The management of primary spinal cord tumours

**REFERENCES**


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Primary spinal cord tumours are rare but patients may present with multiple problems, although pain is generally the primary symptom. Following diagnosis, surgery tends to be the first treatment option. However, even after microscopic surgical excision tumour remnants may still be present, which can necessitate further treatment with radiotherapy and chemotherapy. Patients who have had surgery may have to adjust to significant changes in body image but can be supported by expert nurses who help them to adjust and encourage them in their recovery.

Primary spinal cord tumours are rare, but for the individuals concerned, and for those close to them, they are devastating. Diagnosis is frightening, and the fear of paralysis, sexual, bowel and urinary problems may give rise to physical and psychological problems, which can lead to social isolation. For patients’ partners and carers, new roles may need to be learnt. These changes in roles may not just alter future life plans but may also have more immediate catastrophic effects on finances, lifestyle, and the intimacy of their relationships.

Nurses play a major role in the support and care of this group of patients and their families. They can provide ongoing education, support and information that may help patients and their carers to adjust to the changes in their lives.

**Anatomy of the spine**

The spinal cord is a cylindrical structure that descends from the foramen magnum in the vertebral canal to between the first and second lumbar vertebrae. The spinal cord serves as the main pathway for ascending and descending fibre tracts that connect the peripheral and spinal nerves with the brain. There are 33 spinal vertebrae and 31 pairs of spinal nerves.

Like the brain, the spinal cord is enclosed and protected by the meninges and surrounded by cerebrospinal fluid. The meninges consist of the dura mater (external layer), arachnoid mater (middle layer) and pia mater (internal layer). The cerebrospinal fluid circulates in the subarachnoid space, which is between the arachnoid mater and the pia mater. The cord is well protected by the wall of the vertebral canal, the meninges, cerebrospinal fluid and the vertebral ligaments.

**Primary spinal cord tumours**

Primary spinal cord tumours are rare, with an annual incidence ranging from 0.5 to 1.4 per 100,000 of the population (Marmor and Gokaslan, 2002). They account for approximately 10-20 per cent of all tumours within the central nervous system. To put this in perspective, a new busy neurosurgical unit would see five to eight new patients per year on average, of whom a radiation oncologist would treat no more than three.

**Classification**

Tumours of the spinal cord are classified as being extradural or intradural. Extradural tumours are mostly metastatic tumours that can originate from breast, lung, kidney and prostate. Other extradural tumours are from lymphomas, myelomas and neurofibromas.

Intradural tumours are classified as intradural extramedullary (arising inside the dura but outside the spinal cord) and intradural intramedullary (arising within the spinal cord). Intradural intramedullary tumours are primary cord tumours, because they arise within the spinal cord.

Intradural extramedullary tumours include schwannomas, meningiomas and a range of other rare benign tumours. Intradural intramedullary tumours generally comprise tumours of glial origin such as astrocytomas and ependymomas (either low or high grade).

Glial cells are the supporting tissue of the brain and spinal cord and make up approximately 40 per cent of the central nervous system. There are three main types of glial cells: astrocytes, oligodendrocytes and ependyma.

Primary spinal cord tumours generally originate from %

**BOX 1. CASE STUDY: PETER’S STORY**

It all started in February 2000 when I was working as a sales manager, which involved a lot of driving. I noticed that my legs were aching, with occasional fleeting pains from one leg to the other. I put it all down to the driving and hoped that things would eventually improve. However, they didn’t and I made an appointment to see my GP, who prescribed anti-inflammatory tablets and referred me to the local physiotherapist. Physiotherapy did not help; in fact the pain was gradually getting worse. I was referred to a neurosurgeon with a diagnosis of sciatica. The neurosurgeon arranged for an urgent magnetic resonance imaging (MRI) scan, which revealed a primary spinal cord tumour.
astrocytes (giving rise to astrocytomas) or ependymal cells (giving rise to ependymomas). Unlike neurones that lose their mitotic capacity (ability to divide) at the time of birth, glial cells are able to divide throughout an individual's life and therefore can become malignant and lead to tumours.

Primary spinal cord ependymomas occur more frequently in adults, while primary spinal cord astrocytomas account for 60 per cent of paediatric tumours (Marmor and Gokaslan, 2002; Britton and Evanson, 2000).

Clinical presentation

Primary spinal tumours usually evolve over a period of months to several years. Pain is generally the primary symptom and is due to the growing tumour expanding the spinal cord and pressing spinal roots (Box 1).

The pain experienced is continuous, deep rooted and can be just as severe on movement as at rest. The pain occurs at the level of the tumour, with neurological dysfunction below that level. As the tumour grows, the patient may experience motor weakness, gait disturbances and sensory changes. Some patients may also present with bowel and bladder problems as well as sexual dysfunction.

As primary cord tumours arise within the substance of the cord, they may diffuse over several segments. They may also be associated with the development of a syrinx – a fluid-filled cystic cavity (Sharpe, 1998). Laterally placed lesions may lead to what is known as a Brown-Sequard-type syndrome (ipsilateral – that is same side – paralysis and loss of proprioception/touch – a fluid-filled cystic cavity (Sharpe, 1998). Laterally placed lesions may lead to what is known as a Brown-Sequard-type syndrome (ipsilateral – that is same side – paralysis and loss of proprioception/touch with contralateral – that is opposite side – loss of pain and temperature control).

BOX 2. PETER’S STORY CONTINUED

Following the MRI scan, my life changed completely. Within a week I was admitted to the neurosurgical ward for spinal surgery. I was told that my life expectancy was anything from five to 20 years, with a high risk that I would become paralysed. I was left in a side room with a very sick man who died later that day. I was frightened and, as a consequence, I discharged myself, against medical advice, a few hours before the scheduled surgery.

For the next three years I had MRI scans every six to eight months to monitor the progress of the tumour. My legs continually ached and I had to dose myself regularly with painkillers to be able to work. As a consequence I was often tired and could not undertake as much work as previously.

By January 2003 I was badly limping on my left leg and I had a severe burning sensation in my hip joints. I wanted to believe that this was as a result of a slipped disc and not the tumour as I could still feel my legs and had no problems with my waterworks or bowels. I was readmitted to a neurosurgical unit. This time it was far scarier. I was told that my condition was now very serious and I was at a high risk of paralysis and Prompt diagnosis followed by appropriate treatment is necessary to preserve neurological function and, whenever possible, maintain the patient’s independence.

Diagnostic imaging

In the diagnosis of primary spinal cord tumours, magnetic resonance imaging (MRI) has replaced myelography and computerised tomography (CT) as the investigation of choice. This is because contrast-enhanced MRI with gadolinium (Gd-DTPA) is significantly more sensitive in detecting multiple lesions as it shows up soft tissue in greater detail (Box 2).

An MRI scan of the entire brain and spinal cord (craniospinal axis) should be undertaken to rule out tumour spread. Low-grade ependymomas and astrocytomas generally remain localised (Lindstadt, 1998). However, high-grade ependymomas and astrocytomas can spread via the subarachnoid cerebrospinal fluid throughout the craniospinal axis.

Being able to differentiate reliably between astrocytomas and ependymomas is not always possible with MRI, but to the experienced neuro-radiologist the presence of haemorrhage may suggest ependymoma. Ependymomas are generally better defined than astrocytomas with MRI as they tend to expand the cord and commonly enhance with contrast (Britton and Evanson, 2000).

Tumours can erode into blood vessels, giving rise to leakage of contrast agents, and this is seen as enhancement on scans. The main value of neuro-radiological investigations is that they allow assessment of the actual site of the tumour and assist in determining the neuro-surgical approach; that is, whether it is to improve symptoms or to provide histological diagnosis (Britton, 1998).

During oncological management, MRI scans are vital for monitoring the response of the tumour to radiotherapy and/or chemotherapy as well as for detecting tumour recurrence.

Treatment options

Surgery

Surgery is generally the first treatment option for patients with primary spinal cord tumours because it allows the cord to be decompressed, thus relieving the symptoms of spinal cord compression (Addison and Shah, 1988).

The surgeon will locate the tumour and remove as much as possible, at the same time obtaining tissue for histological diagnosis. As primary cord tumours are tumours of the cord itself, complete surgical excision may be very difficult (Box 3).

Before surgery, the surgeon will discuss with the patient the potential risks of the procedure, such as infection, worsening of existing deficits and, in some instances, the possibility of death.

Other complications may include transient deterioration of the patient’s neurological condition, which can last from a few days to months, loss of proprioception (joint position sense), the development or progression of

REFERENCES


I underwent surgery and made a good recovery. I was reassured by the fact that the surgeon was pleased with the operation and by how much of the tumour he had been able to remove. I saw a physiotherapist and an occupational therapist, and as I was able to mobilise and was generally fairly independent, I was able to return home within two weeks. The catheter that had been inserted prior to surgery was removed before my discharge; my bowels were also functioning well.

Within three days of being discharged, I was having problems passing urine, my stomach was swollen, and I was having trouble controlling my bowels. There was also discharge from the operation site. I was readmitted to hospital.

Following surgery, I was told that my urinary and bowel function should improve, as this was likely to be a result of temporary surgical cord bruising. The discharge from my back ceased.

I remained in hospital for a further four weeks. During this time there were a couple of attempts to remove the urinary catheter. However, I was unable to pass urine independently and I still had no bowel control. I also noticed that I had lost sensation in my right leg above the knee, which was new and really frightening. Getting comfortable was difficult as I had severe burning and aching in my legs when standing or sitting. My legs also felt heavy, and trying to walk was like wading through a bath of cement.

I was told that the bowel and urinary problems might be permanent but not to give up hope, as improvement often takes months. I was distraught and shattered.

Bob, my nurse, was there to listen to me and offer practical help and advice; he was reassuring but realistic. He taught me how to self-catheterise. I found the thought of self-catheterisation extremely upsetting, but it allowed me to be in better control.

Unfortunately the nightmare got even worse.

The doctor arranged for me to have a further scan, after which I was told that there was still quite a lot of swelling but that, worse still, more tumour could be detected. I would now need to be referred to an oncologist for radiation therapy.

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Radiotherapy

Primary spinal cord tumours can be large and extensive, and after microscopic surgical excision, tumour remnants may still be present. If there is a concern about residual microscopic disease, most surgeons will refer patients to an oncologist for postoperative external beam radiotherapy (teletherapy) (teletherapy refers to the projection through space of X-rays or gamma rays by machines).

In general, postoperative radiotherapy is not indicated in patients with low-grade ependymomas if the tumour has been totally removed. A ‘watch and wait’ approach is often used by the surgeon and/or oncologist with regular spinal MRI to identify further evidence of disease, leading either to further surgery or radiotherapy should the tumour recur (Box 4).


There appears to be no debate about the postsurgical management of patients with high-grade gliomas of the spinal cord. In general, these tumours are treated with radiotherapy, although there is no clinical evidence of its efficacy (Marmor and Goskalan, 2002).

In this group of patients, neurological deficit as a result of tumour or surgery is rarely relieved by radiotherapy, and patients with high-grade gliomas have a very poor prognosis (Brada et al, 2001).

Side-effects

The side-effects of radiotherapy depend on the total dose given and the extent and levels of cord irradiated. The aim of radiotherapy is to destroy tumour cells, but because radiation cannot discriminate between normal and malignant cells it potentially causes damage to all living cells. Doses of radiation are typically prescribed in Gray (Gy).

A Gy is the absorption of one-joule (a unit of energy) per kilogram by material exposed to ionising radiation (Glance et al, 1986). Each body organ has a radiation tolerance, which must be adhered to so as to prevent injury to the surrounding normal tissue. The spinal cord level of tolerance is 2 cent per month of risk of damage at 50Gy.

The prognosis for patients with low-grade gliomas can run into years and therefore minimising side-effects is a major oncological issue.

kyphoscoliosis (curvature of the spine) and cerebrospinal fluid leaks (Marmor and Goskalan, 2002).

Minimising deficits and other treatment complications is paramount in order to maintain the patient’s well-being and promote ongoing rehabilitation.

The educational and supportive role of neurosurgical nurses should not be underestimated. Patients are often frightened and may have major concerns regarding loss of mobility, and sexual, urinary and bowel function.

They may worry about loss of independence and control and fear having to rely on others for intimate care such as bowel and urinary management (Guererro, 2003). Neurosurgical nurses play a major role in reducing anxiety for patients and their families as well as in providing pre and postsurgical education and care.

The expert neurosurgical nurse can often anticipate a patient’s needs and adapt care pathways and education programmes in ways that may promote better recovery and ongoing rehabilitation. For example, giving adequate and timely information may allow the patient to make more informed decisions and feel more in control of the situation.

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Erythema of the irradiated skin causes discomfort and presents as redness and inflammation. The treatment for radiation-induced erythema tends to be controversial, though hydrocortisone cream can alleviate discomfort.

When the cervical cord is irradiated, transient myelopathy (Lhermitte’s phenomenon) is a common syndrome. It occurs approximately 12 to 20 weeks after spinal irradiation in 25–40 per cent of patients who receive over 35–40Gy to significant lengths of the cord (Kun, 1994). Lhermitte’s phenomenon is characterised by transient ‘electric shocks’ that spread down the body when the head is flexed forward. It resolves after a few months to a year following treatment.

Other side-effects can include tissue necrosis and, occasionally, rapidly evolving permanent paralysis is seen, possibly resulting from an acute infarction of the spinal cord (Rubin et al, 1975).

In general, radiotherapy for primary cord tumours is confined to the region of the tumour and the margin of potential spread. More extensive irradiation, including brain and whole spinal cord, is not advocated (Brada et al, 2001) because the risk of isolated brain seeding is approximately six per cent and is marginally more likely to occur in patients with high-grade tumours (Whittaker et al, 1991). However, should brain and spinal cord irradiation be considered, fertility issues must be discussed, with females being given the option of undergoing oophoropexy (repositioning of the ovaries) or another means of egg collection, and male patients offered sperm banking where appropriate.

Because of the large volume of bone marrow within the radiation field, patients undergoing brain and spinal cord irradiation require weekly full blood counts to detect bone marrow suppression. If this is apparent the patient should be rested from treatment for a few days and full blood counts repeated to ensure recovery before recommencing radiotherapy.

Radiotherapy treatment tends to be daily (weekdays) for six to six-and-a-half weeks. During this time, the patient should be seen at least weekly, so progress can be monitored, any questions answered and reassurance offered. Research indicates that the provision of information, education and treatment monitoring can be carried out effectively by trained nurse specialists both in an outpatient setting and over the telephone (Curran, 2001; Guerrero, 1994; James, 1994). Nurses therefore, must have expert knowledge and their judgement must be backed by research-based practice.

Chemotherapy
The adjuvant role of chemotherapy in the treatment of primary spinal cord tumours is even less clear than that of radiotherapy, with no established guidelines for its use or indication of its true benefit.

In principle, chemotherapy may be used when there is evidence of recurrent local disease, particularly as radiotherapy cannot be repeated to the previously irradiated area because of the danger of myelitis and necrosis to the spinal cord. However, at this stage further surgical intervention should be given full consideration, although this is not always a realistic option.

Conclusion
Patients with spinal cord tumours have to adjust to significant changes in their body image. They can also experience social isolation as they adjust to coping with a new self, which can often lead to what has been described as ‘life surrounded by losses’. These losses can have a serious impact on a patient’s quality of life.

Nurses can work alongside these patients, teaching them, supporting them, encouraging them and relating to them on a personal level. Expert nurses are able to provide realistic goals that take into account a patient’s potential level of recovery, both physical and emotional, and to direct care, so inhibiting dependence and encouraging recovery.

### BOX 4. PETER’S STORY CONTINUED

The cancer specialist reassured me that although radiotherapy was necessary, there was no immediate rush for this and that I would need some special preparation before starting the treatment. I was seen by the nurse specialist and also referred to the physiotherapist and occupational therapist. Visits to the simulator were arranged to plan the radiotherapy, and the occupational therapist visited me at home, while the physiotherapist arranged to see me on a regular basis.

The radiotherapy took six-and-a-half weeks to complete. It was a tie getting to hospital, as I was unable to drive and therefore relied on family and friends. Hospital transport was suggested, but I felt that I wanted to be more in control of my day, as I was already feeling very vulnerable. Because I needed to self-catheterise and occasionally my bowels were unreliable, I did not wish to be taken to hospital by strangers, as I felt embarrassed.

The only problem that I had during treatment was a mild redness around the treatment area, which caused me slight discomfort. The most difficult and frustrating thing during this period was not being in control of my life, and having to rely on family, friends and hospital staff when I finished the radiotherapy.

Things are now slightly better; thankfully, no worse. I am still able to get around without major difficulties. I still take painkillers, although not as frequently as before. I am adjusting to the fact that my bowel and urinary problems are now chronic and I have to live with this. I self-catheterise around five times a day, and have my own routine for managing my bowels. Thankfully, I have a good nurse continence adviser whom I see as required.

I have managed to cut down on my hospital visits and have regular telephone contact with the hospital nurse specialist. I am also attending a support group, which is of benefit to me as I meet others who understand my situation.

### REFERENCES