Tumours of the cauda equina

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SUMMARY A retrospective study of 70 consecutive patients with a cauda equina tumour who were admitted to the Neurosurgical Department at the Radcliffe Infirmary, Oxford is presented. The diagnosis of these tumours is often difficult and delayed. The quality of life largely depends upon the neurological disability at presentation. The diagnostic features and investigations are discussed together with the treatment and prognosis.

Patients with pain in the back or lower limbs are commonly seen in hospital or family practice. While many of these are suffering from lumbar spondylosis or a prolapsed intervertebral disc, a small but important group harbours a tumour of the cauda equina. There can be few other tumours which present in such a varied and subtle fashion to so many different specialists. While generally benign and curable by surgery, they are often unrecognised for many years, condemning the unfortunate patient to a life of pain and of misery. It is our purpose in this paper to examine the clinical features and investigations leading to the diagnosis of these tumours together with the treatment and outcome.

Methods

Seventy consecutive patients admitted to the Neurosurgical Department at the Radcliffe Infirmary, Oxford over a 40 year period and having a tumour of the cauda equina were reviewed. Only those with wholly intrathecal disease were included. This accounts for the small proportion of patients with metastatic disease.

The degree of postoperative disability was assessed by placing patients in one of four groups: group 1— asymptomatic and in full-time employment; group 2— minimal symptoms or sphincter disturbance; group 3— significantly disabled and partially dependent with severe sphincter disturbance; and group 4— severely disabled or paraplegic, or with an indwelling urinary catheter.

Results

INCIDENCE

The diagnostic groups are shown in Table 1. Five of the 29 patients with neurofibromas had evidence of generalised neurofibromatosis. Four patients with metastatic disease had primary tumours within the central nervous system (two medulloblastomas and two oligodendrogliomas). The other four patients had tumours with the primary outside the central nervous system, two of these primary tumours in the breast and one in the lung. One patient had a reticulum cell sarcoma of the parotid gland and two years later developed back pain. Both ependymomas and neurofibromas were more common in males, neurofibromas showing a particular male predisposition as noted by Rubinstein (1972). The age distribution of the patients is indicated in Fig. 1. While there were no children under 10 years of age in the series, all other age groups were represented. Onset was during the fourth and fifth decades in almost half the patients.

LENGTH OF HISTORY

Forty-one of the patients had histories longer than two years and in over half of these, longer than

Table 1 Incidence and sex distribution of 70 patients with tumours of the cauda equina

<table>
<thead>
<tr>
<th>Diagnostic group</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ependymoma</td>
<td>18</td>
<td>12</td>
<td>30</td>
</tr>
<tr>
<td>Neurofibroma</td>
<td>20</td>
<td>9</td>
<td>29</td>
</tr>
<tr>
<td>Metastasis and lymphoma</td>
<td>4</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Meningioma</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Lipoma</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>44</td>
<td>26</td>
<td>70</td>
</tr>
</tbody>
</table>

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Patients presented with one of six groups of symptoms (Table 2). Pain was the most common complaint. Approximately equal numbers had back pain (20) and back pain with sciatica (26). Eleven patients suffered with sciatica only. Thirty-five patients complained of pain in recumbency—that is, nocturnal pain.

Case report (RI No 581452)
A 54 year old male complained of back pain and right sided sciatica for 18 months. It had started with a jolt while riding over a rough road in Africa and became progressively severe, necessitating his return to England. A particular feature was pain in recumbency and he was woken regularly at 0200 or 0300 each morning. Relief was obtained by getting up, walking around, and eventually sleeping in a chair for the rest of the night. He had undergone various treatments including plaster of Paris jackets, continuous and intermittent traction, manipulations, a series of 'spinal injections' and prolonged periods

Table 2  Main symptoms at presentation in 70 patients with tumours of the cauda equina

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Back pain and/or sciatica</td>
<td>57</td>
</tr>
<tr>
<td>Painless weakness of the legs</td>
<td>6</td>
</tr>
<tr>
<td>Sphincter disturbance</td>
<td>3</td>
</tr>
<tr>
<td>Papilloedema and raised intracranial pressure</td>
<td>2</td>
</tr>
<tr>
<td>Subarachnoid haemorrhage</td>
<td>1</td>
</tr>
<tr>
<td>Sensory disturbance</td>
<td>1</td>
</tr>
</tbody>
</table>

Fig. 1  Age at presentation of patients with cauda equina tumours

five years (Fig. 2). Those with metastatic disease had short histories, all less than one year. Patients with ependymomas had particularly long histories, and of the 30 patients, 22 had histories longer than two years.

Fig. 2  Length of history in patients with cauda equina tumours.
Papilloedema and raised intracranial pressure were seen in only two patients. Each had other complaints at presentation.

**Case report (RI No 3097)**
A 27 year old male RAF sergeant was seen by Professor Cairns in the summer of 1939. There had been episodes of back pain and bilateral sciatica since 1937, and he had occasional urinary incontinence. Pain in recumbency had been a prominent feature of his story. While being repatriated from India by ship in February 1938, he developed severe headache and lost consciousness for 10 days, apparently recovering without ill effect. He had noted some weakness of the legs but was able to walk six miles in December 1938. By July 1939, he had developed severe frontal headache and blurring of vision. The headaches were worst in the morning and caused him to vomit, these being the reason for his referral.

Examination showed bilateral papilloedema with retinal haemorrhages. The lower limbs were wasted, weak, and areflexic. There was a vague bilateral sensory loss at T10 dermatome, and urinary retention was present. A complete manometric block was found on lumbar puncture and the protein content of the cerebrospinal fluid was 6g/l. The patient died of septicaemia while in hospital and no operation was performed. Necropsy revealed a haemorrhagic tumour in the cauda equina, extending from the conus to the fourth lumbar vertebra. There were many adhesions between the nerve roots, the dura mater, and the tumour. Histology showed a meningioma with evidence of old and recent haemorrhage. The cranial contents were normal and there was no ventricular enlargement.

Spinal subarachnoid haemorrhage was seen in only one patient.

**Case report (RI No 419662)**
A 19 year old female was admitted because of severe sacral and coccygeal pain and bilateral sciatica. There had been two episodes of back pain over the previous 18 months but each had settled with 10 days' bed rest. On this occasion, the pain had begun during the night, waking her from a sound sleep. Several hours later she developed headache, neck stiffness, and vomiting. Examination showed tenderness to percussion over the lumbar spine. The lumbar lordosis was absent and all movements of the lumbar spine were limited by pain. Neck stiffness and limited straight leg raising were present. There were no abnormal physical signs in the legs. A lumbar puncture revealed uniformly blood-stained cerebrospinal fluid, the supernatant of which was xanthochromic and the protein content was 10 g/l. A myelogram showed a filling defect opposite the body of L3 vertebra. An ependymoma, the upper pole of which was haemorrhagic, was completely removed at operation.
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One patient presented with a curious sensory ataxia.

Case report (RI No 394050)
A 73 year old male was seen with the complaint of unsteadiness of gait for six months. For three months his legs had become slightly weaker, and he had suffered several episodes of acute urinary retention, each requiring hospital admission and catheterisation. Examination showed absent vibration and joint position sense in the lower limbs with preservation of touch, pain, and temperature sensation. There was slight weakness of dorsiflexion of the left ankle but power was otherwise normal. There was no limb ataxia. He was quite unable to stand unsupported and was worse with closed eyes. On rectal examination, the prostate was only moderately enlarged. In view of the ataxia, he was considered to have degenerative cerebellar pathology. A lumbar puncture revealed a protein content of the cerebrospinal fluid of 7 g/l, and there was no manometric block. Myelography, however, showed a complete block at L1 vertebral level. A WR on blood and CSF were normal. At laminectomy, a hard, neurofibroma containing calcium and lying in the roots of the cauda equina and compressing the lowermost part of the conus medullaris was removed. Six days after operation joint position sense and vibration sense were normal in the lower limbs and the patient could walk unaided.

By the time patients presented to the neurosurgeon, there was usually a combination of symptoms of weakness, sensory disturbance, or sphincter disturbance, but pain was the most common.

There was no characteristic pattern of physical findings in the spine. Limitation of movement was the most common sign and was seen in 28 of the 70 patients. Limitation of straight leg raising (17), loss of the lumbar lordosis (12), scoliosis (10), and tenderness on vertebral percussion (seven) were less common. Similarly, there was no characteristic pattern of physical signs in the legs. Some of the patients had no signs, some had one or two nerve roots involved, and some had involvement of many. In five of the patients the physical signs were confined entirely to the lumbar spine. There was often no correlation between motor and sensory signs. However, bilateral motor signs tended to accompany bilateral sensory signs and together with sphincter disturbance were seen most commonly in patients with ependymomas (17 of 30).

All patients had plain radiographs of the lumbar spine and abnormalities were present in 18. The most common finding was of a widened interpedicular distance (14 patients). Vertebral body erosion was seen in six, laminar erosion in two, and a cavitated sacrum produced by a giant intrathecal neurofibroma in one.

Lumbar puncture was attempted at least once in 69 patients. In 23 of these, a dry or bloody tap was obtained on one or more occasions.

The cerebrospinal fluid was examined in 61 patients (Fig. 3). The protein content was usually raised, in most cases over 5 g/l. It is of importance, however, that in three patients the levels were normal, and nine had levels of between 0.5 g/l and 1.0 g/l.

It was possible to correlate manometric and myelographic block in 41 patients. The differentiation between 'partial' and 'complete' manometric block was often difficult, so that a block was described as either present or absent. Twenty-seven patients had a manometric block noted, and all of these had a myelographic block. In 13 of the remaining patients, a manometric block was absent, and all of these showed a myelographic block. In one patient with no manometric block, a capacious sac and small 'suspicious' filling defect was found at myelography. A small neurofibroma was removed at surgery.

Fifty-nine patients underwent lumbar or cisternal myelography and in all 59 a block or filling defect was found.

![Fig. 3 Protein content of cerebrospinal fluid in patients with cauda equina tumours.](http://jnnp.bmj.com/)

INVESTIGATIONS
The relevant investigations were plain radiographs of the lumbar spine, lumbar puncture and cerebrospinal fluid investigation, and myelography.
TREATMENT
Sixty-eight patients underwent surgery. One patient with a meningioma died from septicaemia before operation, and one patient died during induction of anaesthesia. Two patients died in the immediate postoperative period.

A ‘complete’ naked eye removal was performed in 42 of the 68 patients. Of the 30 patients with ependymoma, however, complete removal was possible in only 13. The remaining 17 patients had incomplete removals and were treated with radiotherapy. One of these was lost to follow-up. In one patient with a large neurofibroma causing sacral erosion, complete removal was not possible but no symptomatic recurrence occurred. Complete removal was not possible in seven of the eight patients with metastatic disease.

Urinary retention was the most common postoperative complication and was seen in 21 patients. All required an indwelling catheter and five of these developed urinary tract infections. In three patients the urinary symptoms became permanent after surgery. Transient increase in motor weakness occurred in two and sensory loss in five patients. These changes were noticed in the first few days after surgery and all had disappeared by two months.

PROGNOSIS
Sixty-four of the 68 patients undergoing surgery were followed for periods between two months and 27 years. Four patients were excluded, two being lost to follow-up and two dying in the postoperative period. The deaths were from septicaemia due to pneumonia and from the intracranial effects of a primary hypothalamic oligodendroglioma. The average length of follow-up of the whole series was two years.

Forty patients having ‘complete’, naked eye removals of their tumour were followed. Three of these had recurrences requiring further surgery. One had a histologically ‘active’ neurofibroma and the other developed multiple ependymomas, disseminated throughout the cerebrospinal fluid, seven years after the original laminectomy. The third patient had a metastatic medulloblastoma. Table 3 indicates the frequency of recurrence.

Of the 16 patients with incompletely removed ependymomas, five had symptomatic recurrences requiring further treatment. These recurred at 12 and 18 months and at two, six, and nine years after the first operation. Then they were treated with either surgery or, if severely disabled, with radiotherapy only, each with minimal or no improvement. The average length of follow-up for these patients with incompletely removed ependy-

<table>
<thead>
<tr>
<th>Tumour</th>
<th>Removal complete</th>
<th>Removal incomplete</th>
<th>Symptomatic recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ependymoma</td>
<td>13</td>
<td>16</td>
<td>1</td>
</tr>
<tr>
<td>(29) Neurofibroma</td>
<td>24</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Metastasis</td>
<td>1</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>(7) Meningioma</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>(1) Lipoma</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

momias was 57 months. Two patients each with a medulloblastoma developed a recurrence, two and five years after the original laminectomy. Each benefited from further surgery and postoperative radiotherapy.

The degree of disability in the surviving patients is shown in Fig. 4. Over half were in groups 1 and 2, these being mainly patients with neurofibromas and little neurological disability. As a group, however, those with ependymomas were more disabled. Of the 29 patients with ependymomas who were followed, 15 were in groups 3 or 4, and of these 15, 10 had incomplete removals.

The prognosis for sphincter involvement was bad. Thirty-two of the 37 patients with preoperative sphincter involvement were followed and only seven improved. Twenty-two did not improve and three deteriorated after surgery.

**Table 3**  Frequency of symptomatic recurrence in followed patients with cauda equina tumours (64 patients)

![Fig. 4 Degree of postoperative disability in 64 patients with cauda equina tumours.](http://jnnp.bmj.com/)

**Discussion**

The protean nature of the early course of these tumours meant that many patients had histories
longer than two years and often longer than five. Allen (1930) reported a range of three months to 20 years in his patients, with an average length of four years. Milnes (1953) found the average time from onset until diagnosis was five years for encapsulated tumours, and 21 months for the more invasive types.

Pain was the most common presenting symptom, and while in some early cases it fluctuated in intensity, the tendency was for it to become constant and progressive. Pain in recumbency was a feature recorded in half the patients and deserved emphasis and special enquiry. Typically they were awakened at 0200 or 0300 and forced to get up and walk around to get relief, often sleeping the rest of the night in a chair. Box (1903) first described pain in recumbency, and it was noted also by Allen (1930), Scott (1966), and Rasmussen et al. (1940). Allen (1930) suggested that recumbency causes a lengthening of the lumbosacral canal due to a decreased lumbar lordosis, thus causing increased tension in the caudal nerve roots. An alternative hypothesis of relaxation of the paraspinal muscles during sleep was suggested by Falconer (1966). Rasmussen et al. (1940) considered a ball-valve action of the tumour being forced down by periods of raised cerebrospinal fluid pressure during sleep to be the cause.

The pain is unlike that of intervertebral disc prolapse in which exacerbations and remissions over a period of months or years are usual. Whereas patients with disc disease may be wakened at night by pain, they rarely get up and walk around or sleep in a chair. An occasionally helpful point is that the pain of a tumour seems to be worsened by jolting or jarring rather than bending or twisting.

Campbell (1963) found painless weakness and wasting of one or both legs to be associated with a tumour near the conus medullaris. In all six patients in the present series, the tumour was situated anterior to the conus medullaris and compressing the ventral roots, sparing the posterior roots. While this is the likely mechanism of motor deficit without sensory loss, Campbell (1963) suggested compression of the radicular arteries. As illustrated by Milnes' patient (Milnes, 1953), the natural history of this group seems to be the development of sensory loss and pain, as the tumour causes progressive distortion.

Disorders of sphincter function may precede neurological deficit by long periods. When examined, each of the three patients had a saddle sacral sensory loss, a patulous anus, and absent anal reflex. Two had an ependymoma and one patient had a neurofibroma. Garfield and Lytle (1970) emphasised the value of the finding of a patulous anus in the early diagnosis of these tumours. Urinary frequency with an inability to empty the bladder completely is the most usual complaint, eventually progressing to urinary retention. Anal incontinence is rather less common. Milnes (1953) and Norstrom et al. (1961) each considered sphincter disturbance as a part of the clinical course to be common, and it occurred in over half the patients in this series.

Presentation with headache and papilloedema was rare. Each of the two patients in the series had a history of back pain and some sphincter disturbance. In the operated patient, cerebrospinal fluid obtained at cisternal puncture was xanthochromic with a protein content of 0.6 g/l. At operation, there was evidence that the tumour had bled. The development of a communicating hydrocephalus after a haemorrhage from the tumour may be the mechanism of papilloedema in this case. The protein content of the cerebrospinal fluid in Professor Cairns's unoperated case was 6 g/l.

Love et al. (1931) described three cases of low spinal tumour presenting with symptoms of raised intracranial pressure. Two of the cases had cauda equina ependymomas and one had an intramedullary glioma of the lower thoracic spinal cord. They commented that papilloedema in two of the cases was due to the development of hydrocephalus, demonstrated by ventriculography, but that papilloedema could occasionally be due to an increase in the protein content of the cerebrospinal fluid alone. Gardner et al. (1954) considered that the communicating hydrocephalus was the direct result of a raised protein content, causing a block in the absorption of the cerebrospinal fluid.

Sudden onset of low back pain and sciatica with meningism should alert the clinician to a spinal subarachnoid haemorrhage. Nassar and Correll (1968) in adding four cases to the literature of 11, noted a male preponderance and that most patients were young, in the second or third decades. Meningeal signs were predominant. They found evidence of raised intracranial pressure in 10 patients and six of these had papilloedema. There seems little doubt that bleeding from the tumour may be related to the development of raised intracranial pressure and papilloedema, usually due to a communicating hydrocephalus.

Ataxia of a sensory nature must be extremely rare, and no parallel case could be found in the literature. If it were not for the immediate discovery of function postoperatively, some cause other than the tumour would be suspected. Allen (1930) considered that posterior column function
was never affected without previous involvement of pain, temperature, or touch sensation.

Plain radiographic abnormalities are unusual. The interpedicular distance should be measured with a rule, and Falconer (1966) considered distances of greater than 3 mm between consecutive vertebrae to be significant. Jefferson (1955) reported that abnormalities can occur as congenital defects. Scalloping of the posterior surfaces of the vertebrae is occasionally present, but Miles et al. (1969) noted that this does not necessarily indicate a neoplasm.

A high incidence of dry or bloody lumbar punctures was obtained, confirming that an unsuccessful procedure is not necessarily the fault of the operator. This was particularly noted in patients with ependymoma, a reflection of the size of these tumours. Jennett (1956) reported that failed lumbar puncture may also occur with a large central disc prolapse.

The protein content of the cerebrospinal fluid was usually raised. Cushing and Ayer (1923) found that this occurs even if the lumbar puncture is performed above the tumour. In three patients, the cerebrospinal fluid protein was normal (Fig. 3), and nine had marginally raised levels. A normal cerebrospinal fluid protein level, then, does not exclude a tumour, and differentiation of a tumour from prolapsed intervertebral disc cannot be made on the protein level in the cerebrospinal fluid.

While manometric block is difficult to assess, all patients in which it was found had a myelographic block. However, the absence of a manometric block in no way negates the diagnosis of tumour. The presence of a block depends on the size of the tumour and the level of the lumbar puncture.

Myelography is an essential investigation. In all cases in which it was performed, the correct anatomical level was indicated and the indication for operation confirmed. It will not, however, give the likely pathological diagnosis, and Jennett (1956) found it may not differentiate tumours from central intervertebral disc prolapse. Surgeons tackling such lesions must be prepared to remove an intrathecal tumour.

The treatment of choice is surgical and where complete macroscopic removal of the tumour was possible, the prognosis was excellent and recurrence unusual.

Two-thirds of patients with incompletely removed ependymomas and who were treated with radiotherapy did well and required no further treatment. The remainder developed recurrent symptoms. While it is impossible to know whether radiotherapy has cured any patient with an incompletely removed ependymoma, it would seem advisable to give radiotherapy in these circumstances. Certainly, it was found to be useful in palliation where pain was a problem. Rasmussen et al. (1940) and Van Duinen (1976) have advocated total excision of these large ependymomas where possible, rather than radiotherapy, but the results of this series suggest that conservative surgery with postoperative radiotherapy produces acceptable results. Rubinstein and Logan (1970) reported that metastasis outside the nervous system may occur with caudal ependymomas, but none was noted in this series. The treatment of intradural metastatic disease was generally unsatisfactory in that local recurrence was usual despite surgical removal and radiotherapy. Radiotherapy was of value for the relief of the pain of intradural metastatic disease.

The amount of residual disability was directly related to the clinical picture at presentation. Patients with pain and minimal signs, as commonly seen in neurofibromas, did well and were able to return to work early. However, patients with ependymomas, while surviving well, were a more disabled group (Fig. 4). The quality of survival was related largely to sphincter complications and muscle weakness. The prognosis for sphincter disturbance was bad and rarely influenced by surgery. These patients were severely disabled and only rarely able to return to work. Garfield and Lytle (1970) noted that only one of their patients was able to return to work and all three were significantly disabled.

Conclusions

Back pain is a common complaint and cauda equina tumours are a rare condition. Back pain with or without sciatica is the usual presenting symptom and pain in recumbency a useful diagnostic feature. Less commonly, patients may present with painless weakness and wasting of the lower limbs, sphincter disturbance, papilloedema and raised intracranial pressure, subarachnoid haemorrhage, or sensory disturbance. There is usually a combination of symptoms at presentation and no characteristic pattern of physical signs. Valuable investigations are plain radiographs of the lumbar spine, lumbar puncture and manometrics, cerebrospinal fluid examination, and myelography.

The treatment of choice is surgical and, where complete removal of the tumour is possible, the prognosis is good. Radiotherapy is of value in those patients with ependymomas in which complete removal is not possible. It is perhaps less valuable in the palliation of patients with intra-
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