THE EARLY DIAGNOSIS OF TUMOURS OF THE CAUDA EQUINA

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At present low backache, pain in the distribution of the lower lumbar or first sacral roots, or the combination of these symptoms, are so often found to be due to protrusion of an intervertebral disc that other and less frequent causes may be forgotten. The protruded disc often causes symptoms in the form of recurrent lumbago over a period of many years before symptoms of root compression appear, and then as a rule only a single root is affected. There are, however, exceptions to these general rules. The protrusion may cause root compression without preceding backache, and if it be centrally placed may compress the cauda equina. The symptoms may then be hardly distinguishable from those of some other space-occupying lesion such as a tumour. The majority of tumours of the cauda equina are benign and easily removable. They often cause pain and other symptoms for some time before there are any abnormal physical signs, and when such signs are present they may markedly resemble those of a protruded disc.

The purpose of the present paper is to analyse the symptoms and signs in a series of 20 proved cases of cauda equina tumours with special emphasis upon the differential diagnosis from disc protrusion.

Material

The 20 cases have been subdivided as follows:—

(A) <i>Fibrous Encapsulated Tumours</i>			
Neurofibroma	..	..	12
Meningioma	..	..	2
(B) <i>Other Primary Tumours of the Cauda Equina</i>			
Glioma	..	..	3
Ependymoma	..	..	2
Haemangioblastoma	..	..	1

The average age of patients in Group (A) is 43.5 years and is nearly twice that for Group (B) which is 23. The youngest patient in Group (A) was 12 years old and had a neurofibroma. The difference in sex incidence does not appear to be significant.

Symptoms at the Onset

There are no particular symptoms associated with either group of tumours, but the average time before the diagnosis is established is significantly different. This was found to be nearly five years for the encapsulated tumours, as opposed to 21 months for the more invasive types.

Pain.—Pain was the first symptom for 19 of these 20 patients. In only one did the story start with weakness. In half the patients the pain began in the lumbar region. In three it started in both thighs, in two in the knee, and in the others either in the foot, buttock and groin, pubic region, or the back of one or both legs.

The pain may be sudden and severe or begin insidiously. Even when the onset is insidious the pain frequently becomes severe and this was so in four of six such patients. It was then particularly unpleasant and exhausting and was subjectively described as “cutting”, “a deep, unbearable ache”, “enough to make me scream”, “tearing”, “agonizing and causing me to shout out”. The remaining 13 had a sudden, severe onset and in five of these it was initiated by muscular effort. For instance, one patient had a sudden severe pain from the sacral to the mid-thoracic region of the spine which prevented him from moving and which occurred for the first time when he was lifting a heavy weight. A second patient, on rising from a stooping position, experienced a sudden, tearing pain in the medial side of the knee.

In two-thirds of the cases the pain was aggravated by sudden movement and by coughing or sneezing. Ten patients found that to get up and walk about relieved pain which had become worse while they were recumbent. The duration of the initial pain was very variable. In three patients it continued for several weeks and then ceased for a time; in eight, stabs or short bouts of pain initiated a more

continuous phase, while in the remainder it was virtually continuous, but often with severe exacerbations.

Weakness.—The exception referred to in the previous section found that his left leg was gradually becoming weaker and that he tended to lose his balance. It was not until seven years later that the first sensory symptom occurred. This was a sensation of heat in the dorsum of the left foot, to be followed 20 months later by severe, immobilizing pain in the lumbar region on getting out of bed.

Course of the Illness

Though some of the patients had a bout of pain lasting for several weeks and initiated by muscular effort, in none of them did it recur in a similar way; they might be free of pain for as long as a year, but on returning it would be of a more chronic and permanent nature with exacerbations, often provoked by sudden movement or straining.

The onset of weakness was nearly always insidious and might be noticed either at the periphery or in the proximal part of one or both legs. It might appear after two or three months or be delayed for 14 years. Considering each group as a whole, the invasive tumours produced weakness after about 10 months and the encapsulated ones after 40 months.

Although irritation of sensory roots occurred early, an actual sensory deficit would not be noticed until late in the illness, and when it did appear, the area affected was often small. Seven patients complained of numbness of part of one or both feet, one of numbness of the thigh, and another three were not fully aware of the passage of urine and faeces.

Micturition became affected in eight cases. There was either difficulty in passing or in holding urine. This disturbance appeared late in all seven patients with an encapsulated tumour. In none of them did it occur earlier than two years after the onset of the first symptom and on the average six years had elapsed. The remaining patient had a glioma and other symptoms had been present for seven months. At the time of diagnosis all eight patients had marked motor and sensory signs in both legs. The diagnosis was usually quickly arrived at after the onset of disturbed micturition, but in two patients a further two years elapsed before the true cause was determined. A tumour involving nerve roots innervating both legs may spare the more medial roots and not give rise to a disturbance of micturition. In this series there were six such cases, all having bilateral motor signs and three an extensive bilateral sensory loss in addition.

Signs at Diagnosis

The principal physical findings are summarized in Table I. Unfortunately the motor findings do not permit a more detailed analysis.

Examination of the spine is also important, though no abnormality may be detected and straight leg raising may be adequate. Eleven patients had limitation of movement associated with pain and muscle spasm; eight had local tenderness, three without limitation of movement; six patients did not show any abnormality.

Investigations

X-ray Examination of the Spine.—Positive findings were infrequent. Five of the patients with encapsulated tumours showed bony changes, but on the average these had had symptoms for nearly four years. Erosion of the posterior part of the vertebral bodies is shown in Figs. 1 and 3, and flattening of the pedicles in Figs. 2 and 4.

Of the remaining patients, one showed an increase in the interpedicular distance from D12 to L3; this was in a young girl having a short history of three months.

Lumbar Puncture.—The abnormalities concern the manometrics and protein content of the cerebrospinal fluid. They are arranged in Table II. It will be seen at once that the protein content is significantly raised in all except one case even if there is no block.

Myelography.—This investigation was carried out on 14 patients and in every case there was either a complete block or a filling defect. Three of the remaining six patients were amongst those showing bony changes in the plain films.

The patient who had only 60 mg. of protein per 100 ml. in the cerebrospinal fluid showed an oval filling defect opposite the body of L1. This is clearly seen in Fig. 8. Fig. 7 shows another example of a local filling defect, though this is also associated with a complete block at the upper border of L2.

Figs. 3 and 4 show a complete block at the lower margin of D12 and the myodil can be seen to be encircling the upper borders of a neurofibroma. Figs. 5 and 6 show a rather similar picture and there is also an increase in the interpedicular distance with some flattening of the pedicles.

Conditions Simulated by Cauda Equina Tumours

Pain is usually the first symptom causing the patient to seek medical advice, and the greatest difficulty in making a diagnosis is at this stage when

FIGS. 3 and 4.—Neuro-fibroma : cisternal myelogram showing a complete block and outlining the upper margins of the tumour. There is erosion of the posterior aspect of the body of the second lumbar vertebra, an increased interpedicular distance, and flattening of the pedicles of several vertebrae.

FIGS. 1 and 2.—Neuro-fibroma : lateral and antero-posterior projections of lumbar vertebrae showing erosion of the posterior aspect of the bodies, an increased interpedicular distance, and flattening of the pedicles.

FIGS. 5 and 6.—Ependymoma: cisternal myelogram showing a partial block. There is an increase in the interpedicular distance and flattening of the pedicles below this level.

FIG. 7.—Two neurofibromata: lumbar myelogram showing a complete block at the upper border of the second lumbar vertebra and a local filling defect on the left side.

FIG. 8.—Neurofibroma: lumbar myelogram showing a local filling defect at the level of the first lumbar vertebra.

TABLE I
SIGNS AT DIAGNOSIS

Case No.	Pathology	Length of History (years)	Motor Signs	Sensory Roots Involved		Knee and Ankle Reflexes		Sphincters Affected
				R.	L.	R.	L.	
1	Neurofibroma	$\frac{3}{4}$	Slight weakness dorsiflexion R. hallux	—	—	++	+	—
2	Neurofibroma	2	Marked wasting L. buttock and hamstrings; slight weakness inversion and dorsiflexion L. foot	—	—	+	0	—
3	Neurofibroma	1 $\frac{1}{2}$	Slight wasting and weakness quadriceps and dorsiflexors feet; L. worse than R.	L5-S2	L5-S2	0	0	—
4	Neurofibroma	15	Gross wasting and weakness both limbs, R. worse than L.	T10-S5	T11-S5	0	0	Anal, bladder
5	Neurofibroma	9	Slight wasting calves and L. thigh; marked weakness L. limb and below R. knee	S1-S5	L5-S5	+	+	Bladder
6	Neurofibroma	3	Slight wasting both limbs; weakness mainly on knee flexion, dorsiflexion, and eversion ankles	L5-S5	L5-S5	0	0	Anal, bladder
7	Neurofibroma	2 $\frac{1}{2}$	Little movement possible at ankles; marked weakness on knee flexion but slight elsewhere	L4-S5	L4-S5	0	0	Anal, bladder
8	Neurofibroma	10	Slight wasting and weakness L. hip flexors, thigh adductors, and knee extensors	L2-3	L2-3	+	++	—
9	Neurofibroma	7	Marked wasting and weakness R. buttock and below knees	S2-5	L5-S5	+	+	Bladder
10	Neurofibroma	4	Weakness hip flexors and knee extensors; wasting and weakness below knees	L5-S5	L5-S5	0	0	Bladder
11	Neurofibroma	$\frac{1}{2}$	Moderate weakness R. quadriceps, R. anterior and posterior tibial muscles	—	—	++	++	—
12	Neurofibroma	1 $\frac{1}{2}$	Slight wasting and weakness L. quadriceps and below knee	—	L5-S1	+	++	—
13	Meningioma	2 $\frac{1}{2}$	Marked wasting and weakness pelvic girdle and below knees, especially R. calf	L4-S5	L4-S5	+	+	—
14	Meningioma	6 $\frac{1}{2}$	Slight wasting both limbs with moderate generalized weakness	L5-S3 Hyperaesthesia D9-L2	L4-S3 Hyperaesthesia D9-L2	+	0	Bladder
15	Glioma	2	Slight generalized wasting and weakness both limbs, especially R.	L5	—	+	+	—
16	Glioma	1 $\frac{1}{4}$	No wasting; moderate generalized weakness both limbs	S3-5	S3-5	+	+	Anal, bladder
17	Glioma	3	No wasting; moderate generalized weakness both limbs	D11-S5	D11-S5	0	0	—
18	Ependymoma	$\frac{1}{4}$	Slight weakness hip flexors and knee extensors	—	—	0	0	—
19	Ependymoma	$\frac{1}{2}$	Moderate wasting and weakness R. buttock and limb, excluding the calf	L3-5	—	0	+	—
20	Haemangioblastoma	3 $\frac{1}{2}$	Slight wasting below knees; movement ankle and toes absent on R. nearly on L.	—	L4-S3	0	0	Anal

there are usually no neurological abnormalities.

The backache may suggest the possibility of a local structural defect. A typical case history is as follows:—

Case 19.—P. H. was awakened from sleep by a sudden, sharp, and severe pain in the lumbar region which was aggravated by moving; this lasted half an hour and did not recur for another month. On this occasion he was running to parade when a similar pain was experienced

in the same site. This necessitated walking slowly, doubling up, and avoiding any sudden strain. A diagnosis of "spina bifida" was made at a neurosurgical unit, but six months later an inoperable ependymoma was revealed.

Fibrositis is still a popular explanation of these cases of backache in spite of the publicity given to prolapsed intervertebral discs. The latter diagnosis is more likely to be suggested when there is radiation

of the pain into one or both legs and was made in both of the following cases :—

Case 13.—L. B. developed a severe, sharp, cutting pain in the lumbo-sacral region which was worse on coughing and movement. Treatment comprised diathermy, massage, mud packs, baths, and subsequently manipulation three months after the onset of definite weakness of the legs.

Case 3.—J. P. developed a dull ache in the lumbar region after hyperextending her back at school. This did not subside and a plaster jacket was fitted. Nine months elapsed before foot drop developed ; a further six months passed before the diagnosis was made.

The backache may be interpreted as a referred pain from visceral disease but even when this is present it is not necessarily the cause of the symptoms as in Cases 6 and 14.

Case 6.—E. J. had severe pain in the left lumbar region and this continued in spite of the removal of a cystic kidney. She then had a slight vaginal discharge. Curettage under general anaesthesia revealed nothing serious, but subsequently there was great difficulty in walking. It was a further two and a half years before the explanation for this pain was discovered.

Case 14.—A. S. developed a continuous ache in the left loin, and there were exacerbations preventing sleep. Relief was obtained by getting out of bed and walking. Three years later a hysterectomy was performed but this had no effect on the pain, which was now beginning to radiate down the back of the legs. After a further three years colpo-procto-perineorrhaphy was carried out without effect. A few months later the legs became numb, micturition was affected, and the pain became exceedingly severe at this juncture ; diagnosis was finally established.

Root pain without backache may suggest some local cause in the legs as in Case 8.

Case 8.—M. D. had a sudden, severe, tearing pain in the inner side of the left knee when straightening up from a bending position. She rubbed the knee for about half an hour, at the end of which time the pain and difficulty in moving the leg had disappeared and she continued her work. These attacks occurred periodically for the next seven years and gradually began to trouble her at night, awaking her from sleep. The pain also began to spread up the thigh into the buttock and to localize there permanently. Treatment included two courses of physiotherapy, a vaccine, and extraction of all teeth before the diagnosis was made three years later.

Persistence of an unexplained pain without abnormal signs may lead to the diagnosis of a neurosis as in Case 2.

Case 2.—F. W. started to experience pain in the left buttock and groin two years before the diagnosis was

made. It was described as a “ kind of drawing pain ” and would radiate down the thigh and the outer aspect of the leg into the toes. Initially it was intermittent and tended to come on after walking or stooping ; it was aggravated by coughing, sneezing, or straining at stool. A year later the patient noticed that the left thigh was becoming weaker but there was still no wasting. She was admitted to hospital, the condition was said to be functional, and electric convulsion therapy was administered. She continued to deteriorate while a course of 10 shocks was administered.

It is possible for local trauma or infection to produce very suggestive symptoms and signs. This will be discussed more fully later, but the following case illustrates the difficulties.

Case 4.—S. S. had had intermittent pain in the back and thigh for 13 years before he started to have some difficulty in passing water. Absence of ankle jerks was thought to be due to peripheral neuritis and the prostate was cauterized. After a temporary improvement, his condition worsened ; this deterioration was said to be due to the spinal anaesthetic.

Discussion

Compression of the nervous structures within the spinal canal is suggested by the following features.

Pain.—Pain in the lumbar region is often the presenting symptom but it may occur in root distribution in one or both legs. It is often severe and aggravated by movement and coughing. This type of pain may occur with either a prolapsed intervertebral disc or with a cauda equina tumour, but it is often particularly unpleasant in the latter and unrelieved by complete rest. Patients with tumours are often more comfortable while walking about than when resting in bed.

The mode of onset does not help. O'Connell (1943) states that over 50% of his cases with a prolapsed lumbar intervertebral disc had an onset associated with trauma. In 25% of the patients with cauda equina tumours the pain started during muscular effort. The occurrence of recurrent episodes of sudden, immobilizing pain brought on by effort is much more suggestive of a prolapsed disc. As already mentioned, none of the patients in this paper had a second episode of this kind, though such cases have been described. Pomeranz (1945) records a case in which, for five years, a man had recurring attacks of low back pain brought on by lifting and made worse by movement or jarring.

Paraesthesiae.—These subjective sensory disturbances are common with both prolapsed discs and cauda equina tumours as they are produced by the

irritation of nerve roots. Extensive numbness is not found except with a large central prolapse or in the late stages of tumour compression.

Weakness.—Pain will prevent a person from using the limb properly, but actual weakness immediately suggests that there is interference with nerve pathways. There is no characteristic picture here, but motor symptoms are relatively uncommon with prolapsed lumbar discs (O'Connell, 1943). Ultimately they invariably occur with tumours but the aim is to make the diagnosis before there is any serious involvement of motor function.

Disturbance of Micturition.—This may occur with any form of pressure on the sacral nerve roots. It is a late occurrence with tumours and does not necessarily appear when both legs are involved. It is relatively uncommon with disc protrusions, but the extrusion of a large part of a disc may cause paraplegia and incontinence of urine and faeces. The story of characteristic, intermittent pain with a sudden serious deterioration is very suggestive of a disc lesion, but the precise diagnosis may be impossible until operation. Dandy (1942) drew attention to the need for urgent surgical treatment of patients with a large central protrusion, and Ver Bruggen (1945) fully described the clinical picture presented by nine such cases.

Abnormal Neurological Signs.—As will have been seen in the analysis of the cases presented here, abnormal signs may be absent, confined to the territory of one nerve root, or be very extensive. There is no specific arrangement of abnormalities typical of cauda equina tumours, but the presence of very severe pain accompanied by little in the way of signs should arouse suspicion.

Methods of Investigation.—In the investigation of these patients the following methods are of value.

Plain X-ray Films.—Early positive findings are infrequent, but erosion of the bodies and pedicles of the lumbar vertebrae may be visible. Any local structural abnormality, or narrowing of a disc space suggestive of a prolapse, may only be incidental. It is also important to exclude any gross bony disease.

Lumbar Puncture.—If the puncture is below a block abnormal manometrics will be obtained, but the protein content of the fluid is much more important for this may be raised, even above the block. Attention was drawn to this as long ago as 1923 by Cushing and Ayer who reported five cases with xanthochromia and excess protein above a cauda equina tumour. As will be seen in Table II,

TABLE II
MANOMETRICS AND C.S.F. FINDINGS

Case Number	Pathology	Degree of Block	Protein (mg. per 100 ml.)	Length of History (years)
1	Neurofibroma	Complete	10,000	$\frac{3}{4}$
2	"	Partial	200	2
3	"	Complete	2,500	$1\frac{1}{2}$
4	"	No record	No record	15
5	"	None	12 ⁺	9
6	"	Complete	3,800	3
7	"	Partial	6,400	$2\frac{1}{2}$
8	"	Complete	500+	10
9	"	None	150	7
10	"	No record	4,000	4
11	"	Partial	1,200	$\frac{1}{2}$
12	"	None	60	$1\frac{1}{2}$
13	Meningioma	Partial	500	$2\frac{1}{2}$
14	"	Puncture above lesion	190	$6\frac{1}{2}$
15	Glioma	Partial	3,200	2
16	"	Partial	260	$1\frac{1}{2}$
17	"	Partial	5,000	3
18	Ependymoma	Complete	5,000	$\frac{1}{2}$
19	"	Complete	3,200	$\frac{1}{2}$
20	Haemangioblastoma	Partial	160	$3\frac{1}{2}$

the protein is markedly increased in all but one case. Love (1944), when discussing the differential diagnosis of intraspinal tumours and protruded discs, emphasizes this point and says that at the Mayo Clinic it is very rare for the protein to be higher than 100 mg. per 100 ml. with a disc alone.

Inflammatory lesions of the cauda equina do occur, but most of the papers on the subject were written before the prolapsed intervertebral disc was a well recognized clinical entity. Elsberg and Constable (1930) analysed 45 cases with cauda equina lesions. Twenty-eight of these had tumours and 17 had congested, swollen roots. Twelve of the latter underwent lumbar puncture and in all the protein level was normal.

Cramer (1934) studied the records of 26 patients who were said to have cauda equina radiculitis, 15 being verified surgically. He admits, however, that 69% had local arthritic changes of such a degree as to suggest a direct relationship to the symptoms of root compression. In another 15% other pathological conditions were discovered at a later date, leaving 15% unaccounted for and possibly true examples of a myeloradiculitis. The cerebrospinal fluid protein level was often normal and usually lay between 50 and 80 mg. per 100 ml. The highest figure is not disclosed.

A high protein level may be found with spinal arachnoiditis. Elkington (1936) described the clinical and operative findings in 41 cases, five of which showed signs of disorder of the lumbosacral enlargement or of the cauda equina. He concluded that a marked increase in protein only occurred as a result of occlusion of the subarachnoid space.

Myelography.—This has become an essential part of the investigation of patients with symptoms and signs suggestive of spinal compression. When it is fairly certain that operation will be required, it is advisable to carry out the myelography on the same day. "Myodil" itself is rarely harmful but the alteration of the pressure within the canal may lead to a sudden deterioration in the condition of the patient.

Toumey, Poppen, and Hurley (1950) felt that the single most important factor in the diagnosis of cauda equina lesions was the myelogram, and held that it should be used as a routine in all cases admitted for surgical treatment of a suspected disc lesion. Apart from a few examples, they do not give any details but at the same meeting Ford and Key (1950) analysed the myelographic findings in 206 patients who had been operated on for low back pain. The findings were accurate in 149, and in another 17 a mid-line disc protrusion had been indicated instead of a lateral one. This left 40 with major discrepancies, 10 having a disc protrusion at another level, 27 a lesion in spite of a normal myelogram, and three no lesion but a positive myelogram.

Dodge, Svien, Camp, and Craig (1951) at a discussion on tumours without neurological manifestations but producing low back and sciatic pain stressed the importance of running the oil as high as D8, for there had been several patients at the Mayo Clinic with a tumour higher than a lumbar disc protrusion.

Results.—The importance of early diagnosis was emphasized by Allen (1930) after reviewing the literature and his own cases. This is clearly seen when investigating the eventual fate of the patients discussed in this paper.

The complete removal of the tumour is usually possible when it is a neurofibroma even if the history is a fairly long one. Of the 12 patients with this type of lesion, 10 had it completely removed and eight made a good recovery; one did not improve and one died from a pulmonary embolus.

With any other type of tumour early diagnosis becomes of paramount importance. Even with a meningioma, complete removal was impossible

after two and a half years. Of the remaining patients, only one did well; she had an ependymoma completely removed after a short history of three months. After seven months the other case with an ependymoma had become inoperable.

Summary and Conclusions

The abnormal signs on examination and the development of symptoms in 20 patients with a surgically verified cauda equina tumour are outlined.

Details are given of the cerebrospinal fluid findings and the results of straight x-ray and myelographic examination. A high protein content of the cerebrospinal fluid is an important early change.

Difficulties in differential diagnosis are discussed and illustrated with examples.

Pain is the most important early symptom and its characteristic features should arouse suspicion. Abnormal neurological signs may be absent or long delayed and the picture may become obscured by psychological difficulties.

A tumour of the cauda equina should always be considered as a possible diagnosis in a case of low back pain which cannot certainly be ascribed to another cause.

I wish to thank the Medical Committee of the National Hospital for permission to use the hospital case records and Drs. Hugh Davies and E. Rohan Williams for the radiographs. I am particularly indebted to Sir Charles Symonds for his constant encouragement and helpful interest.

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