Current Practice

PRACTICAL NEUROLOGY

Spinal Cord Compression

BERNARD HARRIES,* M.B., F.R.C.S.

British Medical Journal, 1970, 1, 611-614

The spinal cord and cauda equina are ensheathed within the bony spinal canal by the dura mater (theca) and the arachnoid membranes. Coating the cord is the vascular pia mater, and the extrathecal fat contains a venous plexus and many small arteries. Some of these enter the cord along certain of the nerve roots and are derived from the vertebral, intercostal, and lumbar vessels. Through the thoracic intervertebral foramina the vascular fatty extrathecal tissue is in direct relationship to the mediastinum and its contained lymphnodes. The intrinsic

Figure 1.— Diagram to show principal pathways in spinal cord related to known function.

arteries of the cord—the anterior and posterior spinal arteries—lie in the pia mater, the former within the anterior sulcus and the latter more superficially on the dorso-lateral aspect in relation to the posterior columns. From these vessels small vessels enter the substance of the cord, many of them anastomosing only poorly or not at all with vessels in adjacent segments above and below.

Paraplegia

An understanding of the significance of the four different types of paraplegia (or quadriplegia) is important.

Upper Motor Neurone Flaccid Paraplegia

This is a condition of total loss of function below the level of the lesion in the spinal cord when the onset has been rapid

or abrupt. Thus it is seen after severe cord injury when it is known as "cord shock." It is also produced when, after pre-monitory symptoms and signs, the final complete interruption of function has been rapidly progressive or sudden. This occurs frequently in the case of metastatic neoplasms affecting the spine or spinal canal and in some cases of tuberculous caries of the spine, in both examples with or without collapse of the vertebral bodies. It is probable that the cause is ischaemia of the cord followed by thrombosis in the affected segment or segments. Once thrombosis has occurred, with resultant softening, little or no recovery of function can be expected whatever therapy is carried out.

On examination all motor, sensory, reflex, and autonomic function is lost below the level of the lesion and the affected limbs are toneless and flaccid.

There is painless retention of urine, followed by overflow incontinence. Abdominal distension due to ileus is not uncommon. There is an absence of sweating below the lesion, but profuse compensatory sweating may be seen above the lesion in the normally innervated skin.

The state of flaccidity and areflexia gradually wears off over a period of three to six weeks and is replaced by a state of paraplegia in flexion or occasionally when the functional interruption has been incomplete by paraplegia in extension.

Upper Motor Neurone Spastic Paraplegia in Flexion

This may follow the state of flaccid paraplegia described above or may be the eventual result of a slowly progressive compressive lesion, such as a benign spinal cord tumour. It is characterized by the absence of voluntary function below the level of the lesion accompanied by impairment or complete loss of sensation. Tone in the limbs is increased, the tendon reflexes exaggerated, and the plantar responses are frankly extensor. Painful paroxysmal flexion spasms are a prominent feature and may be induced by quite slight cutaneous stimuli and be accompanied by involuntary evacuation of the bladder and bowel, the so-called "mass reflex." Complete recovery of function in a patient with established paraplegia in flexion can seldom be expected except in the case of compression due to a benign spinal cord tumour (when the compression has been very slowly progressive). Removal of such a tumour even at this late stage may result in appreciable and useful recovery from the paralysis.

Painless retention of urine occurs when cord function is completely interrupted whether the lesion is sudden in onset or slowly progressive. Retention unrelieved by catheterization is followed by dribbling incontinence due to overflow.

At a later state in certain cases of severe paraplegia a primitive autonomous reflex arc mediated through the afferent and efferent nerves to the bladder wall and sphincters may

*Consultant Neurosurgeon, University College Hospital; Honorary Consultant Neurosurgeon to the Whittington Hospital.

take over control and result in periodical spontaneous filling and emptying. This is the so-called "automatic bladder." The cell stations for the reflex lie in the lumbar segments of the cord. When it occurs the automatic bladder is of considerable advantage to the patient. It arises in a minority of patients who have a severe chronic paraplegia and becomes manifest after the stage of flaccidity has passed. It occurs most frequently in paraplegia due to trauma and is dependent upon a sterile urine, good general health, and other factors which are incompletely understood.

In certain cases of cauda equina paralysis affecting the sacral nerve roots dribbling incontinence of urine may result without retention. Most cauda equina lesions, however, develop retention with overflow.

Upper Motor Neurone Spastic Paraplegia in Extension

This is a condition of weakness or total paralysis of the lower limbs accompanied by heightened tone (spasticity) which principally affects the extensor musculature. The condition is associated with incomplete lesions of the cord, and as these may be variable in degree and extent the clinical picture covers a spectrum from a very mild paraparesis on the one hand to a state which merges into paraplegia in flexion on the other. Paroxysmal spasms of the extensor muscles occur which, if the lesion progresses, may alternate with or be replaced by flexor spasms. The tendon reflexes are exaggerated and the plantar responses are extensor. Impairment of sensation is variable, ranging from complete loss of all modalities below the lesion to little or no disturbance (as in cervical spondylosis or anterior spinal artery thrombosis). Bladder control is similarly variable in degree, ranging from normal function, or relatively minor disturbances such as hesitancy or precipitancy, to complete loss of voluntary control with retention and overflow.

Paraplegia in extension is characteristic of the earlier stages of progressive spinal cord compression, such as may be found to be due to benign tumours or cervical spondylosis, but equally it may be the result of intrinsic cord disturbances such as multiple sclerosis or to certain relatively mild cord injuries.

Brown-Séquard's Syndrome.—The anatomical peculiarity of the cord whereby the ascending (spinothalamic) pathways for pain sensation are crossed relative to the descending pyramidal (motor) tract may result in a paraparesis in which motor symptoms and signs prevail in one lower limb while sensory impairment to pain is discovered on the opposite side rather below the level of the lesion. Such a situation occurs when mainly one half of the cord is affected by the lesion, and is a not uncommon mode of presentation of benign spinal cord tumours producing mainly lateral pressure. It is seldom that the syndrome is seen in "pure" form, and usually there is evidence of some bilateral motor, reflex, or sensory impairment. If the compression is not relieved the condition will merge gradually into one of the forms of spastic paraplegia already described. (Fig. 1.)

Lower Motor Neurone Flaccid Paraplegia

This is characteristic of acute and chronic lesions of the cauda equina. The lower limbs are flaccid, and wasting of muscle groups eventually occurs. The tendon reflexes are diminished or absent at the knee or ankle, and the plantar responses are flexor or unobtainable. Sensory impairment involves the sacral and lumbar dermatome distribution—so-called "saddle anaesthesia." Control of micturition is usually impaired, and urethral, vaginal, and rectal sensation may be absent. Painless retention with overflow or dribbling incontinence will result depending upon the anatomical pathways involved.

There are two zones of the spinal cord lesions of which have rather special characteristics. These are the "conus medullaris" and the cervical cord. Lesions of the conus result in a mixed upper and lower motor neurone paralysis—usually a flaccid paraplegia, absent tendon reflexes, extensor plantar responses, saddle anaesthesia, and retention of urine.

Lesions of the cervical cord at or above the 4th segment will lead to respiratory failure. Acute lesions such as trauma to the cord may cause sudden death while in chronic lesions, causing progressive paresis of the intercostal muscles and diaphragm, respiratory failure occurs as a terminal event. Chronic lesions of the upper cervical cord and of the cervical enlargement often result in wasting of the small muscles of the hands, forearms, and arms, depending on the particular anatomical disturbance, and are accompanied by symptoms (often paraesthesiae) and signs of sensory impairment in the affected dermatomes. The tendon reflexes below the lesion will be exaggerated, while those at the level of the lesion will be absent—often a useful sign for clinical localization.

Early Symptoms of Cord Compression

Since the early diagnosis and treatment of cord compression of gradual onset offers the best hope for useful recovery and the avoidance of the miseries of an irrecoverable state of paraplegia in flexion, the premonitory symptoms are all important.

The diagnosis of established paraplegia is generally an easy matter. It is less easy to recognize or suspect incipient cord compression from among the variety of isolated, albeit persistent, symptoms which so often herald its onset. There may be little time for conjecture. This is particularly the case when a localized infection or a malignant tumour is the cause.

Suspicion should be aroused when the patient complains of persistent localized pain in the back or when the pain is referred girdle fashion, on one or both sides, to the chest wall or abdomen. In such cases a thoracic or abdominal viscus is often blamed even in the absence of supportive evidence of disease of the heart, lungs, stomach or biliary tract. (Carcinoma of the body of the pancreas as a cause of thoracolumbar backache should always be kept in mind.) Pains referred to the groin, perineum, or the legs may similarly cause confusion. Such root pains are common in association with benign extramedullary tumours of the dorsal cord and may persist for weeks or months before definite neurological signs of paraparesis become manifest. Root pains are also common in the case of extrathecal malignant tumours, such as metastases from breast or lung, but in these the time scale is much shorter and may be limited to a few days or a few weeks before paretic symptoms and signs develop. Weakness, or sometimes "stiffness," of the legs and difficulty in maintaining balance are often complained of. It is usual for one limb to be mainly affected at first, and the patient to describe it as dragging or that he is "catching his toe." In other cases unheeded premonitory symptoms may be followed abruptly by an inability to stand on getting out of bed. Sensory symptoms are also common and include numbness or tingling in the legs or feet or described as a sensation of "walking on cotton wool." The symptoms may be unilateral (as in a Brown-Séquard paresis) or bilateral. Numbness or loss of urethral, vaginal, or rectal sensation are common with cauda equina compression.

Involuntary nocturnal jumps or spasms of the lower limbs are sometimes an early symptom but are more usual when the paraplegia is established. They require to be distinguished from the common myoclonic "jerks" which precede sleep in some individuals and which have no pathological significance.

When the cervical cord is affected weakness and progressive

wasting of the hands may be remarked on by the patient in addition to the more common complaint of numbness or woolliness of the fingertips. Root pains in these cases are often referred to the shoulders, the occiput, or behind the ear.

Disturbances of micturition are often early signs in any cord lesion. Hesitancy and precipitancy may progress to retention which is painless, and thence to an overflow incontinence. Such disturbances are often quite insidious and overshadowed in the patient's mind by the, by now, serious weakness of his limbs.

Outlines of Causation and Treatment

Spinal cord compression may be caused by injury, tumour, localized infection, and by certain less readily defined lesions of the intervertebral discs and vertebrae. For clinical purposes these require to be distinguished from those intrinsic disorders of the spinal cord which produce a paraplegia or quadriplegia in which compression plays no part.

Trauma

Injuries to the spinal cord giving rise to a paraplegia or tetraplegia may be compound (penetrating) or closed. The former are generally due to gunshot or stab wounds and are not necessarily associated with compression of the cord. Immediate surgical exploration and wound toilet followed by closure in layers is essential for these injuries.

Closed injuries of the cord are frequently but not invariably (as in "concussion" or mild contusion) accompanied by compression and are due, in the main, to dislocations, fractures, or fracture dislocations of the vertebral column. Acute traumatic compression of the cord results in contusion, softening, or pulping. Very seldom is the paralysis due to "simple" compression by a displaced bony or ligamentous fragment. Thus the injury to the cord may be best classified clinically rather than pathologically as (a) partially or wholly recoverable, or (b) irrecoverable. The diagnosis must in many cases be retrospective.

If a severe vertebral injury is accompanied by a complete paraplegia the latter is initially of the flaccid ("cord shock") type. In the majority of such cases recovery of function does not take place and a state of permanent spastic paraplegia in flexion results whatever treatment is instituted.

Lesser injuries may give rise from the beginning to a spastic paraplegia in extension from which state a greater or lesser degree of recovery may be expected.

In those relatively few cases where the cord has been merely bruised or nipped rather than seriously compressed (e.g. some cervical spine dislocations) good recovery may be expected with sometimes only relatively slight permanent disability.

Rarely the cord may become compressed by an extradural or subdural haematoma, which after the initial injury causes a progressive increase in paralytic symptoms. In such cases the vertebrae are commonly intact.

Generally the treatment of closed cord injuries is conservative, though in some centres early operative fixation of unstable fractures and fracture dislocations is advised in order to encourage rapid mobilization. Urgent stabilization and reduction of dislocations of the cervical spine by means of skull traction is invariably required whether the cord is affected or not. Subsequently internal fixation by open operation is often indicated.

Tumours

Spinal cord compression may be caused either by primary tumours—which are rare—or by metastatic tumours which are common.

Primary Tumours.—These generally present with a long and progressive history extending over months or occasionally years. Root (or girdle) pains are common and frequently early symptoms in the case of extramedullary and extradural tumours.

Primary tumours may be:—

- (1) *Intramedullary* (i.e. growing within the cord).
 - Astrocytoma (especially dorsal cord).
 - Ependymoma (especially cervical cord and filum terminale).
 - Cysts (Cystic astrocytoma, enterogenous cysts, and syringomyelia).
 - Angiomatous malformations (these may give rise to subarachnoid haemorrhage).
 - (These are all rare, the first two being least uncommon.)
- (2) *Extramedullary* (i.e. intradural tumours).
 - Meningioma (especially dorsal cord).
 - Neurofibroma (especially cervical cord).
 - (These are the commonest primary tumours.)
- (3) *Extradural* (i.e. outside the dura within the vertebral canal).
 - Meningioma.
 - Lipoma.
 - Angioma.
 - Dermoid cysts.
 - Chordoma (slowly growing from notochord remnants).
 - Chondrosarcomas. (Malignant, but initially tend to be only slowly invasive and metastasizing late.)
 - Solitary myeloma (plasmocytoma). (May subsequently disseminate "myelomatosis.")
 - (These are all rare.)

Most intramedullary tumours can be "decompressed" by means of a laminectomy and opening the dura mater but less often can they be satisfactorily removed. Occasionally, using special techniques for haemostasis, partial or complete removal may be achieved when a plane of cleavage is discoverable between the tumour and the cord, but generally the management must be limited to decompression and radiotherapy. The prognosis varies with the growth rate of the tumour but many patients progress only slowly over a period of years. Occasionally the tumours are cystic and transient improvement takes place following aspiration at laminectomy.

Most benign extramedullary tumours are completely removable by means of an appropriate laminectomy. A few neurofibromas of the cervical or dorsal region protrude through an intervertebral foramen to produce the so-called "dumb-bell" tumour, for which a second extraspinal operation may be required for complete extirpation. Some dumb-bell tumours take origin in the neck or in the paravertebral region of the dorsal spine and protrude secondarily into an intervertebral foramen to reach the spinal canal and may eventually cause spinal cord compression. Such tumours are usually neurofibromas but malignant neuroblastomas of the sympathetic chain can behave similarly.

The prognosis for patients with benign extramedullary tumours is good if removal has been complete. The paraplegia is likely to recover either completely or to an extent which leaves only manageable neurological sequelae.

Benign extradural tumours are usually removable either in part or in toto. The ultimate prognosis is dependent very much upon the type of tumour, its extent, and whether it involves other structures.

Metastatic tumours.—The history is generally short (two to three weeks) or very short (a few days), though backache and girdle pains may have been complained of without evident signs for longer periods of weeks or sometimes months.

The primary condition may be a carcinoma (lung, breast, prostate, kidney and bowel are the commonest), reticulosis (lymphoma or lymphosarcoma) or myelomatosis. Two groups of metastatic tumours may be distinguished. These are: (a) when there is major bony involvement, i.e. collapse of vertebral bodies or malignant erosion of pedicles, laminae or

spinous processes radiologically; (b) when radiologically there is no visible bony involvement.

Diagnosis and localization are easier in the former group, and generally the prognosis is worse than in the second group even with prompt treatment. Because the reticulosos seldom involve bone so extensively as the carcinomas and are generally highly radiosensitive the prognosis in these cases is better. Metastatic growths reach the spinal canal either by direct spread from an invaded vertebra, by extension from the mediastinum through an intervertebral foramen, or (uncommonly) by direct haematogenous spread to the extradural fat. Such metastases invade the dura with difficulty and seldom if ever penetrate it to gain access to the leptomeninges and the cord itself. Very rarely haematogenous metastases are deposited intrathecally in the arachnoid or the cord itself.

Until recent years paraplegia due to metastatic malignant growths tended to be managed rather inactively, though the suspected area of the spine was often irradiated in an attempt to relieve local pain. Irradiation alone may sometimes achieve worthwhile improvement but accurate localization by myelography followed by subtotal removal of the extrathecal tumour is now to be preferred. Furthermore laminectomy no longer carries any serious risks to the patient nor demands

any unusual expertise from the surgeon. In addition to providing immediate relief of cord compression a positive histological diagnosis is obtained in those cases where the nature of the metastasis is unknown. Appropriate radiotherapy and, where required, chemotherapy or endocrine therapy may be begun a few days before the sutures are removed. A more resourceful approach of this kind has resulted in an improvement of the lot of these patients—not least from the fact that they receive active nursing rather than “terminal care.”

Enthusiasm for such activity must naturally be tempered by an awareness that in many patients, perhaps a majority, little or no functional improvement will occur in the paralysed limbs or bladder. In my experience the best results have been obtained with the reticulosos, carcinoma of the prostate and breast, and some of the rarer radiosensitive tumours such as seminomas. Almost invariably poor results occur when the metastasis is from the bronchus. When a complete paraplegia is associated with vertebral collapse recovery of function is least likely to occur. Vertebral collapse associated with early or only moderately severe paraparetic symptoms and signs—before irreversible ischaemic changes have taken place in the cord—carries a rather better outlook.

(This article will be concluded next week.)

TODAY'S DRUGS

With the help of expert contributors we print in this section notes on drugs in current use.

Treatment of Menière's Disease

Menière's disease is characterized by the occurrence of attacks of vertigo in the presence of fluctuating tinnitus and of deafness which tends to progress. It affects about 1 in 1,000 of the population, with a slight predominance of males. It may begin at any age, but the maximum incidence of onset is between 40 and 60 years. While initially the attacks of vertigo may increase in frequency and severity and the deafness may progress, the disorder is in fact self-limiting, and spontaneous arrest may occur at any stage. The few cases which have been available for post-mortem examination have shown dilatation of the cochleosaccular endolymph system at the expense of the perilymph system in the fixed volume of the bony labyrinth. This is thought to be associated with biochemical changes in the perilymph and endolymph affecting particularly sodium and potassium ions and water.

Clinical Features

The first symptom in over half the cases is sudden onset of vertigo. In others this may be preceded by months or years of deafness or tinnitus. The vertigo may vary in severity and character; usually it consists of a feeling of rotation of objects or of the subject in relation to the surroundings. It may take the form of a sensation of being forced from one side to the other. Occasionally the patient may be suddenly thrown to the ground, either backwards or forwards, as though hit with a hammer. Rarely patients may experience merely a persistent feeling of unsteadiness.

The attacks may occur at intervals of months or even years. There may be premonitory symptoms such as the appearance of, or an increase in the intensity of, tinnitus, fullness in the affected ear or in the head, nausea, vomiting, prostration, faintness or even loss of consciousness, transient diplopia, or cere-

bellar dysfunction with the appearance of nystagmus. The duration of the vertigo varies from minutes to hours, and it may be followed by a period of unsteadiness. There may be a headache, usually in the occipital region. The tinnitus may be paroxysmal or constant, unilateral or bilateral, fluctuating in intensity, being worse at the times of the attacks. The deafness may also be unilateral or bilateral, usually with one side more severely affected; while progressive, it rarely becomes high-grade unless bilateral at onset. In the early stages there may be distortion of hearing.

The psychological aspects of Menière's disease are of considerable importance. As in many other conditions with an undetermined aetiology, various personality traits are alleged to predispose to it, and stress situations have been thought to trigger off attacks. While such theories remain unproved there is no doubt that attacks of vertigo, whatever their cause, lead to anxiety and even depression, the severity of which depends probably on the underlying personality of the patient. Similarly, persistent tinnitus may trigger off psychiatric symptoms in predisposed patients. This aspect of the illness is often more disabling than the actual attacks, which may be infrequent and short lived, and treatment must be directed as much to this as to the attempted reduction of the frequency and severity of the attacks.

While acute attacks are treated with vestibular and central sedatives and drugs to reduce nausea, the treatment for the prevention of attacks is more varied and its effects much more difficult to assess. There is great natural variation in the rate of progression of the illness and in the severity and frequency of the attacks of vertigo. The associated anxiety and tension are often the most disabling aspect of the illness in between the attacks, and there is no doubt that sympathy, encouragement, and firmness on the part of the physician are of great importance in the management of patients—but this adds further to the difficulty of objective assessment of the effects of various treatments. Surgical treatment is indicated in only perhaps 10% of patients, who fail to respond to medical measures and who are significantly disabled by their attacks.