Idiopathic Hypertrophic Pachymeningitis at King Chulalongkorn Memorial Hospital

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Background: *Idiopathic Hypertrophic Pachymeningitis (IHP) is a rare chronic inflammatory disorder of the dura. Classic clinical symptoms include headaches and cranial neuropathy. Because of scarce clinical data from Thailand, the present study aimed to determine the clinical features, neuroimaging findings, natural histories, therapeutic options, and outcomes for treatment of IHP in a tertiary care center.*

Material and Method: *A retrospective study was carried out on all adult IHP patients hospitalized at King Chulalongkorn Memorial Hospital, Bangkok, Thailand, between January 2000 and November 2011. Diagnostic criteria included 1) clinical symptom compatibility with IHP, 2) neuroimaging to reveal enhanced hypertrophic dura compatible with clinical syndrome, and 3) ruled out secondary causes of IHP, using appropriate clinical profiles and investigations including tissue biopsy.*

Results: *Thirty-two patients were enrolled with 21 females and 11 males, mean age of 49.03±16.12 years. The two most common symptoms were headache (93.8%) and diplopia (43.8%). The most common neurological finding was multiple cranial neuropathies (84.4%). Cranial nerve III was affected in 56.3% of the patients, followed by other cranial nerves including CN VI, IV, V, and II. Headache without a neurological deficit was observed in 12.5% of the cases. Focal and diffuse enhanced thickening of the dura were observed in 96.9% and 3.1% of the cases respectively. Focal thickening in the supratentorium included the cavernous sinus, orbital apex, sphenoid wing, and superior orbital fissure. Focal thickening in the infratentorium included the falx cerebelli, the dura at the base of the skull, Meckel's cave, and foramen magnum. CSF examination showed lymphocyte pleocytosis with a slight increase in CSF proteins. Headache subsided in all of the patients after treatment with corticosteroid. In relapsing and recurrent patients, a combined treatment of steroids and azathioprine was prescribed. With the combined treatment, clinical complete recovery, relapsing and recurrence were detected in 40%, 40% and 20% of the cases respectively. All relapsing and recurrence were due to rapid tapering off or early discontinuation of the steroids treatment. Only one patient had a spontaneous remission.*

Conclusion: *The most common clinical manifestations of IHP were headache and multiple cranial nerve involvement. Almost all of the patients had good initial response to steroid therapy. Relapse or recurrence was usually caused by rapid tapering off or early discontinuation of the steroid treatment. Long-term treatment with combined immunosuppression may be necessary in some cases.*

Keywords: *Idiopathic hypertrophic pachymeningitis, Cranial neurophathies, Tolosa Hunt syndrome, Polyneuritis cranialis*

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Idiopathic Hypertrophic Pachymeningitis (IHP) is a rare local or diffuse chronic nonspecific inflammatory disorder of the cranial and spinal cord dura mater. Pathologically, IHP is characterized by fibrosis and thickening of the dura[1]. Headache with multiple cranial neuropathy including Tolosa-Hunt syndrome are the most common clinical manifestations. Other neurological deficits such as cerebellar syndrome may be encountered. Nonspecific symptoms such as low-grade fever and anorexia may also be found[1-4]. A high indication of occurrence of this condition consists of painful cranial neuropathy combined with dural thickening observed through neuroimaging. Exclusion of secondary causes of dural thickening such as systemic autoimmune/vasculitic disorders (Wegener granulomatosis, rheumatoid arthritis, Behcet disease, Sjogren syndrome), malignancy (meningioma en plaque, dural carcinomatosis or lymphomatosis), infectious processes (Lyme disease, syphilis, tuberculosis) and other disorders such as sarcoidosis, hemodialysis, intracranial hypotension must be performed by using clinical profiles as well as appropriate investigations[5,6].

There are only few small series cases of IHP in the literature. This paper presents one of the largest single center series of IHP, and may document more

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thoroughly the clinical features, neuroimaging findings, natural histories, therapeutic options, and outcomes of treatment of IHP in a tertiary care center.

Material and Method

Medical records were obtained from the database of adult inpatients, 15 years or older, whom were admitted to King Chulalongkorn Memorial Hospital, Bangkok, Thailand, between January 2000 and November 2011. Search keywords included idiopathic hypertrophic pachymeningitis, multiple cranial neuropathies, polyneuritis cranialis, Tolosa Hunt syndrome, and undetermined headache. There were two inclusion criteria used to select the cases. The first inclusion criteria looked at clinical syndrome of headaches, cranial neuropathy or other neurological deficits with clinical profiles related to the location of hypertrophic pachymeninges. The second inclusion criteria looked at neuroimaging that revealed enhanced hypertrophic dura in post contrast CT scan of the brain and/or MRI of the brain. Patients with history of clinical symptoms of systemic autoimmune/vasculitic disorders, systemic infections, malignancy, and other types of secondary hypertrophic pachymeningitis were excluded. In cases of mass liked lesion of the dura or cases who did not respond to corticosteroid, dural biopsy were performed and only cases with microscopic findings compatible with IHP were included in the study. A corticosteroid was the first line of drug treatment. The patient who did not respond to corticosteroid, azathioprine was prescribed. The demographic data, clinical features, neuroimaging features, natural history, treatment modalities, and response to the treatments were analyzed by using SPSS version 17.0.

Ethical consideration

The Institutional Review Board of the Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand had approved the present study.

Results

Thirty-two patients were enrolled in the present study, consisted of 11 males and 21 females. The gender ratio of male to female was 1:1.9. The age of the patients ranged from 33 to 66 years with a mean age of 49.03±16.12 years. The observed symptoms and clinical findings of the patients were summarized in Table 1. The most common clinical presentations were headaches (93.8%) and diplopia (43.8%). The most common clinical finding was multiple cranial nerve palsy (84.4%). Motor weakness and ataxia were rare. Regarding cranial nerve palsy, the third cranial nerve was the most common affected nerve (56.3%). Headaches without neurological deficit were observed in 12.5% of cases.

All of the 32 patients were diagnosed by neuroimaging such as CT scan and/or MRI of the brain. The locations of the dural thickening are summarized in Table 2. Focal and diffuse enhanced thickening of the dura were observed in 96.9% and 3.1% of the cases respectively. Focal thickening in the supratentorium included the cavernous sinus, superior

Table 1. Clinical features of IHP patient (n = 32)

Clinical feature	n (%)
Symptom	
Headache	30 (93.8)
Focal headache	23 (71.9)
Diffuse headache	7 (21.9)
Diplopia	14 (43.8)
Blurred vision	2 (6.2)
Spastic paraparesis	1 (3.1)
Dysphagia	1 (3.1)
Anorexia	1 (3.1)
Sign	
Cranial nerve (CN) involvement	27 (84.4)
CN II	1 (3.1)
CN III	2 (6.3)
CN IV	1 (3.1)
CN VI	3 (9.4)
CN II, III	1 (3.1)
CN II, III, IV	1 (3.1)
CN II, III, VI	1 (3.1)
CN II, III, IV, V_1	1 (3.1)
CN II, III, IV, V_1, VI	2 (6.3)
CN II, III, IV, VI	1 (3.1)
CN II, III, V_1, VI, VII	1 (3.1)
CN II, III, IV, V_1, V_2, V_3, VI, IX, X, XII	1 (3.1)
CN III, IV	1 (3.1)
CN III, VI	1 (3.1)
CN III, IV, V_1, V_2, V_3	1 (3.1)
CN III, IV, VI	1 (3.1)
CN III, IV, V_1, VI	1 (3.1)
CN III, IV, V_2, VI	1 (3.1)
CN III, IV, V_1, V_2, VI	1 (3.1)
CN V_1, VI	1 (3.1)
CN V_1, V_2, V_3, VI, VII, VIII, IX, X	1 (3.1)
CN VI, XII	1 (3.1)
CN VIII, X, XII	1 (3.1)
Motor (spastic paraparesis)	1 (3.1)
Ataxia	1 (3.1)
No focal neurological deficit	4 (12.5)

IHP = idiopathic hypertrophic pachymeningitis

Table 2. Imaging findings of IHP patient (n = 32)

Imaging findings	n (%)
Location	
Focal	31 (96.9)
Diffuse	1 (3.1)
Supratentorium	23 (67.6)
Infratentorium	8 (23.5)
Both supratentorium and infratentorium	1 (2.9)
Spinal cord	2 (5.9)
Supratentorium	
Cavernous sinus	17 (53.1)
Orbital apex	7 (21.9)
Superior orbital fissure	3 (9.4)
Sphenoid wing	3 (9.4)
Dural cerebral hemisphere	2 (6.3)
Infratentorium	
Falx cerebelli	5 (55.6)
Foramen magnum	2 (22.2)
Meckel's cave	2 (22.2)
Base of skull	2 (22.2)

orbital fissure, orbital apex, and sphenoid wing. Focal thickening in the infratentorium included the falx cerebelli, foramen magnum, Meckel's cave, and the dura at base of the skull. The follow-up neuroimaging was performed in 21 patients. Persisted dural thickening, partial and completed resolution of dural thickening were observed in 12/21 patients (57.14%), 7/21 patients (33.33%), and 2/21 patients (9.52%) respectively. The examples of neuroimaging in IHP were demonstrated in Fig. 1, 2.

Lumbar puncture was performed in 29 patients. Cerebrospinal fluid (CSF) examination revealed lymphocyte pleocytosis with a slight increase in CSF proteins in most cases. The mean number of white blood cells in the CSF was 10.35 cells/mm^3 ranging from 0 to 75 cells/mm^3 and the mean CSF total protein level was 65.54 mg/dl ranging from 4.20 to 294 mg/dl. Examination of the erythrocyte sedimentation rate (ESR) was carried out on 11 patients and C-reactive protein (CRP) was performed on 2 patients. The mean ESR was 34.55 mm/hr and ranged from 8 to 77 mm/hr and the mean CRP was 6.4 mg/L.

Dural biopsy for definite diagnosis was performed in 8 patients. All of the patients had dural thickening and fibrosis. The histopathological study showed chronic inflammatory response such as infiltration of lymphocytes, plasma cells, and epithelioid cells, and 1 patient had associated granuloma.

Regarding the treatments, 27 patients received steroid and 5 patients received only supportive medication for headaches. Patients who received an initial treatment with steroid, all recovered from headache within 16 days. Before the treatment, 26 patients (96.29%) had neurological deficit. Completed and partial recovery of neurological deficit were detected in 11/26 patients (42.30%) and 15/26 patients (57.69%) respectively. Relapsing and recurrence after the steroid treatment were detected in 3/27 patients (11.11%) and 5/27 patients (18.51%) respectively. All patients with relapsing course had rapid tapering off or early discontinuation of the steroid. Recurrence occurred in the range of 4 months to 10 years after initial diagnosis. In 5 patients with relapsing or recurrence, combined treatment with steroid and azathioprine were prescribed. Clinical complete recovery occurred in 2/5 patients (40%). Relapsing and recurrence after the combined treatment were observed in 2/5 patients (40%) and 1/5 patient (20%) respectively. Five patients had

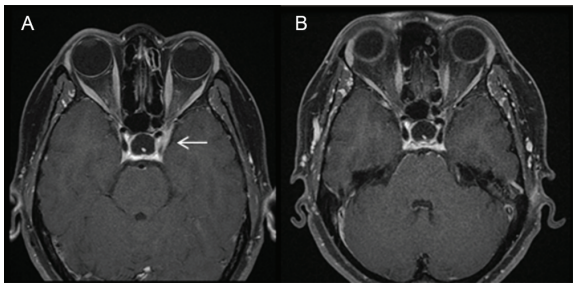

Fig. 1 A) Axial T1 post gadolinium MRI showing enhancing soft tissue thickening at left side cavernous sinus with encasement cavernous portion of the left carotid artery. B) Axial T1 post gadolinium MRI after prednisolone treatment, 10 month after initial MRI showed no abnormal soft tissue thickening.

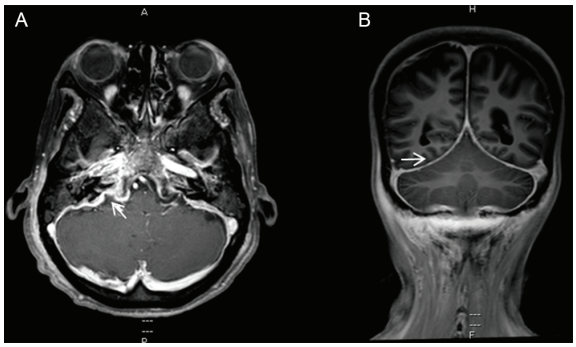

Fig. 2 A) Axial T1 post gadolinium MRI showing dural enhancement at right CP angle (arrow). B) Coronal T1 post gadolinium MRI showed diffuse dural enhancement along tentorium cerebelli.

supportive medication, 4/5 patients (80%) were lost to follow-up and 1/5 (20%) patient had a spontaneous clinical remission without resolution of dural thickening.

Discussion

Hypertrophic pachymeningitis (HP) can be categorized into two subtypes, which are the secondary HP, in which an etiology of the thickened dura has been detected, and the idiopathic HP, in which no etiology except a non-specific inflammation has been found[1,5,7]. The pathological findings of the dura in IHP consist of a dural thickening and fibrosis[1,4-6]. Histopathological findings demonstrate a chronic inflammatory response with lymphocytic infiltration, vasculitis, and granuloma in some patients[6]. The clinical course of IHP is one of a chronic progression or recurrence but spontaneous remission may occur[6,8]. Duration of the disease can be months to years[1]. IHP is usually a steroid responsive disorder[1,6] but some patients had partial clinical improvement or relapses after tapering the dosage. Combined therapy with immunosuppression such as azathioprine or methotrexate may be needed in some cases[9,10].

At the time of the present study, only two large single center series with more than ten cases were reported in the literature[4,6]. The present study enrolled 32 IHP patients in a single center and may reflect a broader spectrum of clinical features, neuroimaging findings, natural histories, therapeutic options, and outcomes of treatments for IHP. The IHP patients in the present study were predominately females, which is different from previous studies[4,6]. The mean age of onset in the present study is 49 years, lower than in previous studies[4,6]. In Riku S et al the mean age was 69 years, while Kupersmith MJ et al had a mean age of 55 years[4,6].

Headache and multiple cranial nerve palsies were the most common clinical findings in the present series, which is similar to other studies[1,4,6,7,11,12]. Headaches were localized in most of the patients and the location of the headaches was related to the site of the cranial neuropathy. In the present series, cranial nerve III's involvement was most commonly observed, followed by cranial nerves VI, IV, V, and II. This observation differs from previous studies. In Riku S et al showed in a series of 14 cases that the most common cranial neuropathy was CN II, VI and III[4] and Kupersmith MJ et al showed in a series of 12 cases that the most common cranial neuropathy was CN II, VI, and III[6].

Dural compression, entrapment, and dural inflammation adjacent to cranial nerves may have been a mechanism of cranial neuropathy. CN III palsy may be associated with pupillary involvement or pupillary sparing which reflected the compression or inflammatory vasculopathy of the CN III from the thickened and inflamed dura. Apart from cranial neuropathy, other neurological deficits included seizure, increased intracranial pressure, venous sinus thrombosis and venous congestion, intracranial mass, and radiculomyelopathy[1,4,6,13,14].

Neuroimaging, especially of the cranial gadolinium enhanced MRI, can demonstrate the location and extent of IHP[15]. The location of dural involvement reflected clinical profiles. In supratentorium, the most common location is in the cavernous sinus, the superior fissure, and the orbital apex. In the infratentorium, the most common location is at the base of the skull and the posterior fossa dura. In the present series, the location of IHP was more frequent in supratentorium, especially in the cavernous sinus and related structures, and the cranial neuropathy was related to these structures. In some cases in the present series, the dural thickening involved tentorium cerebelli and cervical dura and caused ataxia and spastic paraparesis.

CSF examination in most of IHP patients showed lymphocytic pleocytosis with elevated CSF proteins. Indicators for inflammatory response such as CRP and ESR were slightly increased. CSF analysis as well as the indicators for inflammatory response in the present series had similar features to the previous studies[1,4,6].

Dural biopsy for definite diagnosis of IHP is useful in the doubtful cases. The pathology of IHP showed non-specific chronic inflammation such as infiltration of lymphocytes, plasma cells, and fibrotic dura, as well as vasculitis or granulomatous changes. The dural biopsy in the present series revealed the typical features of IHP.

The natural history of IHP is frequently chronic and progressive, spontaneous remission is rarely encountered[16]. Corticosteroid therapy improves headaches and other neurological deficits. Most patients have a good initial response to steroid therapy[10,11,16,17]. Relapsing and recurrence are commonly related to rapid tapering off or early discontinuation of the steroid. The addition of azathioprine is recommended in cases of clinical relapse or frequent attacks in spite of adequate steroid therapy, or in cases of steroid complications[2].

Resolution of the dural thickening was observed in 9/21 patients (42.85%) after treatment with steroid. Spontaneous clinical remission without resolution of dural thickening was observed in 1/5 patients (20%). Persistent dural thickening with clinical improvement may be related to the fibrotic scars from previous inflammation. Spontaneous resolution of the dural thickening without steroid treatment was not detected in the present study. Only one case report revealed a spontaneous resolution of the dural thickening without steroid treatment[8]. The natural history and clinical course of the patients in the present series is similar to the previous studies.

IHP is mysterious in the etiology and the pathogenesis of IHP is still unknown. In a recently published data from a nationwide questionnaire survey of HP in Japan which included 159 cases of HP patients, idiopathic HP, antineutrophil cytoplasmic antibody (ANCA)-related HP, IgG4/multifocal fibrosclerosis (MFS)-related HP and other causes were detected in 70 cases (44%), 54 cases (30.2%), 14 cases (8.8%), and 21 cases (13.2%) respectively[11]. From the information of this survey, ANCA related HP and IgG4/MFS-related HP were the two major causes of all HP and two of these entities should be investigated in the future studies of HP.

Conclusion

IHP is a rare inflammatory disease. From the present series, the most common clinical manifestations were headaches and multiple cranial nerve involvement. Almost all of the patients had a good initial response to steroid therapy. Relapse or recurrence was usually caused by rapid tapering off or early discontinuation of the steroid. Long-term treatment or combined immunosuppression may be necessary in some cases.

What is already known on this topic?

Idiopathic hypertrophic pachymeningitis (IHP) is a rare inflammatory disease with lack of comprehensive clinical features, neuroimaging findings, natural history, therapeutic options, and treatment results. At the time of the present writing only two large, single center series with more than ten cases were reported in the literatures.

What this study adds?

The present study enrolled 32 IHP patients in a single center and therefore may reflect a broader spectrum of clinical features, neuroimaging findings, natural histories, therapeutic options and outcomes of treatments for IHP. From the present series, the most common clinical manifestations were headaches and multiple cranial nerves involvement. Almost all of the patients had good initial response to steroid therapy. Long-term treatment or combined immunosupression may be necessary in some cases.

Potential conflicts of interest

None.

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ภาวะการหนาตัวของเยื่อหุ้มสมองชั้นดูราชนิดไม่ทราบสาเหตุในโรงพยาบาลจุฬาลงกรณ์

ดวงนภา รุ่งพิบูลโสภิษฐ์, กัมมันต์ พันธุมจินดา

ภูมิหลัง: ภาวะการหนาตัวของเยื่อหุ้มสมองชั้นดูราชนิดไม่ทราบสาเหตุ (Idiopathic Hypertrophic Pachymeningitis, IHP) เป็นการอักเสบเรื้อรังทำให้เยื่อหุ้มสมองชั้นดูราหนาตัว เป็นพังผืด ทำให้เกิดอาการปวดศีรษะและเส้นประสาทสมองผิดปกติ ข้อมูลการศึกษาทางคลินิกของกลุ่มอาการนี้มีน้อยในประเทศไทย

วัตถุประสงค์: เพื่อทบทวนอัตราการเกิดโรค ลักษณะอาการทางคลินิก ลักษณะทางรังสีวิทยา การรักษา รวมถึงผลการรักษาผู้ป่วยที่มีภาวะการหนาตัวของเยื่อหุ้มสมอง เพื่อให้เป็นข้อมูลในการศึกษาและดูแลผู้ป่วยต่อไป

วัสดุและวิธีการ: เป็นการศึกษาแบบย้อนหลังในป่วยที่ได้รับการวินิจฉัยว่ามีภาวะ IHP ทุกรายที่เข้ารับการรักษาตัวในโรงพยาบาลจุฬาลงกรณ์ในช่วงปี พ.ศ. 2543 ถึง พ.ศ. 2554 เกณฑ์การวินิจฉัยประกอบด้วย 1) กลุ่มอาการที่เข้าได้กับ IHP, 2) ภาพวินิจฉัยทางระบบประสาทมีการหนาตัวและแสดงการอักเสบของดูราเข้าได้กับอาการ 3) สาเหตุอื่น ๆ ที่จะทำให้เกิดการหนาตัวของดูราได้ถูกแยกออกจากการศึกษาโดยการสืบค้นที่เหมาะสม ข้อมูลทางคลินิก ข้อมูลทางรังสีวิทยา ข้อมูลการรักษา และผลการรักษาได้ถูกรวบรวมและวิเคราะห์โดยใช้โปรแกรม SPSS version 17

ผลการศึกษา: ได้รวบรวมผู้ป่วย IHP จำนวน 32 ราย เป็นหญิง 21 ราย ชาย 11 ราย อายุเฉลี่ย 49.03±16.12 ปี อาการที่พบบ่อยได้แก่ อาการปวดศีรษะ ร้อยละ 93.8 และอาการเห็นภาพซ้อน ร้อยละ 43.8 อาการแสดงที่พบทางระบบประสาท ได้แก่ เส้นประสาทสมองผิดปกติ ร้อยละ 84.4 เส้นประสาทสมองคู่ที่ 3 พบบ่อยที่สุด ร้อยละ 56.3 รองลงมา ได้แก่ เส้นประสาทสมองคู่ที่ 6, 4, 5, 2 ตามลำดับ ผู้ป่วยร้อยละ 12.5 มีเฉพาะอาการปวดศีรษะโดยตรวจไม่พบความผิดปกติใด ๆ ทางระบบประสาท การตรวจภาพวินิจฉัยทางระบบประสาท ร้อยละ 96.9 พบความผิดปกติเฉพาะที่, ร้อยละ 3.1 พบความผิดปกติทั่ว ๆ ไป ความผิดปกติเฉพาะที่บริเวณ supratentorium พบมากที่สุดบริเวณ cavernous sinus, orbital apex, sphenoid wing, superior orbital fissure ตามลำดับ ความผิดปกติเฉพาะที่บริเวณ infratentorium พบมากบริเวณ falx cerebelli นอกจากนี้ยังพบบริเวณ base of skull, Meckel's cave และ foramen magnum ร่วมด้วย การตรวจน้ำไขสันหลังพบลักษณะ lymphocytic pleocytosis ร่วมกับมีการเพิ่มขึ้นของโปรตีนในน้ำไขสันหลัง การรักษาด้วย steroid ในช่วงแรกของการรักษา อาการดีขึ้นทั้งหมด สำหรับผู้ป่วยที่มีอาการกำเริบ (relapse) และกลับเป็นซ้ำ (recurrence) ได้รับการรักษาด้วย prednisolone ตามด้วย azathioprine พบว่าการรักษาได้ผลหายขาด (clinical complete recovery) อาการกำเริบ (relapse) และกลับเป็นซ้ำ (recurrence) ร้อยละ 40, 40, 20 ตามลำดับ ผู้ป่วยทุกรายที่มีอาการกำเริบและกลับเป็นซ้ำพบว่ามีการลดขนาด steroid ลงอย่างรวดเร็ว และหยุดการรักษาในช่วงต้นระหว่างการรักษา ผู้ป่วย 1 ราย ได้การรักษาแบบประคับประคอง พบว่าอาการหายเองได้แม้ว่าการตรวจภาพวินิจฉัยทางระบบประสาทจะยังพบความผิดปกติ

สรุป: ภาวะ IHP จากการศึกษานี้พบว่าอาการทางคลินิกส่วนใหญ่เป็นอาการปวดศีรษะและเส้นประสาทสมองผิดปกติ ผู้ป่วยส่วนใหญ่มักตอบสนองดีต่อการรักษาด้วย steroid อาการกำเริบและกลับเป็นซ้ำพบว่ามีการลดขนาด steroid ลงอย่างรวดเร็ว และหยุดการรักษาในช่วงต้นระหว่างการรักษาในบางรายอาจจำเป็นต้องรักษาด้วย steroid ในระยะยาว หรือ รักษาร่วมกับการใช้ immunosuppressive drug