintense with peripheral dark T2 intense (T2-rim pattern) of the thickened dura along right frontal and right temporal convexity (arrow). Axial (**C**) and coronal (**D**) gadolinium-enhanced T1-weighted images demonstrate rim enhancement (white arrows). Smooth and homogeneous enhanced thickened dura at left anterior cranial fossa (black arrows) is also observed.

(27%). However, our patients did not exhibit hypointensity with thin hyperintense edges on T2WIs as shown in prior studies [10, 11]. Interestingly, three idiopathic HP patients (13%) had thick hyperintense dura with hypointense rim on T2WIs (T2-rim pattern), which was described in the prior study [12]. None of secondary HP showed T2-rim pattern. This finding is promising to be MRI characteristics of idiopathic HP. Some of our patients (13%) showed diffuse T2 hyperintensity of the thickened dura. This finding was also described in the previous study and probably reflects vessels and inflammatory cells in the dural tissue [9–11]. There was no significant T2 intensity difference between idiopathic HP and secondary HP. Different signal intensity may represent different stages of disease with variable fibrosis, collagen, and inflammatory cells.

Majority of idiopathic HP showed homogeneous enhancement, which was well described in prior studies [9, 13, 14]. Prior studies showed enhancement of dural edges in idiopathic pachymeningitis [9, 10]. We found this enhancement pattern in two idiopathic HP and one secondary HP patients. Hypointense/dark SI on T2WIs with homogeneous enhancement pattern were found only in idiopathic HP and reached statistical significance (75% vs 0%, $P = 0.044$). These findings may raise possibility of idiopathic HP.

Major limitations of our study were small number of patients and lack of histopathologic examination in some patients. Future study with larger number of patients may clarify the promising imaging characteristics found in this study.

Conclusion

MRI plays an important role for evaluation of patients with dural thickening. Imaging characteristics of idiopathic HP are T2 hypointensity or dark intensity and enhancement of the dural edge. Central T2 hyperintensity with hypointense rim (T2-rim pattern) is one of the promising MRI findings in idiopathic HP.

Author contributions. KJ contributed to the conception and design of this study. Both authors contributed substantially to the acquisition of data. KJ analyzed and interpreted the data. SW drafted the manuscript and KJ contributed substantially to its critical revision. Both authors approved the final version submitted for publication and take responsibility for the statements made in the published article.

Conflict of interest statement. The authors have completed and submitted the International Committee of Medical Journal Editors Uniform Disclosure Form for Potential Conflicts of Interest. None of the authors disclose any conflict of interest.

References

[1] Goyal M, Malik A, Mishra NK, Gaikwad SB. Idiopathic hypertrophic pachymeningitis: spectrum of the disease. Neuroradiology. 1997; 39:619–23.

[2] Gunia S, Ecke T, Wohlfarth B, Koch S, Erbersdobler A. Dural metastases from disseminated prostate cancer clinically mimicking a benign reactive condition of the dura: case report and review of the literature. Urol Int. 2011; 86:239–41.

[3] Yuh WT, Drew JM, Rizzo M, Ryals TJ, Sato Y, Bell WE. Evaluation of pachymeningitis by contrast-enhanced MR imaging in a patient with rheumatoid disease. AJNR Am J Neuroradiol. 1990; 11:1247–8.

[4] Zelasko S, Hollingshead M, Castillo M, Bouldin TW. CT and MR imaging of progressive dural involvement by nephrogenic systemic fibrosis. AJNR Am J Neuroradiol. 2008; 29:1880–2.

[5] Wallace ZS, Carruthers MN, Khosroshahi A, Carruthers R, Shinagare S, Stemmer-Rachamimov A, et al. IgG4-related disease and hypertrophic pachymeningitis. Medicine (Baltimore). 2013; 92:206–16.

[6] Roongpiboonsopit D, Phanthumchinda K. Idiopathic hypertrophic pachymeningitis at King Chulalongkorn Memorial Hospital. J Med Assoc Thai. 2014; 97:374–80.

[7] Hatano NMD, Behari SMD, Nagatani TMD, Kimura MMD, Ooka KMD, Saito KMD, et al. Idiopathic hypertrophic cranial pachymeningitis: clinicoradiological spectrum and therapeutic options. Neurosurgery. 1999; 45:1336.

[8] Kupersmith MJ, Martin V, Heller G, Shah A, Mitnick HJ. Idiopathic hypertrophic pachymeningitis. Neuropathology. 2004; 62:686–94.

[9] Riku S, Kato S. Idiopathic hypertrophic pachymeningitis. Neuropathology. 2003; 23:335–44.

[10] Mamelak AN, Kelly WM, Davis RL, Rosenblum ML. Idiopathic hypertrophic cranial pachymeningitis. J Neurosurg. 1993; 79:270–6.

[11] Martin N, Masson C, Henin D, Mompoint D, Marsault C, Nahum H. Hypertrophic cranial pachymeningitis: assessment with CT and MR imaging. AJNR Am J Neuroradiol. 1989; 10:477–84.

[12] Keshavaraj A, Gamage R, Jayaweera G, Gooneratne IK. Idiopathic hypertrophic pachymeningitis presenting with a superficial soft tissue mass. J Neurosci Rural Pract. 2012; 3:193–5.

[13] Nishioka H, Ito H, Haraoka J, Takahashi M, Shinmura F. Idiopathic hypertrophic cranial pachymeningitis of the cavernous sinus mimicking lymphocytic hypophysitis. Neurol Med Chir (Tokyo). 1998; 38:377–82.

[14] Zhao. M, Geng. T, Qiao. L, Shi. J, Xie. J, Huang. F, et al. Idiopathic hypertrophic pachymeningitis: clinical, laboratory and neuroradiologic features in China. J Clin Neurosci. 2014; 21:1127–32.