CASE REPORT

Idiopathic Hypertrophic Pachymeningitis: A Report of Two Patients and Review of the Literature

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ABSTRACT: ***Purpose:*** We report the treatment and follow-up, including MRI, of two patients with idiopathic hypertrophic pachymeningitis and review the English language literature, with emphasis on management and outcome in this rare disorder. ***Methods and Materials:*** The files of two patients were reviewed, with relevant histopathology and imaging (MRI). The first patient has been followed for sixteen years (the longest MRI-documented postoperative course reported for this condition) and the second for two years. The English language literature was reviewed, including a summary of all reported patients that have been followed with MRI or CT imaging. ***Results:*** Despite extensive investigation, no underlying etiology was determined in either patient. Histopathological studies revealed a chronic inflammatory dural infiltrate in both patients, with granulomas in the first but not the second patient. The first patient underwent surgery twice and has remained stable for sixteen years, despite persistent neurologic deficits. The second patient was managed with dexamethasone after a surgical biopsy, and experienced complete resolution of all neurological deficits and abnormalities seen with MRI. ***Conclusions:*** Although prompt and extensive surgery has been recommended for this condition, the results from our second patient indicate that complete remission can be achieved in some patients with biopsy and steroid therapy. This also supports the view that autoimmune mechanisms underlie idiopathic hypertrophic pachymeningitis. The first patient illustrates that extensive laminectomies may be an effective therapeutic option but chronic discomfort may result. If extensive surgery must be performed, laminoplasty should be done because of the potential for reduced pain and improved long-term spinal stability.

RÉSUMÉ: Pachyméningite hypertrophique idiopathique: à propos de deux cas et revue de la littérature. But: Nous rapportons le traitement et le suivi, incluant la RMN, de deux patients ayant présenté une pachyméningite hypertrophique idiopathique et nous revoyons la littérature de langue anglaise en insistant sur le traitement et l'évolution de cette maladie rare. ***Méthodes et sujets:*** Les dossiers ainsi que l'anatomopathologie et l'imagerie (RMN) de deux patients ont été révisés. Le premier patient est suivi depuis seize ans (le suivi postopératoire le plus long documenté par RMN rapporté dans cette maladie) et le deuxième est suivi depuis deux ans. La littérature de langue anglaise a été révisée, incluant un sommaire de tous les cas rapportés qui ont été suivis par RMN ou CTscan. ***Résultats:*** Malgré une investigation poussée, aucune étiologie n'a pu être déterminée dans chacun de ces cas. Les études anatomopathologiques ont révélé une infiltration inflammatoire chronique de la dure-mère, avec des granulomes dans le premier cas seulement. Le premier patient a subi deux interventions chirurgicales et il est demeuré stable pendant seize ans, malgré des déficits neurologiques persistants. Le second patient a reçu de la dexaméthasone après la biopsie chirurgicale et a présenté une résolution complète de tous les déficits neurologiques et des anomalies observées à la RMN. ***Conclusions:*** Bien qu'on recommande de procéder rapidement à une chirurgie extensive dans cette affection, les résultats chez notre second cas indiquent qu'une rémission complète peut être obtenue chez certains cas par la biopsie et la corticothérapie. Ceci est en faveur de l'hypothèse d'un mécanisme autoimmun dans la pachyméningite hypertrophique idiopathique. Le premier cas illustre que les laminectomies extensives peuvent être une option thérapeutique efficace, mais qu'un inconfort chronique peut s'en suivre. Si une chirurgie extensive doit être effectuée, une laminoplastie devrait être faite pour minimiser la douleur et assurer une meilleure stabilité de la colonne vertébrale à long terme.

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Hypertrophic pachymeningitis is a rare disorder characterized by marked inflammatory hypertrophy of the dura mater, with subsequent neurological deficits resulting from the compression of adjacent structures. Based on anatomic site, cases of hypertrophic pachymeningitis can be subdivided into spinal, intracranial and the much less frequent craniospinal pachymeningitis.[1-3]

Hypertrophic pachymeningitis involving the spinal meninges was first described in the nineteenth century by Charcot and Joffroy and the intracranial form was reported shortly thereafter (cited in Ashkenazi et al[4] and Parney et al[5]). Hypertrophic pachymeningitis has often been attributed to specific etiologies, such as tuberculosis or syphilis. Most recent cases, however, have been reported as idiopathic, despite intensive investigation

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to implicate other etiologies. The widespread application of gadolinium enhanced MRI has facilitated the diagnosis and follow-up of patients and increased the frequency of reported cases.[6-8]

We present two cases of idiopathic hypertrophic pachymeningitis; one of craniospinal and the second of spinal distribution, with MRI findings, pathology, and postoperative follow-up. These patients illustrate very different approaches to treatment. The first patient underwent surgery twice, initially for biopsy and subsequently for a second biopsy and decompression. Despite persistent pain and numbness, the patient's neurological function has remained stable 16 years after his initial surgery. To our knowledge this is the longest postoperative follow-up reported for this condition. The second patient was treated with dexamethasone after biopsy of the affected dura, with complete resolution of symptoms and MRI changes. Prompt and extensive surgery has been recommended for this condition but experience with our second patient indicates that complete remission can be achieved with biopsy and steroid therapy. This is consistent with the view that hypertrophic pachymeningitis involves an underlying autoimmune etiology.[1,5,6,8-10]

PATIENT REPORTS

Patient 1

In January 1981 this 61-year-old man presented with diplopia and a twelfth cranial nerve palsy. He later developed pain in his upper cervical region. A myelogram revealed a complete block at the cervical level. Surgery revealed "serous material" overlying and encasing the spinal cord and pathologic evaluation of a biopsy disclosed reactive tissue. He was treated with steroids and improved, with resolution of all neurological problems except the 12th cranial nerve palsy.

In June 1982, he suddenly developed ataxia and weakness in his lower extremities. Physical examination in July revealed sensory loss and upper motor neuron signs below T6. Myelograms revealed adhesive, obliterative arachnoiditis in the lumbar area, complete blocks at T9 and T2, and relative obstruction at C5. CSF contained 34 white blood cells and 106 mg of protein. There was no documented fever or leucocytosis.

Re-exploration was undertaken due to the recurrence of the block and failure to establish a diagnosis. Laminectomy, with decompression and exploration of T3 to T5, revealed a thick, nonpulsatile dura and adherent epidural material. The arachnoid was pulsatile but contained opacities throughout.

Pathologic examination revealed a markedly thickened dura with a nodular chronic inflammatory infiltrate. Small granulomas were noted with thick rims of epithelioid cells surrounding central collections of polymorphonuclear cells. Giant cells were present in a few granulomas. Lymphocytes, histiocytes, plasma cells and occasional polymorphonuclear cells surrounded the granulomas. Stains for bacteria including tubercle bacilli and spirochetes, fungi and protozoa were completely negative. The histological diagnosis was chronic granulomatous inflammation of dural and epidural tissues of unknown etiology.

The patient gradually improved, but in April 1991 experienced increased pain in the posterior mid-thoracic region. An MRI revealed localized spinal cord atrophy at upper thoracic levels (T2-T4) and increased cord signal on T2-weighted images from this level distally, consistent with myelomalacia or ischemic/demyelinating changes. Gadolinium-enhanced axial images throughout the thoracic spine revealed no abnormal enhancement.

In 1997, about fifteen years after his second operation, the patient presented with tightness across his shoulders, activity-induced cramping in the upper extremities and increased numbness in his legs. He was still able to walk for several hours each day and his bowel and bladder function was normal. He had a moderate kyphosis and unchanged neurologic signs. Repeat MRI revealed an atrophic cord and probable posterior tethering at the site of his previous laminectomies. On T2-weighted images the thoracic spinal cord demonstrated focal T2 signal hyperintensity centrally but no definite syrinx.

Patient 2

A 30-year-old woman presented in December 1996, complaining of difficulty urinating, with bilateral leg weakness and numbness. MR revealed a dural lesion extending from approximately the C4 to T3 level (Figures 1 & 2). She subsequently underwent a T1 bilateral laminectomy and biopsy of the lesion. At operation, dark and slightly hemorrhagic tissue was observed in the epidural space, dorsal to the dural tube. The dura appeared thickened and bowed posteriorly. Histopathological examination of the biopsied tissue revealed well-organized connective tissue, infiltrated by a mixed inflammatory response, with giant cells, plasma cells, occasional eosinophils, and foci of B lymphocytes (Figures 3 & 4). Discrete granulomas were not identified. Stains for organisms (bacteria, acid fast bacilli, spirochetes and fungi) were negative. There was no evidence of vasculitis. Investigation for lymphoma, infectious, and connective tissue disease was negative. The final diagnosis was idiopathic spinal pachymeningitis.

Postoperatively the patient was continued on dexamethasone, with gradual improvement in her neurologic function. She developed a pulmonary embolus and required intensive care. New defects appeared on ventilation perfusion scans despite aggressive therapeutic measures and an inferior vena cava filter was implanted. Her neurological function continued to improve. She was able to ambulate with a walker and her bladder function returned to baseline. An MRI done before discharge in February revealed almost complete resolution of the cervical and thoracic dural thickening. A small amount of residual enhancement was seen but there was no evidence of cord compression.

A repeat MRI scan done five months after surgery revealed no dural thickening or spinal cord compression. The residual enhancement noted on the first postoperative MRI had completely disappeared. She was seen one month later, at which time her numbness and weakness had resolved. Another MRI scan done 12 months postoperatively revealed no new findings.

DISCUSSION

Idiopathic hypertrophic pachymeningitis is a rare but possibly underrecognized condition. There are few reports of long-term follow-up of treated patients.[6,8,11,12] Optimal therapy is controversial. Our patients initially underwent biopsy and steroid therapy, with improvement in neurological function. In the first patient, an early relapse has been followed by more than 15 years of stable neurological function. Increased thoracic pain and subjective numbness were likely the result of surgical treatment, as radiologic and neurologic signs of progression were absent. Thus our first patient, with the longest follow-up reported, suggests a relatively favorable long-term prognosis for some patients with this disorder.

The diagnosis of idiopathic hypertrophic pachymeningitis depends on excluding causative diseases, particularly those which call for specific treatment. The natural history is poorly defined. A review of the English and Japanese literature on idiopathic hypertrophic spinal pachymeningitis led to the conclusion that patients with inflammatory signs (fever, increased sedimentation rate, leukocytosis, or increased C-reactive protein) had a poorer prognosis than patients without inflammatory signs.[13]

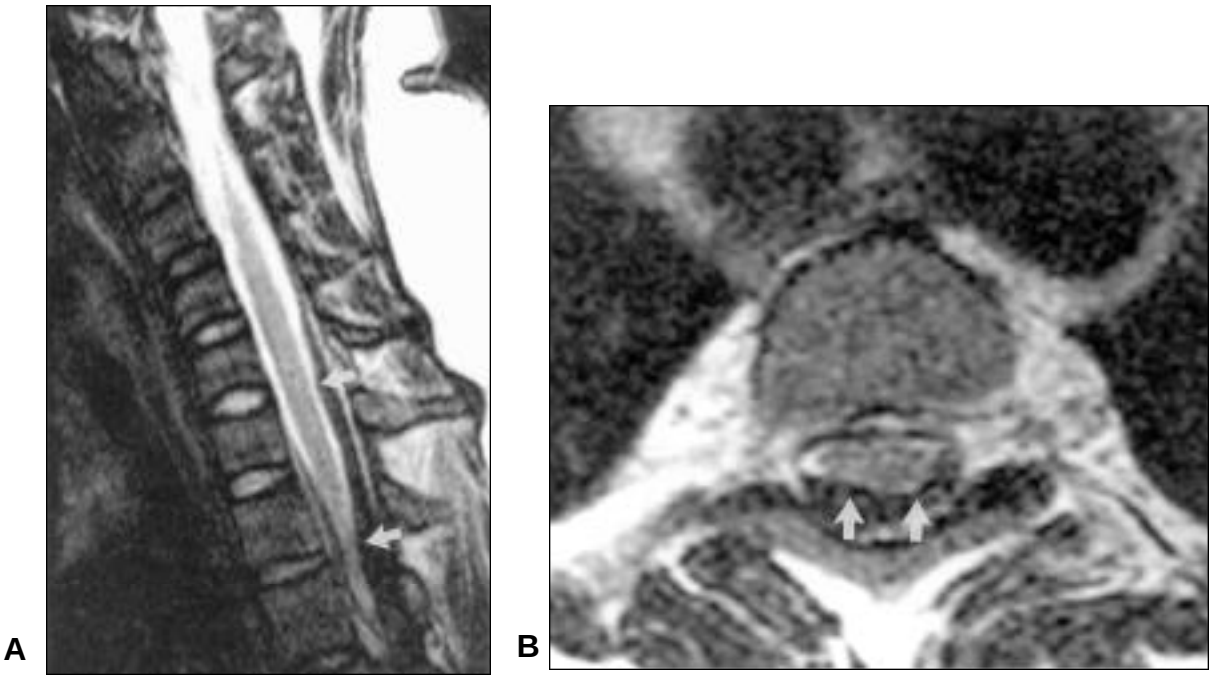

Figure 1: *Patient 2. Sagittal (A) and axial (B) T2-weighted fast spin-echo images of the lower cervical and upper thoracic spine show markedly thickened dura which appears hypointense (arrows), causing mild spinal cord compression.*

Associated diseases

Hypertrophic pachymeningitis can be associated with infectious agents, autoimmune disorders, and other processes.[2,4,14-17] It should be distinguished from dural hypertrophy without inflammation[18] and from enhancement of the dura on neuroimaging without documented hypertrophy or inflammation.[19] Sarcoidosis can present with findings similar to hypertrophic pachymeningitis.[20-23]

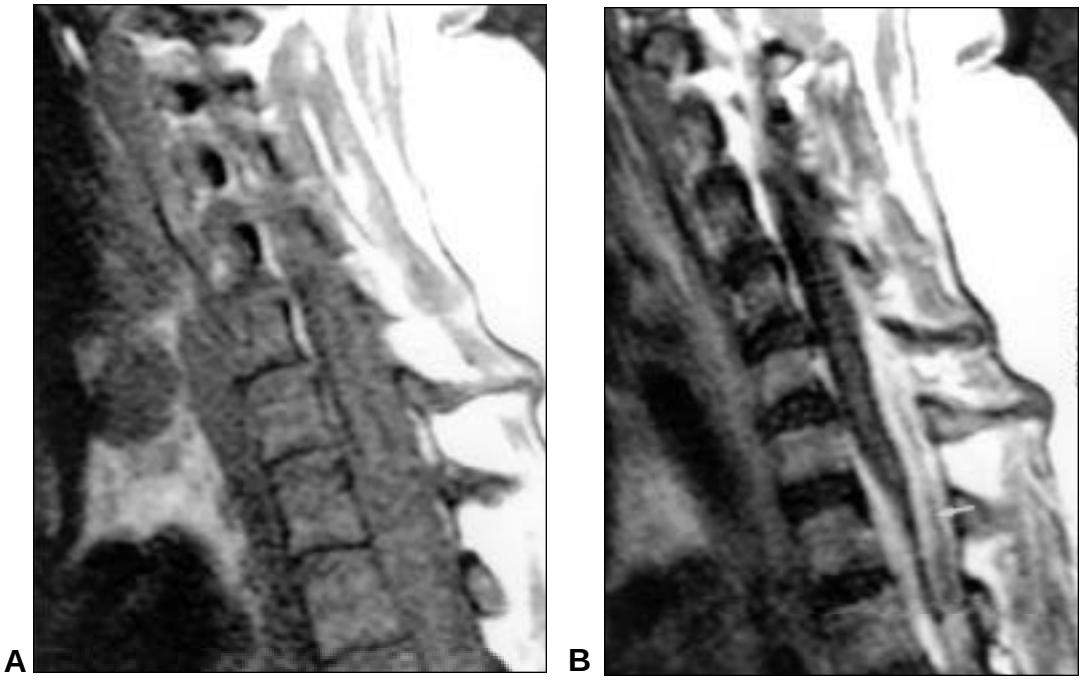

Figure 2: *Patient 2. Sagittal T1-weighted images before (A) and after administration of intravenous Gd-DTPA (B). On the pre-contrast image, spinal cord and dura are isointense and difficult to distinguish. The abnormally thickened dura shows marked contrast enhancement. The enhancement is more marked peripherally with some relative signal hypointensity centrally (arrow).*

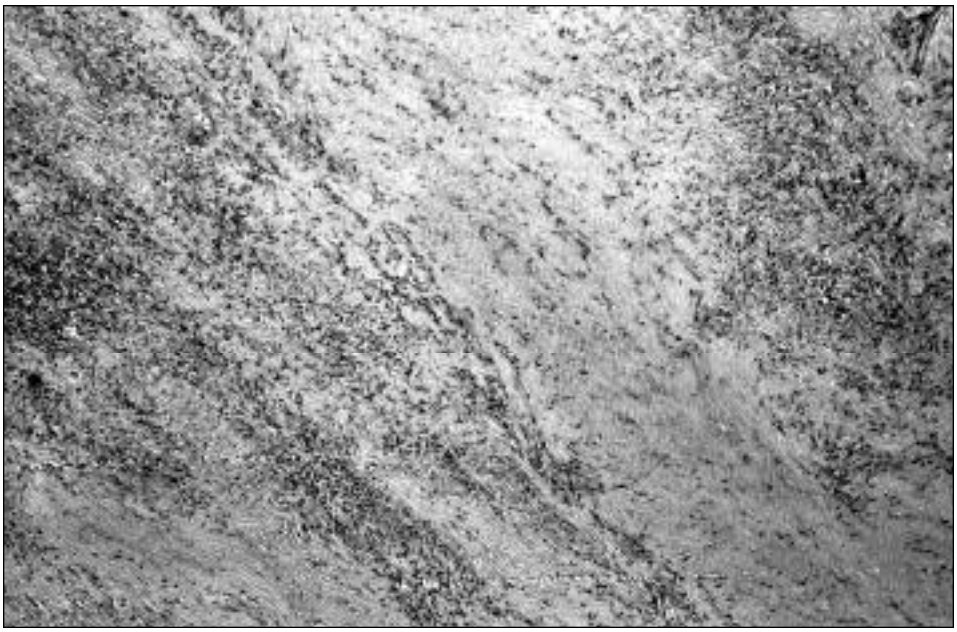

Figure 3: *Patient 2. Periodic acid Schiff (PAS)-stained section showing pathologic features from the surgically excised dura. Markedly thickened dura mater is infiltrated by inflammatory cells (X80)*

Infectious agents reportedly associated with hypertrophic pachymeningitis include syphilis,[24,25] tuberculosis,[26,27] HTLV-I[28] and fungi.[29-31] Pachymeningitis may be the presenting manifestation of adjacent ear or sinus infections.[32,33] PCR diagnostic methods applied to CSF may be helpful in patients where tuberculosis is suspected but routine tests are negative.[5,34]

Hypertrophic pachymeningitis has been associated with various autoimmune processes including rheumatoid arthritis,[35,36] orbital pseudotumor,[37] multifocal fibrosclerosis,[38,39] mixed connective tissue disease[10] and Wegener's granulomatosis.[40] Such association supports an autoimmune pathogenesis for idiopathic cases.[1,5,6,8-10]

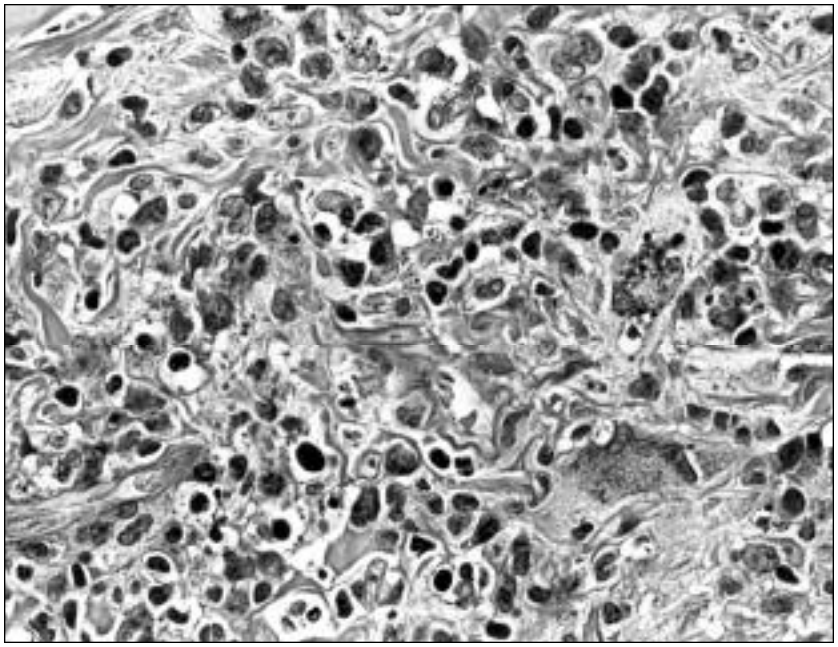

Figure 4: *Patient 2. At higher magnification the inflammatory infiltrate is shown to include giant cells, plasma cells, occasional eosinophils, and lymphocytes (PAS, X800).*

Table: Idiopathic hypertrophic pachymeningitis - Response to Treatment

Author (Ref No)	Age	Sex	Description	Treatment	Outcome
Kanamori et al[11]	28	M	Spinal: T5L2	Steroids (1989)	Remission x2 years
				Expansive laminoplasty (1991)	Improved, then worse at 4 months
				Steroids	Improved; stable at 4 years postop but lesion extended to T2
Mikawa et al[13]	58	F	Spinal: T6T10	Steroids;	Fluctuating course with improvement after third operation
				Laminectomy(x3), dura resection (x2)	
Adler et al[9]	47	M	Spinal: C3T11	Steroids	Stable one week, then rapidly worse
				Laminectomies; longitudinal dura incision & biopsy; steroids; azathioprine	Fluctuating course with postop improvements; stable with deficits at 3 years after third operation
Ashkenazi et al[4]	65	F	Spinal: T1T5	Laminectomy, dura excision	Asymptomatic at 1 yr
	46	F	Spinal: T9T10	Laminectomy, dura excision	Unavailable
Digman et al[61]	70	F	Spinal: All levels	(Antibiotics for concurrent infections)	Progression, died 4 months later due to respiratory failure
Rosenfield et al[41]	25	M	Spinal: C4C7	Laminectomy, steroids	Marked improvement of symptoms at l yr
Kao et al[42]	34	M	Spinal: C7T2	Multiple, transverse durotomies	Symptomatic improvement at 18 months
	28	M	Spinal: C7T2	Multiple, transverse durotomies	Symptomatic improvement at 16 months
Botella et al[1]	55	F	Cranio cervical	VPShunt	Symptoms decreased
				Steroids	Little improvement
				Dura excision	Improvement but recurrence; died of pneumonia
Dumont et al (this report)	30	F	Spinal C4T3	Laminectomy, biopsy, steroids	Complete resolution
	61	M	Craniospinal	Steroids, (laminectomy & biopsy x2)	Improved, recurrent pain at 10 years, spinal cord atrophy, further problems at 15 years but neurologically stable
Friedman et al [2]	65	F	Craniospinal	Surgery	Improvement, no follow-up time given
Goyal et al[15]	28	M	Intracranial	Steroids	Progression at 2 years
	62	F	Intracranial	Steroids	Cranial nerve palsy improvement; MRI improvement at 3 months
	19	F	Intracranial	Steroids	Unavailable
Kitai et al[6]	56	F	Intracranial	Steroids	Symptoms decreased; dura decreased in size at 3 years
Phanthumchinda et al[48]	23	M	Intracranial	Steroids	No symptoms at 1 yr
	30	F	Intracranial	Steroids	MRI improvement at 1 yr
	42	M	Intracranial	Steroids	Symptoms decreased at 10 months
Tanaka et al[8]	45	M	Intracranial	Steroids	MRI and symptomatic improvement at over 2.5 yrs
Jacobson et al[45]	78	M	Intracranial	Steroids, azathioprine	Modest improvement of symptoms; MRI improvement at 18 months
Nishio et al[59]	36	F	Intracranial	None	Spontaneous resolution
Kioumehr et al[47]	35	M	Intracranial	Surgery to release obstruction	Died of postoperative complications
Hamilton et al[16]	55	F	Intracranial	Steroids	Some improvement
				Azathioprine	Marked improvement of symptons at 6 mos
	68	F	Intracranial	Steroids and azathioprine	Improvement of symptoms
	31	M	Intracranial	Steroids	Symptom control
				Methotrexate	No benefit
				Chloroquine and radiation	Marked improvement at 4 months

Author (Ref No)	Age	Sex	Description	Treatment	Outcome
Mamelak et al[49]	67	F	Intracranial	Steroids	Progression
	50	F	Intracranial	Surgical resection (x2), Steroids	Improvement, then recurrence
	75	M	Intracranial	Steroids	Improvement of symptoms; died of cardiopulmonary arrest
Masson et al[12]	58	F	Intracranial	VPshunt, Steroids, Radiotherapy, azathioprine	Little improvement, dead 10 yrs later
	20	F	Intracranial	Steroids and azathioprine	Asymptomatic at 6 years
	57	M	Intracranial	Steroids, radiotherapy, azathioprine	Little improvement, dead 5 yrs later
	41	M	Intracranial	Anticonvulsants, VPShunt	Asymptomatic at 10 years
	50	F	Intracranial		Asymptomatic at 1 year
Lam et al[60]	64	F	Intracranial	Steroids	Symptoms improved at 11 months
	41	M	Intracranial	Steroids	Symptoms improved at 7 months
	70	M	Intracranial	Steroids	Symptoms improved at 12 months
Shintani et al[7]	72	M	Intracranial	Steroids	Symptoms improved No change in MRI at 2 months
Willing and Broghamer[58]	35	F	Intracranial	Steroids	Unavailable
Martin et al[57]	20	F	Intracranial	Steroids, then azathioprine	Asymptomatic at 15 months
	58	F	Intracranial	Steroids and radiotherapy	Progression
	58	M	Intracranial	Steroids and azathioprine	Symptomatic and MRI improvement at 17 months
Kobayashi et al[56]	40	M	Intracranial	Steroids	Little improvement, died soon after
	68	M	Intracranial	Steroids, VPshunt	Little improvement, died of pneumonia

Clinical manifestations

It was the spinal form of hypertrophic pachymeningitis which was first described by Charcot and Joffroy (cited in Ashkenazi et al[4]). They divided the clinical presentation into three distinct stages: 1) intermittent radicular pain that eventually became continuous; 2) muscle weakness and atrophy; 3) spastic paralysis and loss of sphincter control. However, leg weakness or numbness, sometimes associated with bladder dysfunction, and evolving over two weeks to a year, is a characteristic presenting complaint (our patient 2;[4,11,13]). Radicular signs and symptoms confined to the upper extremities may occur.[41] Signs may evolve over a longer time interval.[42] The cranial form of hypertrophic pachymeningitis frequently presents with headache, cranial neuropathies and ataxia.[43]

Radiologic findings

In general, hypertrophic pachymeningitis lesions appear hypointense relative to brain or spinal cord on T1- and, to a greater extent, on T2-weighted images. Contrast-enhanced MRI may show more intense enhancement at the periphery of the lesions with central signal hypointensity, as in our patient 2, possibly due to greater enhancement in an active zone of inflammation peripherally than in a central zone of dense fibrosis.[44] Nonetheless, pathologic confirmation remains essential. It is often only after histopathologic examination of dura mater that the diagnosis can be entertained and a search for potential causes carried out.

Pathology

Grossly observable thickening or inflammation of the dura mater may occur in association with meningioma, craniopharyngioma, lymphoma, metastatic carcinoma, and other tumors. Infectious processes involving the sinuses, middle ear, epidural, or subdural locations can produce inflammation and thickening of the dura. Hence the clinical and radiologic findings and observations at surgery are essential for interpretation of the pathology; and a search for infectious causes is mandatory.

Histologically, distinction between inflammation and a neoplastic proliferation of lymphoid, plasmacytic, or histiocytic elements is fundamental. Most cases of idiopathic pachymeningitis are characterized by a nonnecrotizing chronic inflammatory infiltrate of lymphocytes, plasma cells, and occasional histiocytes, giant cells, polymorphonuclear cells, or eosinophils.[1,4,6,16,17,41,45-48] Granulomas, necrosis and vasculitis are less frequently identified.[2,12-14,49]

Dural involvement has been reported in neurosarcoidosis but involvement of the leptomeninges and substance of the central nervous system are more typical.[20-22,50] Even when involvement is preferentially dural, there is usually an obvious mass, typically subdural, evoking meningioma; but the pattern may be sarcoid en-plaque[51,52] or pachymeningitis.[22,53-55]

Biopsy of affected dura in sarcoid usually reveals the characteristic noncaseating granuloma. In contrast, biopsy of involved dura in rheumatoid arthritis usually reveals only nonspecific inflammation, and the association of pachymeningitis with rheumatoid arthritis relies on other clinical evidence.[35,36] Rheumatoid nodules may involve dura mater in symptomatic pachymeningitis but, to our knowledge, they have been discovered only at autopsy.[35]

Management

The Table summarizes methods and outcomes of treatment in

patients with hypertrophic pachymeningitis in which MRI or CT documentation was available. Most of these patients were idiopathic. Since management differs for the cranial (ICHP) and spinal (ISHP) forms, we separately address their mangement here.

As illustrated in the Table, therapeutic strategies for ICHP have included steroids,[6-8,12,15,16,45,48,49,56-58] azathioprine,[12,16,45,57] methotrexate,[16] chloroquine,[16] radiotherapy,[12,16,57] ventriculoperitoneal shunts,[12,56] antiepileptic drugs,[12] surgery[49] and observation.[59] Corticosteroids can decrease the thickness of dura as documented with MRI,[6,8,15,45,46,48,57] and can result in dramatic reductions of symptoms and complete remission in some patients.[6-8,12,15,16,45,46,48,49,57,60] Patients may become steroid-dependent.[46] Azathioprine may be introduced in an effort to taper the corticosteroids.[16] Early results from the use of azathioprine have been promising[9,12,16,45,57] but the efficacy of chronic azathioprine therapy remains poorly documented.

Radiotherapy probably confers no benefit,[12,57] despite occasional reports of improvement.[16] Surgical techniques and ventriculoperitoneal shunts have been used with variable success.[12,47,49,56] An empiric trial of antituberculous therapy may be warranted in selected patients.[5]

For ISHP, radical surgical treatment has been recommended as the only therapy.[43] Others have emphasized early surgical excision of thickened dura[13] or surgical decompression by laminectomy and excision of the involved dura.[11,61] Immunosuppressive therapy may obviate the need for extensive surgery. Corticosteroids may achieve symptomatic control[9,11] and reduction in dural thickness,[13] which can be virtually complete, as in our second patient.

Decompression may be necessary but our first patient demonstrates that extensive laminectomies can result in chronic discomfort. If decompressive surgery is necessary, laminoplasties should be performed instead of laminectomies, because of the enhanced spinal stability and reduced pain after surgery.[11]

For the management of ISHP, we advocate surgical biopsy and the use of corticosteroids. Azathioprine may be integrated into the therapeutic regimen as noted above in the treatment of ICHP. If stabilization of neurological function is not achieved, surgical intervention with laminoplasty and excision of involved dura should be done.

The need for long-term prospective studies of patients with IHP is apparent. Because of the rarity of the disorder, a North American or worldwide registry of patients may be warranted. Efforts to delineate the epidemiology and underlying autoimmune components of the pathogenesis should be emphasized.

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