

Short report

Non-obstructive idiopathic pachymeningitis cervicalis hypertrophica

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SUMMARY Two young men had similar nonobstructive idiopathic pachymeningitis cervicalis hypertrophica, causing chronic (13 and 11 years respectively) C8-T1 radiculomyelopathy proved by surgical and pathological findings. The preoperative Queckenstedt tests and myelography showed no evidence of CSF obstruction. These unusual findings contrast with previous reports which all described complete or at least partial, block. The findings on metrizamide computed tomogram have not been described before. In the two patients it revealed diffuse cord atrophy from C7 to T2 and hemiatrophy with lateral beaking from C4 to C7. The patients benefited from multiple transverse durotomies. The main pathogenesis of the cord atrophy was the compromising of feeding radicular arteries rather than direct compression.

Hypertrophic spinal pachymeningitis is an uncommon but important disorder characterised by radiculomyelopathy resulting from dural thickening. The few reports have confirmed its diverse aetiologies.¹⁻¹¹ The lesion has been found in all segments of the spinal cord. In the majority of cases the cervical part was involved, giving rise to the name of pachymeningitis cervicalis hypertrophica. Because of the nonspecific clinical manifestations, a definite diagnosis can be made only by surgical intervention with biopsy of the dura,^{1 4 8 9 11 12} or necropsy.^{1 6 7 10} Fortunately, significant myelographic block at the level of the lesion was usually present and led to the early surgical decompression of the cord and nerves.^{2-4 8 9 11 12} Cases with only partial block have been reported as rare exceptions.^{2 12}

High-resolution computed tomography (CT) with intrathecal metrizamide injection has become invaluable in displaying cord morphology¹³⁻¹⁵ but has not been described in pachymeningitis cervicalis hypertrophica. We report two young men with similar chronic idiopathic pachymeningitis cervicalis hypertrophica with unique myelographic presentations and CT scan findings. Both have improved with surgical treatment.

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Case reports

Case 1

A 34 year old male suffered from weakness and wasting of the left hand since the age of 21 years. Three years later, weakness of left leg and increase of tendon reflexes in both legs developed. At that time, CSF, and cervical myelogram were normal. Nine years after onset, the same examinations were repeated and were negative. One year before referring to our hospital he became confined to a wheel-chair. Neurological examination revealed severe muscle weakness and wasting in the left C8/T1 myotomes, moderate on left C7 and mild on left C6 and also right T1. There was hypaesthesia in the left C8/T1 distributions. In the legs, there was severe proprioceptive impairment and bilateral pyramidal signs. Slight tenderness was found on the lower cervical region but the nuchal movement was free.

Serological tests for syphilis, as VDRL, TPHA, and FTA-ABS were nonreactive. ESR, ANA, LE cell and rheumatoid factor were negative. CSF was normal for Queckenstedt test, cells and biochemical contents and negative for variable cultures and VDRL. Electromyography, peripheral nerve conduction and somatosensory evoked potential studies in the four limbs all indicated lower cervical radiculomyelopathy, predominantly on the left side at C8/T1 level. Four view plain radiograph of cervical spines and high cervical myelogram disclosed nothing abnormal. Metrizamide CT from C3 to T3 showed increase in the anterior subarachnoid space, left cord hemiatrophy from C4-5 to C7 with beaking of the left lateral funiculi and diffuse atrophy from C7 to T2 (fig 1, left). There was no compressive lesion.

During exploratory laminectomy from C3 to T1 the dura

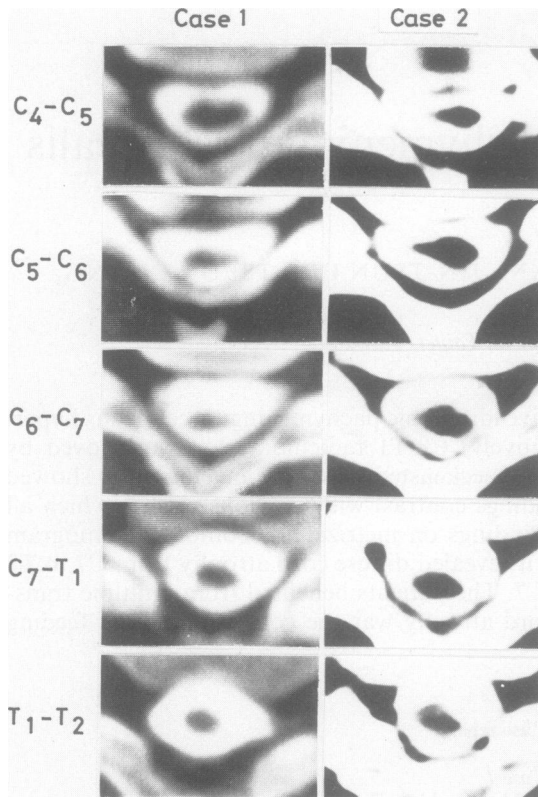


Fig 1 Metrizamide CT scanning from C4/C5 to T1/T2 reveals wide subarachnoid space and no compressive lesion. On case 1 (left column), there is left cord hemiatrophy with lateral beaking from C4 to C7, and diffuse atrophy from C7 to T2. On case 2 (right column) there is right hemiatrophy with beaking from C4 to C7, and irregular diffuse atrophy from C7 to T2.

was found to be abnormally thick, maximal at left C8 and T1 (about 0.8 mm), and tight toward the head. The underlying nerve sheaths were tethered upward, making nerve filaments redundant in the abnormally wide subarachnoid space. The arachnoid membrane seemed slightly thicker but did not compress the cord, which was atrophic and pale. The tightness of the dura and tethering of nerve sheaths were released by bilateral multiple transverse durotomies. Three months after operation, patient could walk unaided. Eighteen months later, he felt that the spasticity and weakness of legs were still diminishing.

Case 2

This man was referred to us at the age of 28 years. Since 11 years previously, he had noticed weakness of the right fifth finger with bilateral numbness in the ulnar distribution, on turning his head. During the past six years, muscle weakness and wasting extended from the hands to the forearms. He

also suffered from numbness up to his nipples, spasticity on walking and incontinence of urine. Three CSF studies and cervical myelographies done at intervals of years in different hospitals were all normal.

Neurological examination revealed weakness and wasting in the upper limbs, severely on the muscles innervated by C8 and T1 spinal nerves, moderately C7 and slightly C6, more pronounced on the right side. Pyramidal sign was present in both legs. There was sensory impairment below the level of C5, more on the left side, with bilateral anaesthesia in C8 and T1 distributions. The right pupil was smaller than the left and light reactive.

Normal laboratory studies included VDRL, TPHA, FTA-ABS, ANA, LE cell, rheumatoid factor, CSF Queckenstedt test, cells, protein, sugar, VDRL test and variable cultures. Peripheral nerve conduction, electromyogram and somatosensory evoked potential studies all indicated an intraspinal disorder predominantly at level of bilateral C8/T1. Plain radiographs of cervical spines in three views, high cervical myelogram and selective vertebral angiogram all were negative. Metrizamide CT scanning from C3 to T3 (fig 1, right) showed right hemiatrophy of cord with lateral beaking from C4 to C7, and irregular diffuse cord atrophy from C7 to T2. The subarachnoid space was wide without a compressive lesion.

Exploratory laminectomy from C3 to T2 (fig 2) disclosed abnormally thick and longitudinally tight dura which made bilateral spinal sheaths strangulated upward. The arachnoid membrane looked normal and was easily detached from overlying dura. The spinal and radicular arteries were hypoplastic. The cord was pale and atrophic. The subarachnoid space was abnormally wide. No cord compression was found. Multiple bilateral transverse durotomies to the dentate ligaments were done and the spontaneous wide separation of the incision margins were not sutured. Two pieces of thickest part of dura, about 0.7 mm in thickness, near the C8 nerve root were taken for H/E, acid fast and PAS staining. Histology showed only connective tissue with a perivascular inflammatory cell infiltrate. Two months after operation, he had no incontinence of urine. Sixteen months later, he reported that the preoperative deteriorating course had been arrested and his legs were stronger.

Discussion

These two men had radiculomyelopathy with similar age of onset, chronicity, clinical and electrophysiological localisation at C8 and T1 level. The dural thickening found by operation, characteristic for pachymeningitis cervicalis hypertrophica, was also maximal at the same level. Other than syphilis,^{1,2} there are many possible aetiologies, such as epidural or subdural suppuration or haematoma, tuberculosis, fungal infection,³ mucopolysaccharidosis,⁴ local irradiation,⁵ intrathecal drug injection,⁶ possible extension of granulomatous sinusitis,⁷ rheumatoid granulation,^{8,9} chronic disc herniation,¹⁰ and, most commonly, idiopathic.^{11,12} The history and laboratory tests indicate that our cases were idiopathic.

Surgical treatment is indispensable for both the

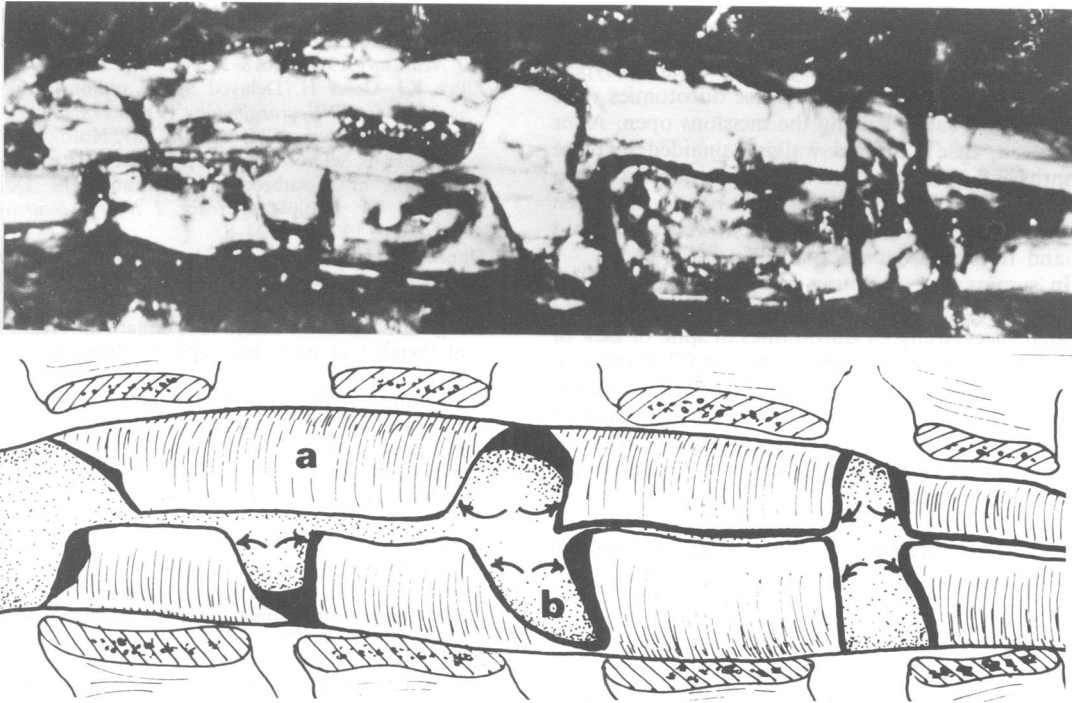


Fig 2 The multiple transverse durotomies cleft widely (↔). The cord (b) under the thick dura (a) seems pale and atrophic.

diagnosis and prognosis.^{1-4 8 11 12} The preoperative periods reported were usually less than a few years. The diagnostic importance of complete myelographic blockage is clear from the reported cases.^{3 4 8 11} Partial block has been exceptionally rare and considered to occur probably only in the earlier stage of the disease process.^{2 12} The repeatedly normal Queckenstedt tests and myelograms, 13 and 11 years after the onset, delayed the correct diagnosis and proper management of our cases, and also make them unusual.

Metrizamide contrast CT scanning has become invaluable in diagnosing cord deformity, especially in the various forms of cord atrophy of different aetiologies.¹³⁻¹⁵ The findings in metrizamide CT have not previously been described in pachymeningitis cervicalis hypertrophica. The findings in our cases were similar. From C7 to T2, which was near the level of the most severe involvement, C8 and T1, CT showed diffuse nonspecific atrophy, as seen in multiple sclerosis¹³ and transverse myelopathy.¹⁴ While, from C4 to C7, which was of less severe involvement, hemiatrophy and beaking of lateral funiculi with the side compatible to the clinical deficits, had been described in Brown-Sequard syndrome caused by injury¹⁴ and spondylolitic

myelopathy¹³ respectively. This combination of different findings might be characteristic for pachymeningitis cervicalis hypertrophica although further experience in this examination is necessary.

Their unexpected operative findings were compatible with the intact patency on myelography and wide subarachnoid space on CT scanning. The arachnoid membranes, grossly normal, were easily separated from dura. There was no real cord compression, which has been described as the main mechanism for myelopathy by most authors.^{1-4 6 11} There was no evidence of constrictive arachnoiditis, which has been associated with the usual myelographic blockage.^{2 8 15} Other than the characteristic dural thickening, the dura was tense which was relieved by the spontaneous widening of the clefts immediately after transverse durotomies. The upward strangulation of nerves and their sheaths caused by the dural tethering could have caused the clinical radiculopathies. The upward stretching was also released by the same procedure. The hypotrophic spinal and radicular arteries and the paleness and atrophy of the cord in case 2 indicate that there had been chronic circulatory deficiency, owing to the compromising of the feeding radicular arteries by the tethered spinal

sheaths.

Few articles describe the surgical technique for treatment of pachymeningitis cervicalis hypertrophica.^{8 11 12} Multiple transverse durotomies were done on our cases, leaving the incisions open. After operation, case 1 started walking unaided in three months and case 2 regained his bladder control in two months. Their preoperative deterioration had been arrested, and subjective improvement were still sensed 18 and 16 months later respectively.

In summary, two young men with chronic idiopathic pachymeningitis cervicalis hypertrophica were treated successfully by durotomies in spite of lack of compressive lesion on myelograms and CT scannings. This form of pachymeningitis cervicalis hypertrophica, although very rare and atypical, should be included in the differential diagnosis of clinical cervical radiculomyelopathy and rendered for early surgical management.

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References

- 1 Agdal N, Hagdrup HK, Wantzin GL. Pachymeningitis cervicalis hypertrophica syphilitica. *Acta Derm Venereol (Stockh)* 1980;**60**:184-6.
- 2 Deshpande DH, Vidyasagar C. Hypertrophic pachymeningitis dorsalis. *Surg Neurol* 1979;**12**:217-20.
- 3 Selby R. Pachymeningitis secondary to *Allescheria boydii*. *J Neurosurg* 1972;**36**:225-7.
- 4 Paulson GW, Meagher JN, Burkhart J. Spinal Pachymeningitis Secondary to mucopolysaccharidosis. *J Neurosurg* 1974;**41**:618-21.
- 5 Zülch KJ, Oeser H. Delayed Spinal radionecrosis—a juridical error? *Neuroradiology* 1974;**8**:173-6.
- 6 Bernat JL, Sadowsky CH, Vincent FM, Nordgren RE, Margolis G. Sclerosing spinal pachymeningitis, a complication of intrathecal administration of Depo-Medrol for multiple sclerosis. *J Neurol Neurosurg Psychiatry* 1976;**39**:1124-8.
- 7 Donishi T, Ishiko T, Kase M, Matsuhta M. Pachymeningitis cervicalis hypertrophica. *Acta Pathol Jpn* 1982;**32**:163-71.
- 8 Hauge T, Magnæs B, Løken AC, Graaf AS. Treatment of rheumatoid pachymeningitis involving the entire thoracic region. *Scand J Rheumatol* 1978;**7**:209-11.
- 9 Markenson JA, McDongal S, Tsairis P, Lockshin MD, Christian CL. Rheumatoid meningitis: A localized immune process. *Ann Intern Med* 1979;**90**:786-9.
- 10 Siqueira EB, Kranzler LI, Dharkar DD. Fibrosis of the dura mater, a cause of "Failed Back" syndrome. *Surg Neurol* 1983;**19**:168-70.
- 11 Guidetti B, Torre EL. Hypertrophic spinal pachymeningitis. *J Neurosurg* 1967;**26**:496-503.
- 12 Wirth FP, Gado M. Incomplete myelographic block with hypertrophic spinal pachymeningitis. *J Neurosurg* 1973;**38**:368-70.
- 13 MaWad ME, Hilal SK, Fetell MR, Silver AJ, Ganti SR, Sane P. Patterns of spinal cord atrophy by metrizamide CT. *AJNR* 1983;**4**:611-3.
- 14 Nakada T, Kwee IL, Palmaz JC. Computed tomography of spinal cord atrophy. *Neuroradiology* 1982;**24**:97-99.
- 15 Donaldson I, Gibson R. Spinal Cord atrophy associated with arachnoiditis as demonstrated by computed tomography. *Neuroradiology* 1982;**24**:101-5.