Syphilitic myelopathy

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SUMMARY Whether the clinical pattern of neurosyphilis has changed since the introduction of penicillin is controversial. This study describes the clinical, laboratory and radiological features of nine patients with syphilitic myelopathy, to assess whether the disease pattern has changed in this sub-group. Four patterns based on clinical course and radiological findings were identified: three patients presented with subacute paraparesis, four with prodromal backache and/or mild leg weakness followed by sudden paraplegia, one patient developed slowly progressive weakness over 4 years and one patient who progressed over one month was shown to have dural thickening on myelography. All patients showed CSF pleocytosis with positive CSF VDRL in seven patients. Despite therapy the prognosis for recovery was not good. Compared with pre-penicillin era studies, the clinical pattern has not significantly changed. Greater alertness to the diagnosis might result in earlier therapy and thus possibly lead to improved prognosis.

The question whether the clinical pattern of neurosyphilis has changed since the introduction of penicillin remains controversial. Although a number of studies have addressed this problem¹⁻⁶ the results are affected by varying and sometimes questionable inclusion criteria and by a lack of detailed clinical information.

Non-tabetic syphilis affecting the spinal cord is rare and only sporadic case reports have been published during the last 40 years.⁷⁻¹¹ This study describes the clinical, laboratory and radiological features of nine patients with syphilitic myelopathy in an attempt to assess whether the disease pattern in this sub-group has changed since the introduction of penicillin.

Patients and methods

The Groote Schuur Hospital computerised record system was used to identify all patients with spinal cord disease and syphilis admitted between January 1977 and June 1984. Patients fulfilling all the following criteria were included in the study:

- (1) Definite clinical evidence of a myelopathy as shown by either upper motor neuron type weakness of the legs with increased tone and brisk tendon reflexes or flaccid areflexic paraplegia in association with a clear truncal sensory level.

- (2) Positive serum *Treponema pallidum* haemagglutination (TPHA) test.
- (3) A positive CSF Venereal Disease Research Laboratory (VDRL) test and/or more than five white blood cells/mm³ of CSF.
- (4) No other cause for meningitis was found by examination and culture of the CSF.

Nine such patients were identified, their charts and radiographs reviewed and the resultant data compiled and analysed.

Results

Six patients were male, three female. The mean age at diagnosis was 36.6 years (range 20-64). Only one patient had a past history of primary syphilis 12 years previously while one had a positive serum VDRL test one year before presentation. A further patient had a negative serum VDRL test five months before presentation.

Clinical and radiological features

The patients could be divided into four groups based on their clinical course and radiologic features. Group A comprised three patients (patients 1-3) with subacute paraparesis progressing over six days to three weeks. Group B comprised four patients (patients 4-7) with a prodromal phase of backache, mild leg weakness or both, lasting between seven days and seven weeks followed by sudden paraplegia evolving over less than 24 hours. Group C (patient 8) comprised a

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Fig Myelogram (patient 9) showing dural lesion at T3 and T4.

single patient with slowly progressive weakness evolving over four years. Group D (patient 9) comprised a single patient with paraparesis progressing over one

month and a cervical dural or extradural lesion on myelography.

The initial symptom was backache in four patients, leg weakness in three and sensory disturbance in the legs in two. Backache occurred in five patients and was localised to the thoracic or lumbar regions. In Group B backache dominated the prodromal phase in three of the four patients overshadowing the initial mild leg weakness. All nine patients had weakness of the legs on examination with four showing complete flaccid paraplegia. The arms were weak in one patient. Sensory abnormalities were noted in all but the single patient in Group D. A clear thoracic or lumbar sensory level for pain and tactile sensation was present in seven patients, six of whom also showed loss of position and vibration sense in the feet. Proprioception alone was disturbed in one patient. Disturbances of micturition were present in all nine patients. Only one patient showed evidence of neurological disease beyond the spinal cord; patient 6 experienced a left midbrain infarct eight months before the myelopathy developed.

Full myelograms were performed in eight of the nine patients and were normal in six. In one patient in Group B the study showed extensive cord swelling from the 2nd cervical to the 9th thoracic segments. Patient 9 (Group D) was found to have an incomplete block due to a dural or extradural lesion at the level of the 3rd and 4th thoracic vertebrae, compatible with hypertrophic pachymeningitis (fig). The lesion had partially resolved on a repeat myelogram 10 days following the initial study.

Laboratory investigations (table)

The serum VDRL was positive in all patients with titres ranging between 8 and 1024. The serum TPHA test was positive in all patients. The CSF protein concentration was raised in all but two patients and ranged between 0.20 g/l and 5.0 g/l. All patients showed a CSF pleocytosis with lymphocytes usually predominating. The cell counts ranged between 7 and 1047/mm³. CSF glucose concentrations were normal. The CSF VDRL was positive in seven patients and the CSF fluorescent treponemal antibody absorption (FTA-ABS) test was positive in all five patients in whom it was performed.

Management and outcome

Patients were treated with varying combinations of intravenous crystalline penicillin G and intramuscular penicillin G procaine for between 10 and 21 days. A limited follow-up study over one to 19 months (mean 6.6 months) showed marked improvement in one patient, slight improvement in four and no improvement in three. Transient improvement occurred in one patient in Group B followed by secondary deteriora-

Table Laboratory results

Patient	Serum VDRL TPHA	CSF protein concentration g/l	CSF cell count (/mm ³)	CSF VDRL and FTA-ABS
Group A				
1	VDRL 64 TPHA pos	1.2	204 L 0 N	VDRL pos FTA-ABS pos
2	VDRL 128 TPHA pos	0.8	16 L 0 N	VDRL neg FTA-ABS pos
3	VDRL 64 TPHA pos	0.2	7 L 0 N	VDRL pos FTA-ABS pos
Group B				
4	VDRL 1024 TPHA pos	0.6	16 L 19 N	VDRL pos FTA-ABS pos
5	VDRL 64 TPHA pos	1.0	920 L 127 N	VDRL pos FTA-ABS pos
6	VDRL 16 TPHA pos	0.8	9 L 0 N	VDRL neg FTA-ABS N.D.
		4.0	99 L 190 N	VDRL neg FTA-ABS N.D.
7	VDRL 1024 TPHA pos	1.2	290 L 30 N	VDRL pos FTA-ABS N.D.
Group C				
8	VDRL 8 TPHA pos	0.4	10 L 0 N	VDRL pos FTA-ABS N.D.
Group D				
9	VDRL 256 TPHA pos	5.0	342 L 7 N	VDRL pos FTA-ABS N.D.

N = Normal; N.D. = not done; L = Lymphocytes; N = Neutrophils.

tion two months later. A repeat myelogram showed the presence of extensive arachnoiditis throughout the thoracic spinal canal. The functional outcome of the patients according to the classification of Paine and Byers¹² was good in one patient, fair in five and poor in three. Insufficient data was available to comment on CSF changes after therapy.

Discussion

In 1944 Adams and Merritt¹³ classified spinal syphilis into (1) syphilitic meningomyelitis, (2) spinal vascular syphilis, (3) syphilitic spinal pachymeningitis comprising spinal cord gummata and hypertrophic pachymeningitis, (4) syphilitic poliomyelitis, and (5) spinal cord compression due to vertebral gumma or aortic aneurysm. Meningomyelitis comprised 15 of their series of 31 patients and consisted of sub-acute or chronic progressive paraparesis associated with variable sensory and sphincter symptoms. The variant of meningomyelitis known as Erb's paraplegia refers to patients with very slow progression over many years and predominantly motor signs.¹⁴ The group of spinal vascular syphilis consisted of 10 patients with sudden onset of flaccid paraplegia, anaesthesia and sphincter disturbance. The other categories of spinal syphilis were rare and syphilitic poliomyelitis was even then considered a dubious entity.

Although the above classification cannot be entirely applied to the present study, it would appear that the

clinical pattern of spinal cord syphilis has not significantly altered. Group A patients fall into the category of meningomyelitis while Group B patients may have had meningomyelitis with superimposed endarteritic cord ischaemia or infarction. The single patient in Group C fulfils the criteria for Erb's paraplegia and the clinical and radiological features of the patient in Group D are compatible with hypertrophic pachymeningitis. Recent case reports^{7,8,11,15} describe patients with either sub-acute paraparesis developing over three to four weeks or sudden onset paraplegia.

A significant clinical feature in the present series is the presence of local backache in five patients, this being the initial symptom in three. The backache and mild leg weakness of Group B patients persisted for up to seven weeks before sudden deterioration, thus providing a period during which astute diagnosis might have prevented the more serious deterioration. Other recently described patients^{7,8} also complained of lower back pain, but not preceding other symptoms. Sensory loss was usually not dissociated in our patients but one patient had only loss of position and vibration sense while another had only loss of pain and tactile sensation. Micturition difficulties occurred in all nine patients.

Only two patients with syphilitic hypertrophic pachymeningitis have been reported in the post-penicillin era.^{9,10} As in the present patient, the lesion was located in the thoracic region in one of these,

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although earlier studies suggest that the cervical meninges used to be the commonest site.¹³ In the rare situation of patients demonstrating an extradural lesion with positive CSF serology and pleocytosis, it is reasonable to give a trial of treatment with penicillin prior to decompression if the patient's clinical status allows.

One of the present patients developed secondary arachnoiditis demonstrated on myelography, but it cannot be certain whether this was due to syphilis or to the previous myelogram. Syphilis as a cause of arachnoiditis is extremely rare and was found in only four of 70 patients in an Indian study.¹⁶ The extensive cord swelling seen on myelogram in one patient is similar to that sometimes seen in acute idiopathic myelopathy.¹⁷

The establishment of laboratory criteria for the diagnosis of neurosyphilis has been fraught with difficulties.¹⁸ The diagnostic criteria used in this study were modified from those suggested by Bracero *et al*¹⁹ and are similar to those used by Burke and Schaberg.⁶ Although the inclusion of cases with a negative CSF VDRL test has been criticised,²⁰ it is widely accepted that 10% to 30% of patients with neurosyphilis have a negative CSF VDRL reaction.²¹⁻²³ Excluding such patients from a reported series may slightly increase the certainty of the diagnosis but will provide a false impression of the clinical spectrum of the condition. In our series the CSF VDRL test was positive in seven of the nine patients and the two patients with a negative test both showed CSF pleocytosis as well as elevated CSF protein concentration.

All nine of our patients showed a pleocytosis and seven of the nine had elevated CSF protein concentration. It has been suggested that these indices are more sensitive indicators of disease activity than the CSF VDRL test alone.²⁴ Although a low CSF glucose concentration has been reported in syphilitic meningomyelitis^{7,8,11} whenever tested this was normal in our patients. It is at present recommended that the CSF FTA-ABS test not be used diagnostically because of problems with its interpretation.^{18,19,25} The test was positive in all five patients on whom it was performed.

The optimal treatment of neurosyphilis remains controversial and it was not the intention of this study to compare different regimes. Our patients were all treated with currently accepted forms of penicillin therapy for neurosyphilis.¹⁸ A short-term follow-up study showed that marked improvement with a good functional outcome was rare and only occurred in one patient. The majority either failed to improve or improved only to a mild degree leaving considerable residual disability. Greater alertness to the diagnosis

may result in earlier therapy and thus possibly improved prognosis.

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