Spinal tumours of childhood

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Summary
Spinal tumours in children have different features and require different management than in adults; 20 patients under the age of 14 years are reported — the common presenting features were limb weakness, pain and spinal deformity. Eight tumours were extradural and 12 intradural, of which 3 were intramedullary. The 6 patients who did not do well postoperatively were found to have a malignant tumour or were completely paraplegic or quadriplegic before surgery. If detected and treated early, results are excellent in many cases, since the tumours are usually benign.

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Spinal tumours are uncommon in childhood but present in a subtle way so that they are easily overlooked as a cause of pain, limb weakness and spinal deformity. If suspected, the diagnosis is easily made and most tumours can be completely removed with good results but permanent paralysis and incontinence is the price of misdiagnosis or delay in treatment.

Although the commonest clinical manifestations are backache, neurological deficit or scoliosis, a loin mass or soft tissue swelling over the spine may be an early sign before there is neurological involvement. We present a series of children with spinal tumours to illustrate the clinical presentation, radiological features and variety of lesions.

Patients and methods
Twenty patients under 14 years of age were seen because of spinal tumours over a 7-year period (1979-1985) in the Neurosurgical Department of the University of Cape Town. The age range was 3 months - 13 years, with 6 patients under 2 years and 10 over 10 years. There were 10 boys and 10 girls, 7 being black, 6 coloured, and 7 white. Pain was the presenting feature in 9 of the children. Abdominal pain led to a search for an intra-abdominal lesion in 3 while lumbar pain was present in 3. Pain in an arm was associated with a high cervical neurofibroma, and pain radiated down both legs in a child with a neurofibroma at the L4 level. Neurological deficit was the commonest clinical feature, being present in 15 children. Weakness in one or both legs was seen in 15 cases and was usually asymmetrical with an associated sensory loss in 6 cases. It was impossible to assess sensory deficit accurately in the very young children. Spinal deformity was seen in 3 cases; 1 patient had a scoliosis, 1 had loss of lumbar lordosis and 1 had marked paraspinal muscle spasm. A mass was palpated in 4 of the children; it was in the loin in 3 of the 4 with abdominal neuroblastomas, and a subcutaneous swelling was found over the dorsum of the sacrum in the patient with a sacrococcygeal chordoma.

Spinal radiographs revealed erosion of pedicles in 3 cases, scalloping of the posterior borders of vertebral bodies at 2 levels, scoliosis in 1 case and a metastatic deposit in a single vertebral body in 2 cases. Myelography was not performed pre-operatively in 3 cases, the child with the sacrococcygeal chordoma and 2 patients who had undergone a laparotomy or thoracotomy for a neuroblastoma which was then found to have an intraspinal extension. A complete block to the contrast medium was seen in 14 myelograms. One patient with a paraspinous neuroblastoma had flattening of the adjacent pedicles at L1 and L2 and myelography showed a focal indentation of the dura. Four tumours were cervical, 8 thoracic, 7 thoracolumbar and 1 sacral.

Of the 20 tumours, 8 were extradural and 12 intradural and in the latter group 9 were extramedullary and 3 intramedullary. The extradural tumours were 5 neuroblastomas, 1 hepatocellular carcinoma that invaded the epidural space by direct spread, 1 neurofibromas, 3 meningiomas, 1 malignant schwannoma (associated with Von Recklinghausen's disease), 1 medulloblastoma seeding to the lumbar sac from the fourth ventricle, and 1 drop metastasis from an intraventricular choroid plexus carcinoma.

At operation an attempt was made to remove the intramedullary neurofibromas completely, with marked postoperative improvement.


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The patient with the intramedullary ependymal cyst was totally paraplegic on admission and remained so after surgery. Six of 8 patients with intradural extramedullary lesions did well. Apart from 1 patient with a meningioma who was quadriplegic before surgery and did not show a functional improvement, the other 2 children with meningiomas did well. The 3 neurofibromas and 1 malignant schwannoma were totally excised with the patients' sustained improvement. The patient with the drop metastasis from the lateral ventricle choroid plexus carcinoma which compressed the cord at T10 was completely paraplegic before surgery and remained so postoperatively.

The little girl who had a sacral chordoma removed 8 years ago remains well. The patient whose cord compression was due to a hepatocellular carcinoma has died, while the boy who had a neurenteric cyst improved after its excision.

The child with medulloblastoma seedings in the cauda equina presented with backache and leg weakness and was subjected to myelography and biopsy; the primary in the fourth ventricle was finally discovered on computed tomography (CT). Radiotherapy relieved the pain and leg weakness but she eventually died 2 years later. Four of the 5 patients with intraspinal involvement due to neuroblastoma have done well neurologically, while the 1 who was paraplegic made a dramatic recovery after surgery and is now walking with support 2 years later.

Discussion

Paediatric brain tumours represent 40–50% of all solid tumours in the population under 15 years of age and the ratio of spinal tumours to brain tumours is said to be between 1:10 and 1:5. During the 7 years that the 20 spinal tumours in this series were treated 145 brain tumours were dealt with in the same department, giving a ratio of about 1:7 spinal to brain tumours. Although others find a slight male preponderance, the incidence was equal for boys and girls and for the three racial groups.

Weakness of one or both lower limbs is the commonest initial symptom; neurological deficit, pain and spinal deformity. Weakness results in an imbalance of muscle action and kyphoscoliosis, while muscle spasm in the neck induces a torticollis or in the lumbar region causes loss of lordosis or a painful stiff back.

Fifteen of the 20 patients in this series had weakness of one or both legs. Cauda equina tumours are associated with lower motor neuron-type weakness in one of the legs with a dermatomal sensory loss that can only reliably be elicited in older and co-operative children. Thoracic level spinal tumours produce bilateral upper motor neuron leg weakness with an appropriate sensory level, whereas cervical lesions often give rise to lower motor neuron signs in one or both arms plus upper motor neuron signs in the legs. Apart from the abnormal curvatures of the spine, paraspinal muscle spasm and tenderness may be found in addition to local sensitivity to firm palpation over the spine. A distended bladder and poor anal tone must be looked for. A sacral sensory level is easily discovered on palpation of the sacrum or on rectal examination in patients with a chordoma whereas a loin mass is often the presenting feature in children with neuroblastoma. Evidence of Von Recklinghausen's disease such as café au lait spots or axillary freckling should be sought because of the association with intraspinal neurofibromas.

Differential diagnosis includes a number of conditions easily identified by careful examination or appropriate special investigations. Congenital disorders such as spina bifida occulta with tethering of the spinal cord as common denominator will be detected by examining the skin in the lumbar region where a lipoma, sinus, dimple, hairy patch or angiomatous malformation may be seen. Spinal epidual abscess is associated with acute local tenderness and pyrexia, and myopathies cause no sensory loss and their presence is confirmed by muscle enzyme studies. Poliomyelitis is fortunately now uncommon but presents as an asymmetrical weakness leading to muscle wasting without sensory loss. Lumbar disc disease has a characteristic clinical pattern but is extremely uncommon before the age of 15 years.

The first investigation required is a full spinal radiographic examination. The dysraphic states will be shown by the presence of one or more bony spina bifida processes. Intramedullary tumours may produce widening of the interpedicular distance over a number of segments and pedicles are often flattened from side to side by compression. Intervertebral foramina are widened by dumbbell neurofibromas extending from the spinal canal into the paraspinous tissues. Scallop ing of the posterior border of one or more vertebral bodies is sometimes seen, being a characteristic feature of cauda equina ependymomas.

Erosion of pedicles or vertebral bodies is a more sinister sign indicative of malignant tumour involvement. CT may be helpful in confirming subtle changes thought to be present on plain radiographs.

Myelography is the investigation of choice and is nowadays most frequently performed with water-soluble contrast medium introduced either at the lumbar or the cervical level. It indicates not only the level and size of the tumour when the subarachnoid space is not totally occluded, but also the lower border of the lesion and thereby for urgent surgery when a block is complete.

Extramedullary intradural tumours have a caudal intradural convexity while intramedullary tumours produce a fusiform enlargement of the spinal cord shadow. Extramedullary lesions are usually distinguished by a 'paintbrush' ending, tapering gradually on each side. The cerebrospinal fluid (CSF) obtained at the time of myelography should be subjected to chemical, bacteriological and cytological examination. High protein concentrations are found in the presence of tumours, especially those producing a complete block, and malignant cells may be seen if the tumour is an ependymoma or if there are 'drop metastases' from a fourth ventricular medulloblastoma or from a rhabdomyosarcoma of the head and neck. New-generation CT scanners are now being used to diagnose spinal tumours but usually require the assistance of a water-soluble contrast medium. Most centres still find myelography the most helpful investigation and isotope scanning is rarely used. Electromyography may be of value in differentiating peripheral nerve lesions from spinal tumours and CT of the head is worth considering if a lumbosacral intradural tumour is found in conjunction with malignant cells in the CSF.

Intramedullary spinal cord tumours usually account for 25-33% of all primary intraspinal tumours of childhood, but in this series formed only 15% of the total. Unlike the adult population, in which ependymomas are significantly more frequent than astrocytomas, approximately two-thirds of paediatric
intramedullary tumours are astrocytomas. These paediatric tumours tend to be more rostrally situated than those in adults, the majority being in the cervical or cervicothoracic cord. Intramedullary tumours generally occupy many cord segments and have been reported to extend the entire length of the cord although in these cases only a short segment is probably tumour with cystic extension below, rather like the cystic cerebellar astrocytoma of childhood with its small tumorous nodule and large non-neoplastic cystic cavity. Until recently it was thought that total removal of intramedullary tumours was impossible but it has been shown that in most cases there is a good plane of cleavage between the cord and tumour which facilitates complete removal. One-third of the childhood intraspinal ependymomas reported by Hendrick1 arose from the filum terminale.

Unlike its incidence in adults, in whom it is probably the most common intraspinal tumour, neurofibroma is reported to be quite rare in childhood. In Ingraham and Matson52 series it constituted about 5% of intraspinal tumours. There were 3 in this series of 20. Progressive scoliosis in a child suggests this type of tumour, and widening of an intervertebral foramen is the characteristic radiographic change often associated with a localized increase in interpedicular distance. Total extirpation is the goal of surgery and a good prognosis can be expected even in patients with a severe pre-operative neurological deficit.

Meningeal spinal tumours presenting in childhood are rare in comparison with those in adults. Ingraham and Matson2 reported 3 such tumours in their series of 134 intraspinal tumours. We encountered 3 in only 20 cases. They should be completely excised with a border of normal dura, the defect being patched with fascia, lyophilized dura or Silastic although it is surprising how well such a defect heals free of CSF leakage without repair.

In one large series nearly half the spinal tumours affecting children were found in the epidural space, one-third belonging to the neuroblastoma-ganglioneuroma group which arise from the adrenal gland or sympathetic chain. Five of our 8 extradural tumours were neuroblastomas. Spontaneous regression sometimes occurs from the malignant neuroblastoma to the benign ganglioneuroma,3 and intraspinal extension of this tumour occurs with significant frequency. Pettersen and Harwood-Nash8 reported that with CT metrizamide myelography on 18 patients with neuroblastoma they found intraspinal extension in 11, unexpectedly from the clinical findings in 6. It is important for the surgeon to know whether there is intraspinal extension pre-operatively to enable him to plan the surgical approach. We have found metrizamide myelography a wise precaution before surgery; in most cases all tumour can be removed via the anterior abdominal approach through a widened intervertebral foramen. Chordoma is an uncommon extradural tumour usually found in the sacrococcygeal area in children and rarely in the upper cervical region or base of the skull.10 The sacrococcygeal chordoma presents with localized back pain, a swelling over the dorsum of the sacrum, and occasionally root irritation. Radiographs reveal bony destruction with preservation of the bony cortex and new bone formation.

It is a slowly progressive malignant tumour, making total excision the treatment of choice but this is often difficult because of the large size and problem in circumscribing the tumour. There have been no documented cases of complete cure of chordomas but many patients survive well for up to 5 years only to develop a recurrence. Our patient with a sacral chordoma is well and free of recurrence 8 years later. Other tumours found in the epidural space are those due to direct or blood spread from carcinomas and sarcomas or result from leukemic infiltration.

Surgical technique requires attention to certain details that are less important in adults. It is well known that multiple-level laminectomy in children predisposes to spinal problems such as kyphoscoliosis and Swan-neck deformity. However, if the posterior articular facets are preserved, removing a number of laminae in the thoracic and lumbar region causes no problem. Kyphosis, anterior subluxation and instability is a distinct hazard in the cervical region and can be prevented by wiring back into position the laminae and interspinous ligaments that are removed en bloc using a high-speed side-cutting drill.10 Small infants are abdominal breathers and in the prone position the abdomen must be allowed to hang completely free by supporting the iliac crests and chest, thereby preventing unnecessary troublesome bleeding from distended epidural veins.1 The use of the operating microscope, micro-instruments and bipolar coagulation has improved technique, especially for the removal of intramedullary tumours.

Factors which influence the outcome are the degree of previous neurological deficit, the extent of tumour removal, the site of the lesion, and its histological appearance and radiosensitivity if malignant.

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REFERENCES