Intraspinal tumours

Spinal cord tumours in children are rare—they occur between five and 10 times less commonly than intracranial neoplasms.^{1 2} The general paediatrician is likely to encounter only one or two children with this condition in a whole lifetime of practice. Presenting symptoms are often vague and it is thus not surprising that these tumours are frequently misdiagnosed. It is not unusual for the interval between presentation and accurate diagnosis to be as long as two or three years.

About 70% of intraspinal tumours in children are benign and slowly growing, with symptoms evolving in a subtle fashion. Nevertheless neurological dysfunction due to cord compression is an ever present danger and the prognosis for neurological recovery is poor once a severe deficit becomes established. Acute spinal cord compression, usually due to an ischaemic insult, may result in a precipitate loss of neurological function below the level of the lesion and even when treated promptly significant recovery may not occur.

Pathological features

The presenting clinical features depend on the cord level of the lesion and also whether the tumour is extradural, intradural but extramedullary, or intramedullary. As a rule of thumb, 40% of intraspinal tumours occur in the thoracic region with the remainder being evenly distributed between the cervical and lumbosacral segments. Extradural tumours (for example, neuroblastoma and sarcoma) account for almost half of intraspinal tumours in childhood, spreading from nearby bone or through intervertebral foramina. Intradural but extramedullary tumours occur in one quarter of patients, the commonest lesions being neurofibromata, dermoids, and lipomata. More than a quarter of all intraspinal tumours in childhood are intramedullary (a significantly higher proportion than in adults) and most of these are slowly growing astrocytomas. Many astrocytomas are relatively well localised but sometimes there is extensive involvement of the cord from the lower brainstem or upper cervical region to the conus. These 'holocord' astrocytomas are associated with large cystic dilations throughout the cord.³

Clinical presentation

Boys are a little more often affected than girls. The highest age specific incidence is found in infancy due to the predilection of neuroblastoma and congenital tumours to present at this time, but no age is exempt.¹

The commonest manifestation of intraspinal tumour in childhood is motor weakness (spastic or flaccid depending on cord level). In the young infant the parents may observe failure or reluctance to move one or both legs, while there may be delay in walking or unwillingness to stand in the toddler. In the school age child progressive motor weakness is often heralded by a limp or other gait disturbance. Occasionally these symptoms are accompanied by segmental muscle wasting.

Back or root pain occurs in at least 40% of children with intraspinal neoplasm but it is a much less common presenting sign in children than in adults. Spinal pain is most often dull and aching in quality and localised to dermatomes adjacent to the tumour. Root pain, which is less frequent, is usually projected over a single dermatome and it is unclear whether this is caused by swelling of the spinal cord with distension of the root, or whether the root entry zone is infiltrated with tumour. Often the site of pain may suggest other more common clinical entities, particularly acute intra-abdominal disease when lower thoracic nerve roots are involved. The child under 4 years will be unable to describe or to localise accurately his discomfort, while in the infant pain must be inferred from irritability.

With slowly growing tumours, neuromuscular imbalance and growth may produce musculoskeletal deformity such as head tilt, torticollis, kyphosis, or scoliosis. Such deformity is commonly accompanied by pain, especially night pain. In any child who presents with a progressive musculoskeletal deformity, even in the absence of neurological deficit, intraspinal tumour must be considered.^{4 5}

Various degrees of bladder and bowel dysfunction and other sensory disturbances are present in about 30% of children at the time of presentation. A history of delayed development of sphincter control is suspicious, but even more significant is a change in bladder or bowel habit in the previously continent child. One of the major pitfalls in diagnosis is the

failure to appreciate the significance of more trivial symptoms such as constipation, anal pain, or secondary enuresis in the child presenting with limp, backache, or leg pain. The symptoms of a patient suffering from intraspinal tumour are often exacerbated by coughing, crying, or straining. Occasionally symptoms are first highlighted by trauma and sometimes these may misleadingly diminish or even disappear, only to recur more insidiously later.

Under exceptional circumstances, intraspinal tumours may present as hydrocephalus, most likely due to raised protein concentrations causing defective absorption of cerebrospinal fluid. Intraspinal dermoid tumours may present as recurrent meningitis if there is a direct extension from the skin surface.

Neurological examination may show a plethora of abnormal signs or no abnormality. Upper motor neurone findings may be less well defined than lower motor neurone findings, the latter signs generally offering a better indication of the level of the lesion, unless the abdominal reflexes are involved. Local tenderness over one of the spinous processes may also suggest a level. Detailed sensory examination is obviously of limited value in the very young patient but some idea of the presence or absence of perianal anaesthesia and pinprick levels can usually be obtained even in infants. Inspection of the skin may suggest neurofibromatosis whereas hepatomegaly provides a clinical clue in neuroblastoma.

Investigation

Plain radiographs of the entire spine (anteroposterior and lateral views) should be carried out as they often show bony abnormalities and strengthen clinical suspicions.⁶ There is no place for routine lumbar puncture in the management of the child. Although examination of the cerebrospinal fluid may be useful for demonstrating a raised protein content or the presence of tumour cells, this advantage is greatly outweighed by the potential hazards of the procedure. Lumbar puncture may lead to further cord damage at the site of compression, particularly in cases of spinal block where tumour has obstructed the cerebrospinal fluid circulation and removal of fluid below the lesion leads to downward displacement of the cord and tumour. Ill considered lumbar puncture may also delay successful myelography for several days.

Further investigation of the child should be undertaken by a paediatric neurologist at a neurosurgical centre. Computed tomography of the spine and in particular magnetic resonance imaging

of the spinal cord are relatively new imaging techniques which may rapidly confirm the presence of an intraspinal mass.^{7,8} High resolution computed tomography with intrathecal contrast enhancement is a valuable but nevertheless invasive procedure and several studies have shown the superiority of magnetic resonance imaging in diagnosing spinal cord tumours in children.^{8,9} Magnetic resonance imaging is a non-invasive method of providing high resolution images which differentiate not only between cystic and solid tumour areas but also between tumour and surrounding oedema.

In most centres myelography remains the definitive investigation. In the presence of an intraspinal mass, myelography itself sometimes leads to rapid deterioration in neurological function. The investigation should thus be regarded as an integral part of the neurosurgical procedure, carried out with the full knowledge and availability of the neurosurgeon. The opportunity for examination of the cerebrospinal fluid should be taken at this time. In very few patients will myelography precipitate urgent exploratory laminectomy and surgery may be safely deferred for several days in most children.

The treatment of the tumour depends on the site and histological diagnosis of the lesion. Advances in microsurgical techniques including the use of the ultrasonic aspirator (Cavitron) and carbon dioxide and argon lasers, along with refinements in radiotherapy have led to a greatly improved outlook, especially in intramedullary tumours.^{3,10,11}

It is clear that early recognition of these rare tumours is important, as their effects are often devastating whereas prompt surgical intervention may produce very favourable results. Although new non-invasive imaging techniques promote swift and sometimes precise definition of such lesions, there is little doubt that a high index of suspicion remains our most valuable diagnostic tool.

References

- Di Lorenzo N, Giuffrè P, Fortuna A. Primary spinal neoplasms in childhood: analysis of 1234 published cases (including 56 personal cases) by pathology, sex, age and site. Differences from the situation in adults. *Neurochirurgia* 1982;25:153-64.
- Till K. *Paediatric neurosurgery for paediatricians and neurosurgeons*. Oxford: Blackwell Scientific Publications, 1975.
- Epstein F, Epstein N. Surgical treatment of spinal cord astrocytomas of childhood. *J Neurosurg* 1982;57:685-9.
- Taylor LJ. Painful scoliosis: a need for further investigation. *Br Med J* 1986;292:120-2.
- Citron N, Edgar MA, Sheehy J, Thomas DGT. Intramedullary spinal cord tumours presenting as scoliosis. *J Bone Joint Surg [Br]* 1984;4:513-7.
- Brett EM. *Paediatric neurology*. London: Churchill Livingstone, 1983:457-60.
- Hagenau C, Grosh W, Currie M, Wiley RG. Comparison of

- spinal magnetic resonance imaging and myelography in cancer patients. *J Clin Oncol* 1987;**5**:1663–9.
- ⁸ Aichner F, Poewe W, Rogalsky W, Wallnofer K, Willcit J, Gerstenbrand F. Magnetic resonance imaging in the diagnosis of spinal cord diseases. *J Neurol Neurosurg Psychiatry* 1985;**48**: 1220–9.
- ⁹ Kucharczyk W, Brant-Zawadzki M, Sobel D, *et al.* Central nervous system tumors in children: detection by magnetic resonance imaging. *Radiology* 1985;**155**:131–6.
- ¹⁰ Edwards MS, Boggan JE. Argon laser surgery of pediatric neural neoplasms. *Childs Brain* 1984;**11**:171–5.
- ¹¹ Garrido E, Stein BM. Microsurgical removal of intramedullary spinal cord tumours. *Surg Neurol* 1977;**7**:215–9.

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