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POST-TRAUMATIC SYRINGOMYELIA

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Abstract. A description is given of the syndrome of post-traumatic syringomyelia amongst patients with traumatic spinal injuries seen at the National Spinal Injuries Centre. The diagnosis was made on clinical grounds. It was confirmed wherever possible by neuroradiology prior to surgery; one case was confirmed only at post-mortem. The incidence of the condition was found to be in keeping with other large series but, in contrast, no difference was found between the time of onset after injury between complete and incomplete lesions of the spinal cord. The commonest manifestation was pain, followed by sensory loss and rarely motor weakness. In the majority of patients the condition eventually became bilateral and in a significant number the lesion ascended to involve the trigeminal territory. The natural history of the condition was followed, and in all cases the condition gradually progressed. The pathogenesis of the condition is briefly discussed in view of the findings.

Key words: Post-Traumatic Syringomyelia; Clinical Features; Natural History.

Introduction

SYRINGOMYELIA as a late sequel to traumatic paraplegia and tetraplegia has been described by Bastian (1876), Strumpell (1880), Holmes (1915), Freeman (1959), Finckle (1960), Barnett, Bottrell, Jousse and Wynn-Jones (1966), Gardner (1973), Barnett, Foster and Hudgson (1973), Shannon, Symon, Logue, Cull, Kang and Kendall (1981). Barnett *et al.* (1966) found a clinical picture suggesting syrinx formation in 10 patients out of a total of 591 paraplegic patients admitted to Lyndhurst Lodge between 1945 and 1966 but were only able to confirm this at operation in one patient. In their monograph of 1973 Barnett, Foster and Hudgson outlined the clinical features of this condition in 17 out of a total of 1,387 patients admitted between 1945 and 1971 and reviewed the findings in a further 56 patients from other sources. This is a rare condition. However, in view of the relatively large number of cases that we have seen at the National Spinal Injuries Centre, Stoke Mandeville Hospital, we are presenting the features of this complication in a further 40 patients.

Clinical Material

Some 6,800 patients have been seen at the National Spinal Injuries Centre since it was opened in 1944 but not all patients had traumatic injuries. Many patients have only been seen on one occasion, some have died, some have been referred to other centres and some have returned overseas. Other patients have been unwilling to return for check ups. We are presenting

TABLE I

Name	Age at Injury	Level	Interval to New Symptoms	New Symptoms	New Motor or Sensory Signs	Duration of Follow up After Injury (years)
R.P.	46	Incomplete L4 Complete S1	13 years	Bilateral girdle pain, mainly right.	Right sided dissociate sensory loss, D5–D8.	17
R.S.	20	Incomplete C7 Complete D3	20 years	Pain in the left shoulder. Bilateral loss of pain sensation in neck.	Bilateral dissociate sensory loss C1–D3. Sweating right side of body.	26
G.B.	15	Complete D11	19 years	Pain in neck, both shoulders and upper limbs. Loss of pain sensation both hands.	Fluctuating sensory loss of trigeminal, sparing C.3, then C5–D11. Pain in first left metacarpophalangeal joints (osteoarthritis). Migraine, dizziness. Ulna sensory changes right and left. (Generalised bilateral weakness).	26
M.M.	31	Complete D8	15 years	Girdle pain at level of lesion. Loss of pain sensation trunk and numbness right upper limb.	Dissociate sensory loss D3–D8 (preservation tendon reflexes).	16
B.H.	26	Incomplete C8	13 years	Pain in both arms, left face, progressive weakness in lower limbs.	Dejerine left face, dissociate loss down to D7. General weakness in all 4 limbs.	21
P.M.	22	Complete D4	8 years	Painful spasms right upper limb followed by loss of pain sensation.	Right dissociate sensory loss C3–D4 and weakness of right hand. Involvement of right trigeminal distribution (migraine).	22

T.G.	21	Complete D7	Less than a year	Sudden onset loss of feeling right upper limb.	Right dissociate sensory loss C7–T2 with minimal weakness right hand.	1
W.W.	23	Complete D7	7 years	Pain at D7 radiating to left shoulder and arm.	Dissociate sensory loss left trigeminal, left upper limb (C3 –D4 with weakness of left upper limb. Severe head pain on stooping.	17
J.M.	18	Incomplete C6	3 years	Pain left upper limb and left chest + spinothalamic loss. No sweating in these parts.	Dissociate sensory loss C2–D3 left. Weak extensor of left elbow. Anhydrosis C2–D3 left. Dissociate sensory loss below C5 on the right.	4
J.D.	18	Incomplete D5	11 years	Pain in neck radiating to left arm. Sweating, weakness left arm.	Weakness left arm. Left trigeminal involvement. Dissociate sensory loss C3–D5.	16
K.T.	33	Complete D7	2 years	Loss of sensation of pain left trunk and left arm.	Dissociate sensory loss C3–D6. Weakness of left finger flexion and intrinsic muscles.	8
J.R.	20	Complete D7	1 year	Chest pain. Weakness with loss of temperature sensation left upper limb.	Dissociate sensory loss C2–D1.	8
K.A.	59	Incomplete C6	8 years	Pain left neck, shoulder. Increasing weakness left hand.	Dissociate loss left C2–C8, wasted triceps and hand intrinsics.	11
D.R.	24	Incomplete D12	18 years	Swelling right forearm, loss of feeling right upper limb with weakness right hand.	Impairment of all modalities of sensation right arm C4–D3. Feels normally until D10.	18

TABLE I continued

Name	Age at Injury	Level	Interval to New Symptoms	New Symptoms	New Motor or Sensory Signs	Duration of Follow up After Injury (years)
D.K.	29	Incomplete D12	15 years	Swelling of right arm. Paraesthesia, sensory loss, progressive weakness right upper limb followed by similar changes on left. Increasing weakness left leg.	Dissociate sensory loss C2–C8 on the right, similar changes on the left. Charcot shoulder on the right.	23
T.M.	26	Incomplete D9	2 months	Tingling numbness, weakness right hand.	Sensory impairment right ulna distribution with weakness, then 14 months later sudden loss of C7, 8, T1 sensation precipitated by coughing. Depressed right triceps jerk.	2
S.K.	8	Incomplete C6	15 years	Pain in left shoulder with weakness and impairment of sensation to involve the whole of left upper limb and face + weakness of right hand.	Bilateral sensory impairment incomplete below C2 on the left, C4 on right with trigeminal involvement on left.	17
D.Ho.	21	Incomplete L3	23 years	Partial loss of pain sensation in right upper limb, denser loss in left hand of sensory modalities.	Dissociate sensory loss C5–L3 on right followed by dense C7–D3 on left.	25
S.W.	18	Incomplete D8	17 years	Back pain. Loss of feeling in left upper limb.	Dissociate sensory loss C1–D4 on left, then complete loss D8–L1 both sides with global weakness of left hand.	31

G.V.	39	Complete D12	2 years	Pain in neck and shoulders.	Dissociate sensory loss C5–D12 on right. Generalised weakness of both (predominantly right hand) upper limbs. Bilateral trigeminal loss.	22
P.W.	44	Complete D7	3 years	Pain in right shoulder, loss of feeling and power in right upper limb.	Dissociate sensory loss C5–D7 right. Weakness of right upper limb.	4
A.Ba.	23	Incomplete L4	19 years	Pain right shoulder. Loss of feeling chest, right shoulder. Cut right hand while doing carpentry. Sensory involvement to T11 on left.	Dissociate sensory loss right arm, C3–D4 (Migraine, Bell's Palsy).	21
T.Mc.	44	Complete L4	8 years	Progressive loss of pain sensation in trunk and right upper limb, lesser degree in left with pain in shoulders. Dissociation of hot and cold and pin prick.	Dissociate sensory loss C4–D4 on right, less marked changes in left, pain in shoulder.	22
J.Re.	37	Incomplete D6	1 year	Pain at D4, loss of sensation left arm leading to pain in left arm.	Dissociate left sensory loss, C3–D6, with global weakness left hand. Late minimal ascent D2 on the right.	8
J.Hu.	20	Incomplete D11	5 years	Paraesthesia right arm, loss of pain sensation in right side of face.	Dissociate sensory loss right trigeminal, C5–D3.	19
J.H.	38	Incomplete L1	9/12	Pain right upper limb and numbness of right side of the face.	Bilateral trigeminal involvement plus right C2–L1 and left C3–L1. Weak right hand intrinsic muscles. Later almost total loss of power in left upper limb resulting in total incapacity.	4

TABLE I continued

Name	Age at Injury	Level	Interval to New Symptoms	New Symptoms	New Motor or Sensory Signs	Duration of Follow up After Injury (years)
E.F.	55	Complete D4	2 years	Pain and weakness left arm and hand.	General weakness of whole left upper limb. Absent reflexes. Dissociate sensory loss C5–D4 on left.	3
R.R.	22	Complete L4	2 years	Loss of pain sensation in right upper limb followed by pain in right chest and shoulder. Loss of power both lower limbs.	Dissociate sensory loss from C3–L4.	28
J.Hi.	23	Incomplete D10	6 years	Pain in right chest followed by loss of pain sensation and hot and cold in right hand.	Dissociate sensory loss between C5–D10 on the right, similar changes 2 years later on left.	9
A.S.	14	Complete D6	2 years	Pain and sensory loss in left upper limb.	Dissociate sensory loss C4–D6.	33
P.B.	15	Complete D6	16 years	Loss of pain sensation in right upper limb, pain in right chest and neck followed by similar changes on left.	Dissociate sensory loss right trigeminal to D6 followed by similar changes on left with generalised weakness of both upper limbs. Commencing in right hand with stepwise progression.	28
S.Wi.	18	Complete D.8	1 year	Pain in left arm, left side of head and leg with global weakness of left hand. Late ascent two segments on right. Then sudden deterioration later.	Dissociate sensory loss between C5–D8. 4 years later sudden deterioration with dense loss all modalities C2–D8 with tremor, dysphagia, hiccoughs and paralysed left diaphragm. C.T. scan confirmed cyst in the medulla.	4

T.C.	46	Incomplete D5	1 year	Pain in right shoulder followed by partial loss of feeling in right chest.	Dissociate sensory loss between D4–D6.	3
G.Y.	58	Complete D5	18 months	Feeling of coldness in right arm accompanied by difficulty in swallowing and talking.	Rotatory nystagmus, loss of upward gaze, paralysed palate, paralysed vocal cord. Global weakness of right arm. Complete loss of reflexes in right arm. Dissociate sensory loss below C2 on right, minimal sensory changes on left.	7½
D.E.	19	Complete D5	9/12	Sensory loss to C5 which remitted for many years then developed weakness in left hand.	Sensory impairment to C5 which remitted then weakness developed in interossei muscles of left hand 17 years after injury accompanied by minimal sensory changes in C8 distribution.	17

here 40 patients with post-traumatic syringomyelia, Table 1. One was caused by a stab wound, two by gunshot wounds, the remainder by direct or indirect violence. We know of other patients who have developed this condition following spinal anaesthesia, osteomyelitis and thoracic disc protrusions but which have not been included in this study. The diagnosis had been made by neuroradiological investigations and operations in 22 patients Fig. 1 and in one of these a post-mortem confirmed the diagnosis. In two patients the diagnosis was made at operation, in one by neuroradiological investigation and in one patient at post mortem. In the remaining fourteen the diagnosis was made on clinical grounds alone. (See Table 2).

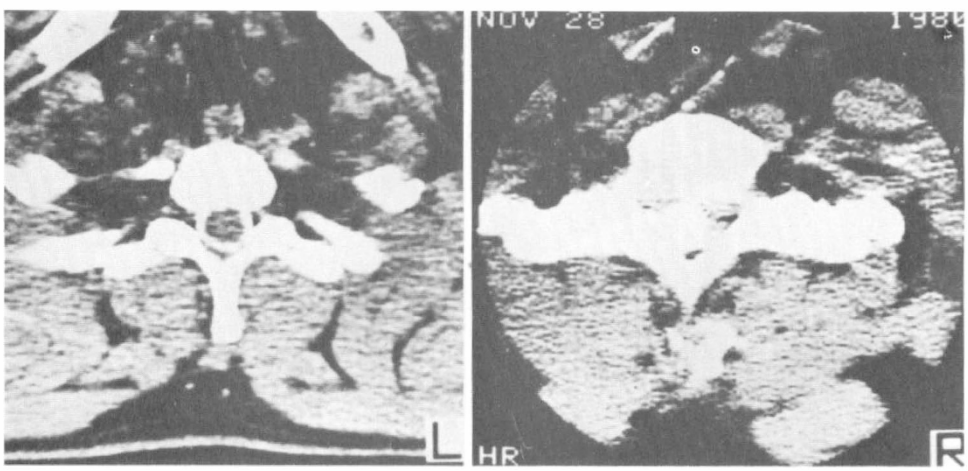

FIG. 1

Augmented metrizamide enhanced computerised tomography.
a. Early scan shows dye around the dilated cord.
b. Eight hours later the dye has entered the cyst.

Incidence

The 40 patients out of 6,800 admissions to the National Spinal Injuries Centre only represents a proportion of the patients with this condition and the figure of 0·6 per cent is unrealistic. There are at least another ten traumatic cases not presented in this publication since the investigations or documentation are not complete.

A much more realistic figure can be obtained by looking at the incidence amongst 370 consecutive traumatic patients personally followed up by one of the authors over a ten year period (1970–1980), 166 cervical, 154 dorsal, 45 lumbar, five not known. There were six patients in this series giving an incidence of 1·6 per cent, a figure similar to that obtained at Lyndhurst Lodge by Barnett *et al.* (1973), where they found that out of a series of 1,387 patients seventeen had this condition, giving an incidence of 1·2 per cent. It should be noted that the period of study of the Lyndhurst Lodge series was between 1945 and 1971, a span of 26 years. This is longer than our own series, 1970 to 1980, and our figure of 1·6 per cent is an under-

TABLE II

Diagnosis

Confirmed				Not Confirmed		
Neuroradiological Investigations and Surgery	Neuroradiological Investigations Alone	Post Mortem Alone	Operation Alone	Neuroradiological Investigations Negative	No Investigation	Total
22*	1	1	2	6	8	40

*One also had an autopsy.

estimate since it is known that this condition can manifest itself as late as 18 years after the onset of paraplegia.

Level of Injury in Patients Developing the Condition

Six cervical, 27 thoracic and seven lumbar patients developed post-traumatic syringomyelia. Few cervical injuries showed this condition, i.e. six out of 40 and this is in common with Barnett *et al.* (1973), who found only one cervical patient in their series of 17.

In the early days few patients with cervical injuries were admitted to the National Spinal Injuries Centre. Sir Ludwig Guttmann (1976), in an analysis of the first 3,000 patients found 466 cervical, 1,036 thoracic and 461 lumbar lesions, the rest being paralysed as a result of medical conditions. The mortality amongst the cervical patients was high so it might be argued that the cervical patients were not surviving long enough to develop this condition. This argument cannot be applied to the experience of the last ten years when there was, amongst 166 cervical patients referred to previously, a mortality rate of 10 per cent at ten years, Ravichandran and Silver (1982). Once these patients survive the initial six weeks they can be expected to survive for twenty-five years. The small number of cervical patients who have developed post-traumatic syringomyelia thus represents a diminished risk in this type of patient of developing this condition. This has been confirmed statistically by observing that the proportion of cervical to thoracic lesions among the syringomyelia cases was significantly less than among the patients as a whole ($P < 0{\cdot}001$) whereas the proportion of thoracic and lumbar lesions did not differ ($P > 0{\cdot}05$). Unfortunately, there are not enough cases to determine if patients with thoracic injuries are particularly liable to develop this condition, compared with patients with lumbar injuries.

In view of the rarity of patients with cervical lesions developing syringomyelia, we are presenting the history of one case, K.A., in detail:—

In 1966 a 59-year-old female sustained a gunshot wound of the spine causing tetraplegia incomplete below C5/6 and complete below C8. A laminectomy operation was performed. She was admitted to the 'Young Chronic Sick Unit' as a long-term patient. In 1974 she began to notice a loss of sensation and weakness of the whole of the left arm and also a shooting pain in the left side of the neck and shoulder radiating to the hand. She developed further weakness of her left upper limb in the shoulder and elbow and her previously partially paralysed hands had become more wasted and weaker.

The tendon jerks were normal in the right arm but were depressed in the left and were absent in both lower limbs. Sensation was impaired below the third division of the trigeminal nerve on the left to involve the whole of the left arm to all modalities and in the right arm below C8.

A myelogram showed a block at the lower border of the T3 vertebrae and the cervical cord was dilated in the C5/C7 region. There was an irregular narrowing of the subarachnoid space at the site of the previous trauma. An operation was not carried out at this stage and she was discharged to a nursing home. Following a bumpy car ride she noticed a further deterioration in her condition. Surgery was then carried out and the cord appeared to have degenerated into a system of multiple cysts which communicated with one another to a considerable extent. Longitudinal incisions were made in the cord in the D1/2 region, the cysts being incised on either side to reach the subarachnoid space anteriorly. She developed a cerebrospinal

fluid leak but following a further operation this healed. She was discharged to long-term care elsewhere, and re-admitted in January 1977 with hypothermia and died on 14.1.77. A post mortem examination was carried out:—

Macroscopic appearances of the spinal cord

At the cervicothoracic level the theca was bound down by dense fibrous adhesions to the cord and bone was adherent to the meninges. In the upper cervical region the cord showed marked flattening of the posterior columns and also a slit-like cavity involving grey matter, particularly the anterior horn on the left side. In the midcervical region this cavity was more pronounced. At C5 the whole cord was shrunken, with extensive bilateral slit-like cavitation. At the site of the dense meningeal adhesions, the possible site of the original bullet wound, i.e. the lower cervical, upper thoracic region, the cord was completely replaced by multiple smooth walled cavities with intervening septae of dense fibrous tissue and grey, probably gliotic tissue. Immediately below this mid thoracic level more cord substance was visible but at least two separate cystic cavities were present. In the lower thoracic and upper lumbar levels there was a single, central, smooth-walled cavity. Below this there again appeared to be multiple cavities separated by septae. At about L3 level the cord was flattened antero-posteriorly and had a single slit-like cavity. At the mid-lumbar level there were no obvious syringomyelic cavities but there was a thin grey area of possible demyelination of the gracile tract bilaterally and also greyness in the pyramidal regions. In the conus medullaris there was a central cruciate cavity probably the central canal. The anterior roots generally appeared to be shrunken and grey. Fig. 2.
Comment: After a delay of eight years this patient developed a typical syndrome of post-traumatic syringomyelia which extended both above and below the level of her initial injury.

We know of two other cases, not included in this series, who initially had cervical lesions who developed this condition. Details of one of these is presented:—

On 30.9.76 E.H., a nineteen-year-old male, was involved in a road traffic accident. He sustained a fracture dislocation of C6 resulting in tetraplegia incomplete below C5, complete below D3. There was considerable recovery of power in both his hands, particularly on the right and a little on the left. During December 1981 he developed severe interscapular pain with increasing numbness and weakness of the right arm and hand. Both his hands developed weakness in previously strong muscles. He developed sensory loss over the whole right hand side of the body including the trigeminal distribution, with associated tendon reflex loss.

Time of onset of post-traumatic syringomyelia

The time of onset of this condition is extremely difficult to determine, especially in retrospect, as it was only after the publication of Barnett *et al.*'s paper in 1966 describing the condition and his subsequent studies showing that it could be arrested by operation, that we began to consider its presence in patients who were then referred for investigation at neurosurgical centres. We are now diagnosing the condition earlier and more frequently.

We have made an effort by studying the notes and questioning the patients to determine the time of first presentation, but it is obvious that in

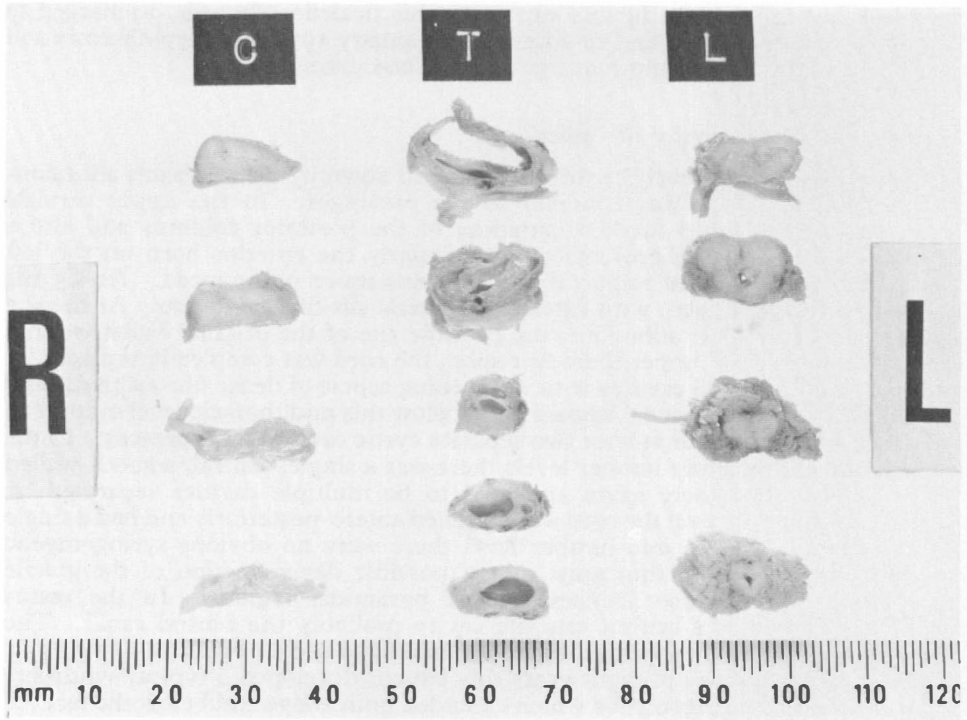

FIG. 2

Post Mortem on K.A. Transverse sections of the cord at cervical, thoracic and lumbar levels. There is cavitation in the cord from the cervical to the lumbar region, in part multilocular. Note, it extends well below the site of the initial injury.

patients whose condition presented some 25 years ago that inaccuracies are present, particularly when the onset was insidious. To avoid doubt we suggested taking the date of onset of the condition as being the time when unequivocal sensory loss was present. Even today when we are more aware of the disorder, and are referring patients for early investigation, we are still surprised by the atypical way in which the condition may present.

In nine patients it presented at a year or less and in four as late as 18 years after injury. The earliest case, T.M., was observed to have tingling of the right hand at two months followed by loss of sensation at 8 months and wasting of the interosseous muscles at 11 months. The diagnosis was not entertained and subsequently electromyography and nerve conduction studies were performed as he was thought to have an ulnar nerve lesion. The diagnosis was not confirmed and it was only some 14 months after injury, after coughing suddenly, that he developed pain together with a wider area of sensory loss. The diagnosis of syringomyelia was confirmed by a myelogram and at operation.

We could not detect any significant difference in the time of onset between patients with complete or incomplete lesions. There were 22 in one group and 18 in the other and statistical analysis showed no significant

difference. ($P > 0.05$; Students T test). We subjected Barnett's figures to a similar analysis and, this too, failed to show any significant difference.

Initial Manifestations

Despite the fact that the development of fresh symptoms and signs above the level of the initial injury would appear to be a frightening experience, only 10 patients requested a special appointment at the hospital because of this. Out of these ten patients seven complained of pain, two of weakness of the hand, and two of sensory loss. In the remaining patients the condition was detected at routine examination when they were seen for check up, Table 3.

TABLE III

Initial Manifestations	
Pain	25
Sensory Loss	8
Motor Weakness	2
Other	5

In 40 patients pain was the commonest presenting symptom. Seventeen had continuous pain, three had pain on straining or coughing and one had both. Eleven patients were found to have an extension of their sensory loss above the level of the lesion; two patients had weakness of the hand and four had paraesthesiae. While the signs and symptoms mentioned above brought the patient to hospital, re-examining them and studying their notes has revealed more subtle symptoms such as excessive sweating, early loss of previously present reflexes and minor alterations in muscle power that were attributed to arthritis around the shoulder girdle. Others had loss of sensation attributed to a carpal tunnel syndrome, pain or difficulty with swallowing (and one patient who presented with 'painless' dislocated shoulder, not included in the series). (Fig. 3).

Pain

Pain was the commonest presenting feature. In 25 patients it was an early finding, and of these, seven requested consultation at the hospital because the pain was severe. In the remaining 20 it was noted during routine examination. Only ten patients had no pain at any time and one of these developed pain for a short period only after surgery had been performed upon the syrinx.

In 12 patients the pain was localised centrally over the lower cervical or upper thoracic spinous processes. It radiated into the arm or trunk in all except one patient. In two patients the limbs were not involved, the pain being in the chest or abdominal wall only, while one patient with a cervical lesion presented with pain in the throat.

The character of the pain was variable, the commonest being a dull ache as seen in 20 patients. It was precipitated by coughing in 12 patients and by exercise in six. It is striking that a small number of patients clearly dated the onset of pain from an episode of coughing, straining or

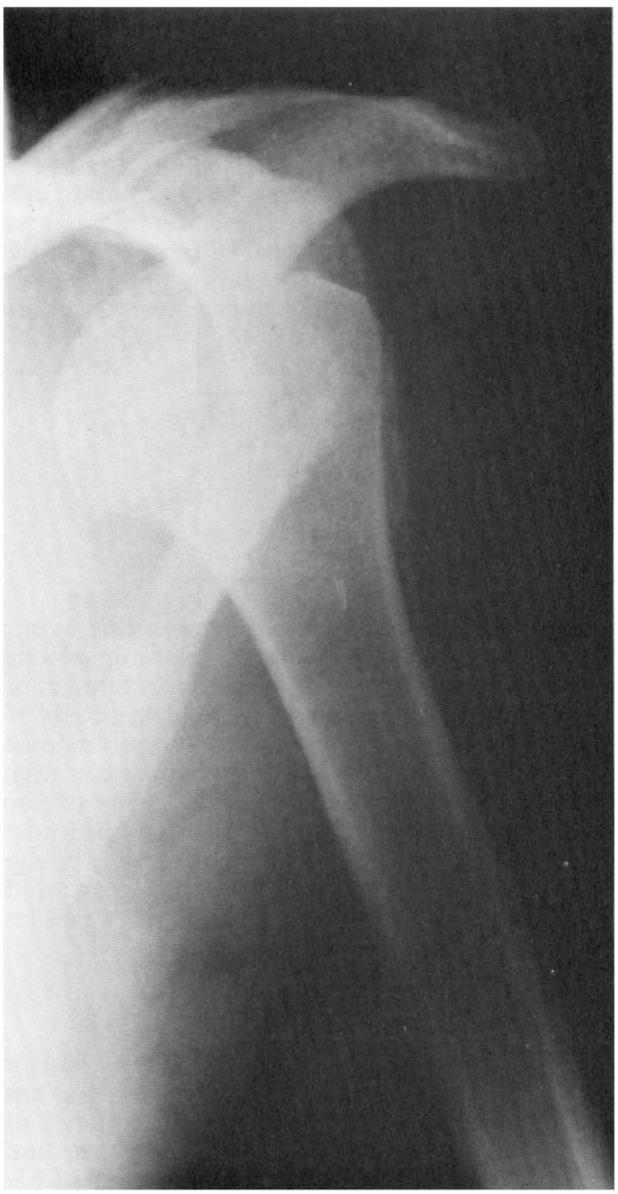

FIG. 3

Painless dislocated shoulder in a paraplegic patient. In a normal person this would be extremely painful.

strenuous exertion. While the pain was usually a dull ache it could be stabbing or burning in nature. When it occurred at or below the level of the cord lesion it was indistinguishable from root pain. When the pain occurred in normally innervated areas of skin well above the cord lesion it was more significant. A striking symptom was the description in four

patients of an itch that could not be scratched, in the anaesthetic areas. We have not seen the dramatic progression described by Barnett *et al.* (1973), of the pain ascending from the level of the lesion, followed by an area of numbness.

The pain fluctuated in intensity and sometimes the patient was free from pain for several days. It was aggravated by tension or bouts of fever but could sometimes be relieved by dexamethasone or an injection of a local anaesthetic. Only one patient, who had also had a severe head injury, became addicted to narcotics.

The most striking feature was the dramatic relief obtained by surgical drainage of the cyst. In some cases the relief of the pain was noted when they recovered from the anaesthetic but in others it took some weeks to disappear. The pain recurred if the drainage became blocked, and was relieved when drainage was re-established. In only one patient was the pain precipitated by the operation, but in this case the pain was of short duration.

Sensory Disturbance

The commonest presenting clinical sign was sensory disturbance, occurring at some stage in virtually every patient. Indeed we would say that its absence would make the diagnosis extremely unlikely, in view of the location of the cyst, demonstrated at post mortem and by myelography, at the base of the posterior horn of grey matter. Sensory changes were very striking in all the patients apart from one in whom it was difficult to detect, even when motor manifestations were well established. Only two patients were so distressed that they sought help from the hospital; one had cut his hand while sawing wood, and the other was frightened by increasing sensory loss. Two other patients first noticed severe swelling of their arms; in one it was due to a Charcot shoulder but the other had cellulitis. Presumably he had neglected a small cut or cigarette burn on his hand. When the swelling subsided following a course of antibiotic therapy he was found to have considerable sensory loss. This swelling has been described in classical syringomyelia by Morvan (1883). Apart from these, the majority of patients were surprisingly little disturbed by the sensory loss, and it was found at routine check up examination.

Mode of Progress

The progress of the sensory loss tends to fall into clearly defined patterns. Firstly, in the majority of patients the sensory loss began at the site of injury and ascended from there to higher levels, the progress usually being insidious. Secondly, in six patients it took place in sharply defined steps. The progress stopped for an interval of months or years and then suddenly following an episode of straining or coughing, the lesion ascended a variable number of segments, sometimes overnight. Thirdly, in 14 patients the pattern was different, presenting with an area of sensory loss over the face or upper limbs with an area of intact sensation between this and the spinal cord lesion, the two gradually merging.

Speed of Progress

The speed and direction of progress of the sensory loss was variable in some

patients taking some years to ascend and spread. Conversely, in others there was a rapid onset and within a matter of hours the condition had spread from the trunk to involve the whole of the upper limbs. In T.G. it presented with a band of numbness in the C8 distribution and, overnight (following a day's vigorous physiotherapy), it had spread from D7 to involve the whole of the upper limb, involving 11 segments. He was at this time undergoing his initial programme of rehabilitation at the National Spinal Injuries Centre, Stoke Mandeville Hospital.

In 12 patients the sensory loss spread to involve the face. No correlation could be found between the duration of the condition and the distance spread. Some patients with cervical lesions did not show any involvement of the face, although they had shown other changes for many years. Conversely, J.H. who had a lumbar lesion, showed within a year a rapid ascent to involve both the upper limbs and face. We did not find the relationship of pain described by Barnett *et al.* (1973) who found that the pain preceded the sensory loss and was associated with, or was gradually replaced by, a sensation of numbness. Some patients on questioning remembered that they had observed either excessive sweating in areas subsequently shown to be involved above the level of the lesion or, more rarely, the sweating disappeared. However, sweating disturbance in patients with spinal cord lesions is such a common occurrence above and below the level of the lesion that fluctuations in its intensity, when significant, is difficult to assess, except in retrospect.

Character of the Sensory Loss

The features of the sensory disturbance were predominantly spinothalamic. Loss was not total or uniform. There was sometimes loss of both deep and superficial pain sensation as shown by the development of Charcot joints. This is particularly liable to occur in these patients as they place undue strain upon their upper limbs which they use to transfer. In other patients deep pain was preserved and one, P.M., developed a painful tennis elbow beneath the anaesthetic skin. S.W. showed severe loss of all modalities and developed pseudo-athetosis and intention tremor in the left upper limb.

There was a dissociation between the temperature modalities so that sometimes cold appreciation was preserved and heat appreciation lost and sometimes the converse. The dissociation over the face was a special situation (Table 4). Some of the patients showed a classical Dejerine (1914) loss whereby peripheral areas of the face lost their sensation, the nose or lip being spared. The onionskin preservation has been described by Dejerine and is indicative of a lesion in the descending spinal tract of the Vth cranial nerve in the spinal cord. We also noticed loss of taste over the anterior two-thirds of the tongue associated with this loss. Appreciation of taste requires normal sensation over the tongue, mediated by the Vth cranial nerve and its loss does not indicate a lesion of the chorda tympani.

A further observation was that these patients showed patches of normal sensation in little islands of skin that were otherwise anaesthetic. Conversely one patient showed an island of total sensory loss.

TABLE IV

Facial Involvement

Level	Name	Time since injury until symptoms developed	Time since injury until facial involvement	Interval from symptoms developing to facial involvement	Dejerine	Bilateral	Non Dejerine
C6	S.I.	1	2	Presenting		+	+
C6	S.K.	15	17	2	+		
C8	B.H.	13	21	8	+		
D4	P.M.	8	12	4			+
D5	I.D.	11	15	4			+
D6	P.B.	16	22	6		+	+
D6	J.D.	9	11	2		+	+
D7	W.W.	7	17	10			+
D11	J.Hu.	5	9	4			+
D11	G.B.	19	36	17	+		
D12	G.V.	2	2	Less than 1 year		+	+
L1	J.H.	1	1	Less than 1 year		+	+

Variability

Daily variations occurred in the sensory loss, particularly in the facial distribution and some striking remissions occurred. One patient D.E. who had mild bilateral sensory loss, completely remitted, but many years later he developed motor weakness. Remissions could be induced by dexamethasone and by surgery.

Motor Involvement

Motor weakness was not a common presenting feature. Only three patients showed this as a first manifestation but the majority of the patients, (28), showed it at some stage (Table 5). This was lower motor neurone in type. Rarely it was preceded by exaggerated reflexes although this usually disappeared as the condition progressed. In patients with complete lesions, weakness could only be seen above the level of the lesion, usually in the upper limbs. The onset of weakness could be both insidious and subtle. The patients did not usually complain until their attention was drawn to the weakness when specifically questioned and tested and one patient only realised that there had been weakness when after surgery he found his hands were much stronger.

TABLE V

Motor Weakness	
Unilateral	
Hand	10
Whole upper limb	7
Shoulder only	1
Elbow	1
Bilateral	
Upper limb only	6
Lower limb above level	1
Upper and lower limbs	
Both arms + 1 leg in a paraplegic	1
Both arms + 2 legs in a quadriplegic	1
No weakness	12
Total	40
N.B. From the above:	
Tetraplegic helplessness with bilateral involvement	1
So severely and suddenly affected in one arm as to be totally dependent	1

In ten patients it was first observed in the thenar and interosseous muscles of the hand. These patients noticed difficulty in gripping and in fine movements. In two patients the weakness and wasting were so severe that a diagnosis of ulnar nerve lesion was considered. They were referred for electromyographic studies and one had his ulnar nerve transplanted before the spinal cyst was diagnosed. The correct diagnosis may be difficult to establish since both conditions can co-exist. Many patients lean

on their elbows in any case and the anaesthesia from the syrinx can mask the pain of the ulnar neuritis.

In five of the ten patients, weakness and wasting was not restricted to the hand alone but progressed to involve the elbow and shoulder muscles. For example, J.B. showed such severe weakness of his whole hand as to render his upper limb virtually useless so that he stopped using a car; as his wife could not drive he became totally housebound. One patient, (J.H.) had bilateral involvement, one hand was weak and then after an interval of 4 years he developed severe weakness in the opposite arm and progressed to the condition of quadriplegic helplessness (described by Barnett). It can be life threatening as in high lesions it can involve the diaphragm.

In seven patients the weakness involved the whole upper limb from the outset. In further patients it did not present in the hand but in one it

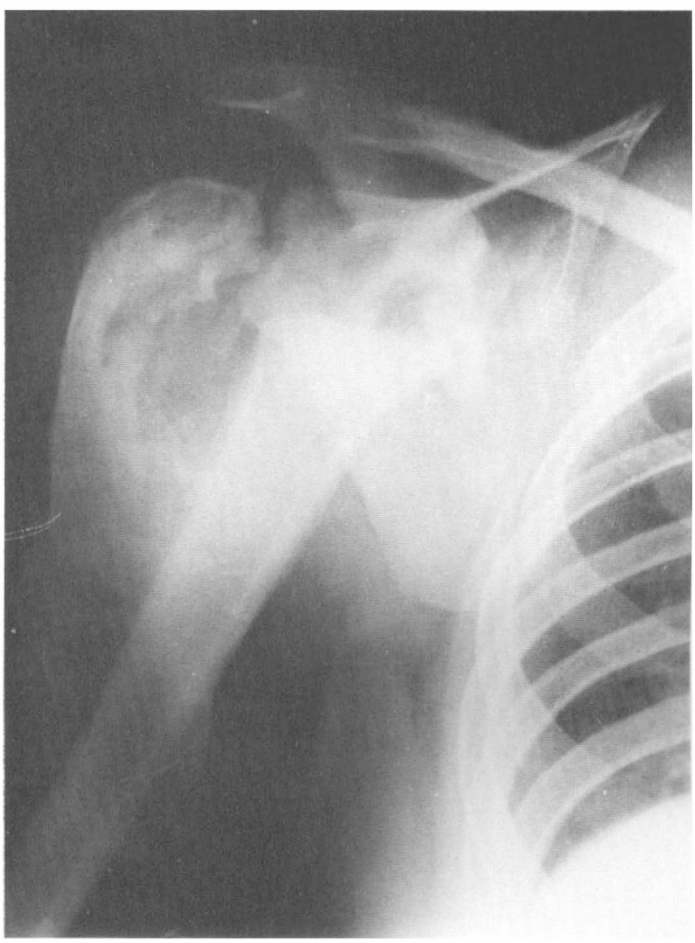

FIG. 4

Severe, painless Charcot joint in D.K.

was localised mainly to the shoulder girdle and in another to the elbow in the triceps muscle. In 6 patients weakness and wasting spread to the other side, often beginning in the small hand muscles.

In patients with a complete cord lesion the weakness appears above the cord lesion, usually in the upper limbs. However, in a patient with a low cord injury the weakness may appear in the lower limbs. R.R. originally had a fracture-dislocation of D12/L1 with a resulting lesion complete below L4. He experienced a deterioration of power initially in his knees and then in his hips. Sensory loss spread up to his neck with pain in his shoulders. He also experienced much pain and spasm in his left knee and ultimately underwent cord section for pain at another hospital. This confirmed the diagnosis of syringomyelia, but unfortunately the pain recurred in the perineum a few years later.

In contrast to the preceding patients with complete cord lesions a small number of patients with incomplete cord transections showed a deterioration in power below the level of the cord lesion. In two patients with bilateral involvement there was also involvement of the lower limbs.

In D.K., an incomplete paraplegic at the D12 level, upper limb weakness of the right side was the first to appear. This predominantly involved the shoulder, resulting in a Charcot joint. (Fig. 4). The left arm was likewise affected but he lost sensation bilaterally below C2. His cyst thus extended above and below the level of the lesion including both sides of the cord, sparing the conus reflexes. In B.H., an ambulant tetraplegic, the weakness involved all four limbs making walking difficult. In this case too there was loss above and below the level of the cord lesion.

Bilateral Involvement

In the vast majority the presentation was unilateral but two patients showed bilateral changes when the condition presented. Other patients rapidly developed bilateral changes. (Table 6). Only twenty-five patients developed this condition. The side that presented first remained the most severely affected. It could occur at any stage. The commonest manifestation was sensory change; the least common was reflex changes and motor loss. It can easily be appreciated why sensory changes are the commonest manifestation in view of the finding of the cyst at the base of the posterior horn of grey matter. Williams (1979) has postulated that in idiopathic syringomylia (by the time the cyst has grown anteriorly large enough to cause motor weakness of a lower motor neurone type) sensory changes should be present on the other side. However, this only occurred in our

TABLE VI

Bilateral Involvement	
Sensory	21
Absent reflex	12
Motor	8
Total with bilateral involvement	25

series in 13 of the patients, approximately half. The others only showed ipsilateral weakness and sensory loss. Conversely, there were a small number of patients (4) who had bilateral sensory loss without showing any motor weakness. The only rational explanation for these findings is that the pathology differs from that found in classical syringomyelia and does not consist of a single large cyst but multiple cysts that intercommunicate and ramify throughout the different parts of the cord. This has been found both by myelography and at post mortem.

Brain Stem Involvement

Only a few patients with brain stem involvement have been described in the literature.

However, some of our patients with proven cysts have had a variety of symptoms from the brain and brain stem which might be attributable to brain stem involvement, such as dysphagia, perversion or loss of taste. Some have had migraine of an unusual type, one has had atypical Bell's palsy and two patients have developed deafness after the cyst manifested itself. However, in both cases this was shown to be peripheral in origin. D.R. had progressive deafness but a C.A.T. scan of the brain excluded syringomyelia, and auditory-evoked potentials showed that this deafness was peripheral and not central in origin. Three patients have had unequivocal clinical evidence of brain stem involvement, in two substantiated by C.T. scans.

G.Y. (Aged 58). He ustained a fracture-dislocation of D4/5 on 11.10.61, when a sack fell on him causing a complete spastic paraplegia below D5. Lumbar puncture showed a complete block. In 1963 he experienced a cold feeling in his right arm followed by 'pins and needles'. Four months later he was referred to a neurosurgeon as he had developed difficulty in speaking. He had nystagmus, preservation of normal sensation over the face, the palate deviated to the left and there was a right laryngeal nerve palsy. He had global weakness of the right arm and there was impairment of all modalities of sensation apart from figure writing in the right arm. At a later stage he developed impairment of sensation in C7 and C8 dermatomes on the left side to two point discrimination. Tendon reflexes were lost in all four limbs. A diagnosis of brain stem ischaemia was made and he was transferred to the National Spinal Injuries Centre, Stoke Mandeville Hospital. In 1969 (aged 65) he died of pneumonia and cardiac failure. Macroscopic section of the brain did not confirm a cyst. Unfortunately, sections were not preserved for histology.

Spinal Cord

The dura and leptomeninges showed an area of scarring extending for 4 cms over D5 segment and partly the segments above and below this. The spinal cord showed atrophy from D2 down to the conus. The anterior spinal artery could be identified throughout most of the cord except where it was obscured by the traumatic scar. Transverse slices through the cord were made at each segmental level. A small syrinx began at C2, situated in the right posterior fossa and neighbouring posterior column. This expanded at C3, involving most of the right posterior column and all of the right posterior horn. The transverse slices caudal to this showed the extent of the syrinx which could be seen as far as D1 segment. D2 appeared to be atrophied with a small trace of syrinx which could also be seen

in D3, D4, D5 and D6 showing atrophied cord segments deformed and associated with connective tissue fibrosis. From D7 down to D11 the cord was atrophied generally but not apparently due to direct trauma. D12 showed some atrophy suggesting gliosis of the cord of the lesion above. The lumbar and sacral cord were normal apart from pyramidal tract degeneration.

Comment. In his case the clinical features were unequivocally those of brain stem involvement. The brain section macroscopically did not show a cyst but this could easily have been missed and, unfortunately, the brain was not preserved for detailed histology.

A further case is presented

In 1976 S.W. aged 18 sustained a complete lesion below D8 as the result of a fracture of his fifth upon sixth thoracic vertebrae due to a motor cycle accident. One year after injury he developed partial anaesthesia of a spino-thalamic type in his left arm between C5 and D8 with minimal weakness of his left hand. As it was causing him little disability he was not investigated further. Four years later he suddenly developed dense sensory loss in the left upper limb with pain in the left shoulder, hiccoughs and dysphagia. Examination showed him to have surprisingly well preserved power in the left upper limb with a paralysed left diaphragm although he had little use of the left arm. He had bilateral depressed corneal reflexes and dense sensory loss involving all modalities including posterior column of the whole of the left upper limb from C2 down to D8. There was

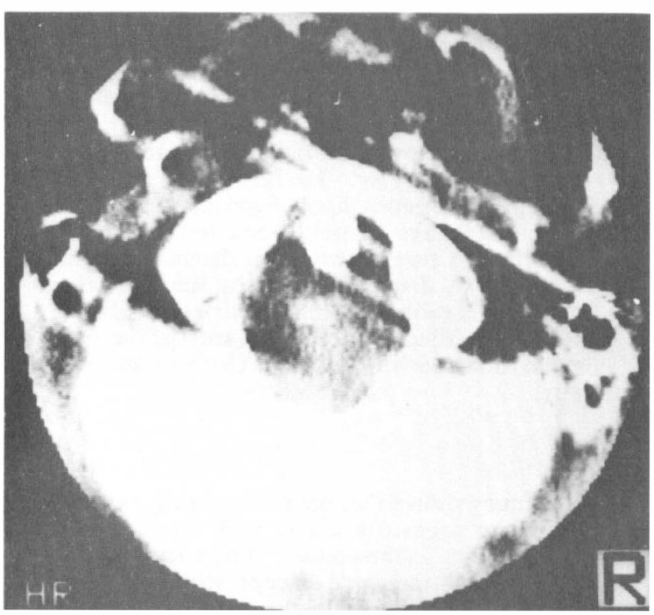

FIG. 5

S.W. Late matrizamide enhanced computerised tomography scan at base of skull, odontoid anteriorly, occipital condyles laterally, basi occiput posteriorly with metrizamide in the syrinx.

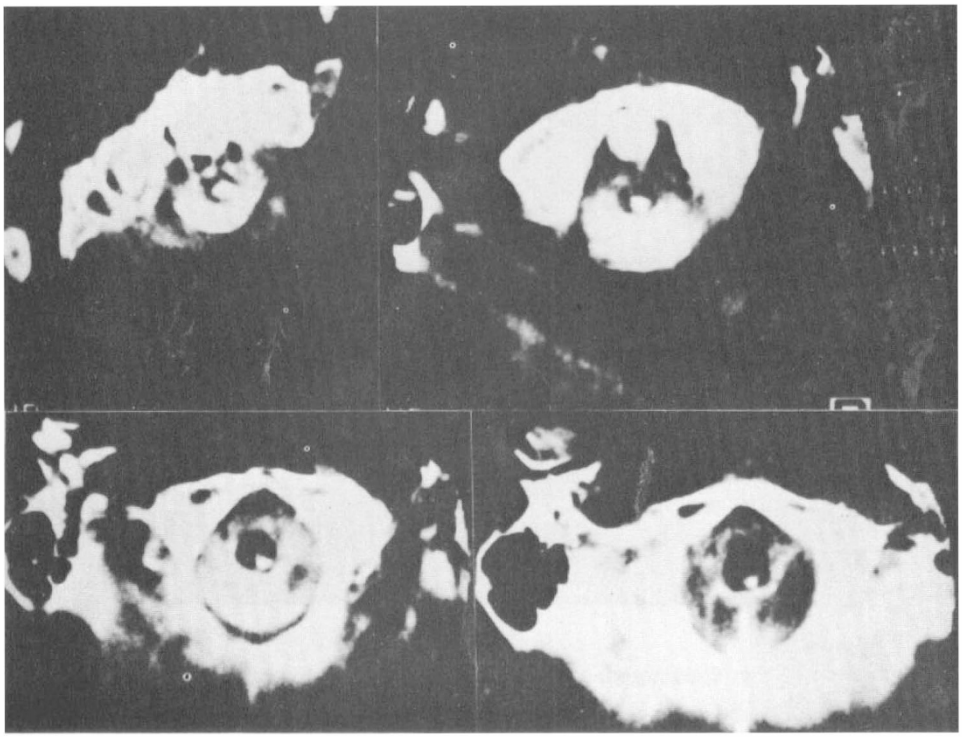

FIG. 6

Late metrizamide enhanced computerised tomography to show the syrinx extending through the foranem magnum into the brain stem.
Top right at the occipital condyles, syrinx clearly visible. Bottom right, through the foranum magnum, the cyst is extending into the brain stem.

tremor of the left arm that was only partially corrected when he concentrated. There was intention tremor. Scan revealed that the upper thoracic cord was dilated with a cyst in the medulla. (Figs. 5, 6 and 7.)

Natural History

There have been only 73 cases described in the literature. The largest series described by a single author until now was 17 cases. (Barnett *et al.*, 1973). The present series of 40 cases does enable us to amplify and delineate the condition more fully.

Conclusions

1. These 40 patients certainly do not represent the total number of patients with this condition since at this centre many patients were lost to follow up and it is weighted by those cases seen more recently when active investigation and exploration has been carried out.

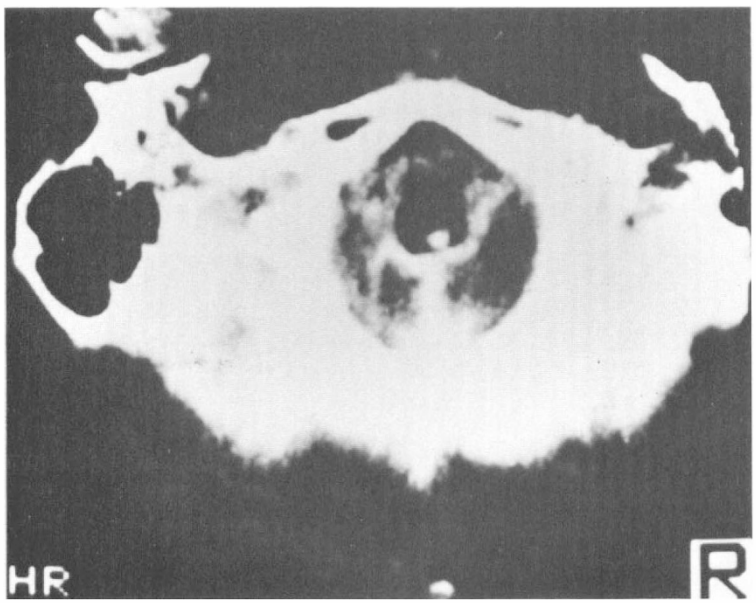

FIG. 7

Bottom right of Fig. 6, enlarged, showing the cyst extending into the brain stem in S.I.

Consequently the progress has been modified by treatment. In many cases it has been arrested but a small number have deteriorated. We have encountered the condition within two months or as long as 30 years after injury.

2. Untreated, all the cases have progressed either slowly or rapidly, some to severe disability. At least three had brain stem involvement. Two progressed to quadraplegic helplessness.

3. There is no way of predicting from the extent, the level, the severity of the injury or its aetiology, how quickly or how slowly the condition will progress, but it does do so and frequently the other side is involved.

4. Secondary changes in the joints are common.

5. From the post-mortem studies it is clear that several cysts may exist and the condition can progress both upwards and downwards confirming the clinical findings.

6. Clinical extension caudally has been found in three patients. In one patient the cyst was demonstrated at post-mortem extending well below the level of the lesion; the other post-mortem showed extensive atrophy of the cord below the fracture site of maximum injury.

7. Remissions, particularly sensory, may take place for as long as 15 years. Surgical treatment does not always improve the condition of the patient and two are known to have deteriorated after surgery.

These findings support the view that at least in some of the patients an injury occurs to the cord not only at the site of maximum trauma but also at a distance from it and as time passes, cystic changes supervene

in those damaged areas and that as the cysts increase in size they compress the adjacent neural tissue causing symptoms and signs.

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Summary

The clinical features of 40 patients with post-traumatic syringomyelia are described. The diagnosis was confirmed in 26 patients by neuroradiological investigations, operation or post-mortem examination. In the remaining 14 it was a clinical diagnosis. There were six cervical, 27 thoracic and seven lumbar patients. The incidence was thought to be at least 1·6% of traumatic injuries. The time of onset was between two months and eighteen years and no difference was found in the time of onset between complete and incomplete lesions. The commonest initial manifestation in 25 patients was pain, sensory loss in eight patients and motor weakness in two patients. The condition was bilateral in 25 and involved the trigeminal territory in 12 patients. Three patients developed brain stem involvement. The natural history of this condition is discussed.

Résumé

On décrit les caractéristiques cliniques des 40 patients avec de la syringomyélie post traumatique. La diagnose était confirmée par des investigations neuro-radiologiques, par des opérations ou par l'autopsie. Les autres 14 cas étaient des diagnoses cliniques. Il y avait 6 patients avec des lésions cervicales, 27 thoraciques oy 7 lombaires. L'incidence était pensée d'être au moins 1·6% des lésions traumatiques. Le tempe d'attaque était entre deux mois et 18 ans. On n'avait pas trouvé une différence entre des lésions complètes et incomplètes dans le temps d'attaque. L'Initiale manifestiation la plus commune était la douleur pour 25 patients, la perte sensoriale pour 8 patients et pour 2 patients la faiblesse motrice. La condition était bilatérale pour 25 patients et 12 patients avaient une implication du territorie du trijumeau. Trois patients développaient une implication du tronc cérébral.

L'histoire naturelle de cette condition est discutée.

Zusammenfassung

Es werden von 40 Patienten die klinischen Merkmale einer postraumatischen Syringomyelie beschrieben. Die Diagnose wurde bei 26 Patienten durch neuro-radiologische Untersuchungamethoden, durch Operation oder Obduktion bestaetigt. Bei den uebrigen 14 Faellen handelt es sich um eine klinische Diagnose. Das Patientengut umfasste 6 cervicale, 27 thoracale und 7 lumbale Laesionen. Man nimmt an, dass das Auftreten mindestens 1·6% der traumatischen Verletsungen susmacht. Die Zeit bis zua Auftreten betrug zwischen 2 Monaten und 18 Jahren. In der Zeit des Auftretens fand man keinen Unterschied zwischen vollständigen und unvollstädigen Laesionen. Die häufigsten anfänglichen Kundgebumgen bei 25 Patienten waren Schmerzen, bei 8 Patienten sensorische Verluste und bei 2 Patienten Schwäche der Motorik. Bei 25 Patienten war der Befall bilateral, bei 12 Patienten war der Trigeminus-Bezirk einbezogen. 3 patienten entwickelten Hirmstammbeteiligung.

Die naturliche Geschichte des Zusandes wird diskutiert.

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