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Clinical Study

Idiopathic hypertrophic pachymeningitis: Clinical, laboratory and neuroradiologic features in China

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ABSTRACT

Hypertrophic pachymeningitis is a rare chronic inflammatory disorder characterized by marked fibrous thickening of the cerebral and/or spinal dura mater. Clinical, laboratory, neuroradiologic and therapeutic data from 12 patients with idiopathic hypertrophic pachymeningitis (IHP) from our department were retrospectively studied. There were four men and eight women with a mean age of 49 ± 15.3 years, and more than half of the patients (58%) were aged 40–60 years. Headache was the most common symptom, occurring in 92% of patients. Headache improved markedly and rapidly after glucocorticoid treatment. Optic nerve involvement was noted in seven patients (58%). C-reactive protein levels increased in 80% and the erythrocyte sedimentation rate increased in 71% of patients. Three patients were positive for autoantibodies, including antinuclear antibodies (ANA), perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA), anti-cardiolipin antibodies (ACA) and rheumatoid factor (RF). Cerebrospinal fluid showed inflammatory changes, and protein levels were low to moderately elevated. MRI revealed a thickened dura in all patients, and five patients (42%) were diagnosed with sinus stenosis/occlusion. IHP is a chronic inflammatory disorder of the dura with three groups of symptoms, namely headache, cranial nerve palsy and symptoms due to sinus stenosis/occlusion. However, IHP has different features in China in that it predominantly affects women and the age of onset is younger. Sinus stenosis/occlusion is relatively common in IHP patients in China.

1. Introduction

Hypertrophic pachymeningitis (HP) is a rare chronic inflammatory disorder characterized by marked fibrous thickening of the cerebral and/or spinal dura mater, with a clinical presentation including headache, cranial neuropathies, ataxia, seizures and myelopathy, occurring either alone or in combination [1]. HP is divided into two types: idiopathic hypertrophic pachymeningitis (IHP), for which no identifiable cause has been found, and secondary HP, for which there are identifiable coexisting causes, including infections (including syphilis [2] and tuberculosis [3,4]), autoimmune disorders (rheumatoid arthritis and vasculitis, such as Wegener's granuloma) [5–8], sarcoidosis [9,10] and neoplasms [11–13]. Thus, the diagnosis of IHP is one of exclusion [14]. The pathogenic mechanisms underlying IHP are unknown, but the condition is considered to be an autoimmune disorder [15]. We present 12 patients with IHP from our department and report their clinical features, significant laboratory tests, radiological findings and therapeutic data.

2. Patients and methods

Clinical, laboratory, neuroradiologic and therapeutic data from 12 patients were retrospectively studied (Table 1). All patients underwent whole blood cell analysis, routine urinalysis, routine stool testing, occult stool blood testing, biochemical investigations, human immunodeficiency virus serology, serum venereal disease reference laboratory testing and imaging studies including a chest radiograph and gadopentate-enhanced MRI of the brain. Tests for autoantibodies, including rheumatoid factor (RF), antinuclear antibodies (ANA), anti-double stranded DNA antibody (anti-dsDNA), anti-cardiolipin antibodies (ACA), anti-neutrophil cytoplasmic antibodies (ANCA) including perinuclear ANCA (p-ANCA) and cytoplasmic ANCA, anti-proliferating cell nuclear antigen, anti-Sm, anti-Jo-1, anti-Scl-70, anti-SSA, anti-SSB and anti-UIRNP, and for tumor

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Table 1
Clinical, laboratory, neuroradiologic and therapeutic data from 12 Chinese patients with idiopathic hypertrophic pachymeningitis

Patient	Age at onset, sex	Duration (months)	Clinical features	Laboratory data	CSF WBC	CSF Pro.	MRI results	Sinus	Treatment	Headache improved
1	44, F	5	Headache, AGS	ESR 127 CRP 5.96	0	0.38	Diffuse dural thickening	DSA: stenosis	DXM Warfarin	2 days
2	49, M	360	Headache	CRP <0.3	0	0.67	Diffuse dural thickening	—	DXM	3 days
3	37, F	2	Headache, low fever	ESR 107 CRP 2.34 N 73% Hb 93	2	0.21	Diffuse dural thickening	MRV (−)	DXM Levofloxacin	3 days
4	61, F	16	Headache, CN II (left)	ESR 81 CRP 6.11 ANA (+) p-ANCA (+)	10	0.82	Diffuse dural thickening	—	MP AZA	3 days
5	40, F	96	Headache, CN II Epilepsy	ESR 14 CRP 1.58 RF (+)	0	0.86	Diffuse dural thickening, venous infarction	MRV, DSA: occlusion	DXM LMH	5 days
6	41, M	22	Headache, CN II Hemihyp-esthesia	ESR 59 CRP 3.52	5	0.42	Diffuse dural thickening, venous infarction	MRV: occlusion	MP AZA Urokinase LMH	4 days
7	55, M	5	CN II, eye pain	—	0	0.48	Diffuse dural thickening	—	MP	Eye pain 3 days
8	50, M	3	Headache, CN II	CRP <0.3 ANA (+) at 1:320 ACA (+)	100	0.99	Localized dural thickening	—	MP	1–2 weeks
9	17, F	3	Headache, CN II	CRP 0.1 ESR 4	5	0.43	Diffuse dural thickening	DSA: stenosis	MP	1 week
10	73, F	2	Headache, CN II, III, IV, V, VI	ESR 71 CRP 2.58	0	0.68	Diffuse dural thickening	—	DXM	2 days
11	71, F	12	Headache, lethargy	CRP 1.16	44	0.71	Diffuse dural thickening	MRV: stenosis	DXM Urokinase, warfarin	4 days
12	51, F	144	Headache, ataxia	—	—	—	Localized dural thickening	—	Surgical removal	—

AGS = amenorrhea galactorrhea syndrome, ANA = antinuclear antibodies, AZA = azathioprine, CN = cranial nerve, CRP = C-reactive protein (mg/dL), CSF = cerebrospinal fluid, DSA = digital subtraction angiography, DXM = dexamethasone, ESR = erythrocyte sedimentation rate (mm/L/hour), F = female, Hb = hemoglobin, IHP = idiopathic hypertrophic pachymeningitis, LMH = low molecular heparin, M = male, MP = methylprednisolone, MRV = magnetic resonance venography, N = neutrophils ratio, p-ANCA = perinuclear anti-neutrophil cytoplasmic antibodies, Pro = protein (g/L), RF = rheumatoid factor, WBC = white blood cell ($\times 10^6$/L), (+) = positive, (−) = negative, — = not performed.