Misdiagnosis of intraspinal lesions in childhood

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Summary

Three children with intraspinal mass lesions in whom the diagnosis was initially missed are described. Their case histories highlight the specific clinical features of and diagnostic difficulties with the syndromes produced by lesions of the craniocervical junction, the mid-thoracic spinal cord and the cauda equina.

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Intraspinal space-demanding lesions often present difficult diagnostic problems resulting in long delays before correct diagnosis. Since many of the lesions are surgically correctable, it is of great importance that the clinical syndromes of early cord compression are recognised before irreversible cord damage results. The anatomy and physiology of the spinal cord as well as the principal syndromes of disease are fully discussed in several excellent textbooks.[1-3] In childhood the diagnostic dilemmas are, for many reasons, of increased magnitude. The incidence of compressive lesions of the spinal cord in children is much less than in adults and these lesions occur much less frequently than the intracranial space-demanding lesions of childhood.[4,5] In the young child the history may be inadequate or even misleading. Furthermore, neurological examination of the baby or toddler may be inconclusive, particularly in the assessment of sensory deficits. However, careful clinical assessment, a high index of suspicion and appropriate investigations should achieve a better diagnostic success rate.

The Paediatric Neurosurgical Service of the University of Cape Town has an incidence per annum of 4 new patients presenting with intraspinal tumours.[6] Some of these patients, in whom the diagnosis was unsuspected, were initially referred for opinion to the Paediatric Department, Groote Schuur Hospital. From these patients 3 have been selected, since they illustrate specific syndromes that, by their location, are distinctive in symptomatology but are seldom diagnosed early in their course.

Case presentations and comment

Case 1 — lesion of high cervical cord-craniocervical junction

Four weeks before referral this previously well 11-year-old girl tripped and fell; this resulted in immediate severe pain in the neck and occiput. The pain improved the next day but over the next 2 weeks she developed attacks of occipital headache of increasing frequency and severity. Over the same period she developed episodes of dyspnoea with sighing respiration and complaints of 'air hunger' especially when recumbent at night. She also complained of paraesthesia in the hands and subsequently in the feet. Neurological examination, radiographs of cervical spine and skull and computed tomography (CT) of the brain were all reported to give normal results. Two weeks after onset of symptoms an acute anxiety state with hyperventilation was diagnosed and amitryptilene and lorazepam prescribed. Over the next 2 weeks she also complained intermittently of increasing weakness and increasing difficulty in walking. During the 5 days before referral the occipital headache, dyspnoea, paraesthesia and limb weakness worsened. Three days before referral she developed urinary incontinence. The limbs were noted to flex spontaneously although she could not move them on command. At this stage the referring doctor realised neurological disturbance was present but could not localise the disease.

Examination revealed a restless, whining and anxious child with sighing respiration. She was fully conscious with no cognitive deficit. Cranial nerves were normal. The neck was held rigidly and attempted passive movement resulted in resistance and pain. Attempted neck flexion caused tingling down the back and legs (Lhermitte's phenomenon). The limbs were hypotonic but all muscle stretch reflexes were symmetrically brisk. There was marked bilateral finger flexion, positive Hoffmann's sign and bilateral extensor plantar responses. No voluntary movement was possible but reflex-flexor withdrawal occurred as a result of noxious stimulation of the limbs. The patient had continuous dribbling of urine and a distended bladder could be felt. Anal tone was decreased. Impairment of finger and toe position sense and arm and leg vibration sense was noted. Cutaneous examination was difficult and no sensory level was obtained. No sweating abnormalities were noted.

Myelography revealed an intradural tumour extending from C3 to above the foramen magnum. The spinal cord was displaced posteriorly and to the right (Fig. 1). Removal of a large angiomatous meningioma at the craniocervical junction was urgently undertaken. The patient's postoperative course was long and complicated. Six months after surgery she was discharged from hospital with residual paraplegia.

Comment

This case illustrates how early symptoms of a craniocervical junction lesion can be mistaken for those of anxiety. The onset of symptoms after minor trauma are well recognised in lesions of the spinal cord.[7] High cervical cord and posterior fossa lesions should be considered in children complaining of bouts of 'occipital headaches'. Neck movements will often exacerbate the pain and result in cervical muscle spasm and splinting of the cervical spine. Fixed painful torticollis or retrocollis may result and, in the younger child, these postural abnormalities may be the presenting clinical problem.[8] Paraesthesia is a well-recognised symptom of peripheral polyneuropathies and psychogenic hyperventilation syndrome, but may also occur with involvement of the posterior columns of the spinal cord. Such involvement may also result in Lhermitte's phenomenon, which is considered pathognomonic of a cervical cord lesion.

By the time the patient was examined on referral obvious deficits of posterior column function with impairment of position and vibration sensation in all four limbs were present. It is imperative to examine these sensory modalities. It is much more difficult to examine touch and pain sensation in a young child and often a sensory level may not be demonstrable despite quadriplegia and urinary incontinence. In addition,

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high cervical cord lesions besides impairing pain over the entire trunk can result in pain impairment over the scalp and part of the face (owing to involvement of the spinal nucleus of the trigeminal nerve). Thus, few areas of normal sensation will be available for comparison.

Such patients are at great risk of developing respiratory problems. Bilateral anterior high cervical cord lesions may result in complaints of 'air hunger' (inability to get enough air), sighing, confusion, sleep apnoea or respiratory arrest.[3] Reflex spinal flexor withdrawal of the limbs is often wrongly considered to mean that the patient is capable of voluntary movement and bladder dysfunction may be misinterpreted as psychogenic bed wetting. Other neurological syndromes associated with craniocervical junction lesions have been well documented.[9]

Case 2 — lesion of cauda equina

This previously healthy 11-year-old boy developed increasingly frequent and severe episodes of lower lumbar sacrococcygeal backache. One year after onset a diagnosis of coccydinia was made and coccygectomy was performed. Postoperatively the low backache worsened and radiated into the left hip and knee. Walking aggravated the pain and resulted in weakness of the left leg and a limp. Four months after surgery he was found to have loss of lumbar lordosis with associated lumbar scoliosis. The results of neurological assessment were interpreted to be normal. The left sacro-iliac joint was tender. Radiography of the sacro-iliac joints was interpreted to show arthritic change and ankylosing spondylitis — tuberculous arthritis or septic arthritis were considered. He was treated for these suspected infections but failed to improve, so he was referred to our clinic 2 years after the onset of symptoms with a diagnosis of ankylosing spondylitis.

Examination revealed a well-looking boy with fixed kyphoscoliosis of the lower dorsal and lumbar spine. He walked with flexed hips and knees and had bilateral Trendelenburg signs. Movements of the lower back were painful and limited by paraspinal muscle spasm, but straight leg raising was full bilaterally. His buttocks were wasted and hip extension and abduction showed 4/5 power bilaterally. Knee and ankle reflexes were normal bilaterally. Plantar responses were flexor. No sensory abnormalities were detected over the perineum, buttocks and the back of the legs. The anal reflex was normal.

A myelogram revealed a large intradural mass extending from the lower border of the L4 vertebra downwards resulting in a complete intradural block (Fig. 2). Following myelography, the patient developed plantar flexion weakness of both feet. Impaired sensation was noted over the buttocks and the back of the legs and the ankle reflexes could not be elicited. Laminectomy revealed a massive intradural tumour, the bulk of which was removed. On histological examination it was shown to be a myxopapillary ependymoma of the cauda equina. The patient received radiotherapy and is well 9 years after surgery. He is able to run marathons and the only residual neurological deficit is bilateral absent ankle reflexes.

Comment

This case illustrates how a cauda equina lesion can be misdiagnosed as skeletal or rheumatic disease.

Lower back pain is the most common presenting feature of cauda equina lesions. It may remain localised or may radiate into the lower abdomen, groin, anterior thigh, back of the leg, buttocks or perineum. Occasionally the patient may only complain of the radiating pain.[10] Since disc lesions are decidedly uncommon in childhood, any radicular pain must be regarded as suspicious.[11]

Paraspinal muscle spasm secondary to pain is the most common finding. This spasm results in limitation of spinal movements with loss of lumbar lordosis. The combination in a child of paraspinal muscle spasm (with or without deformity)

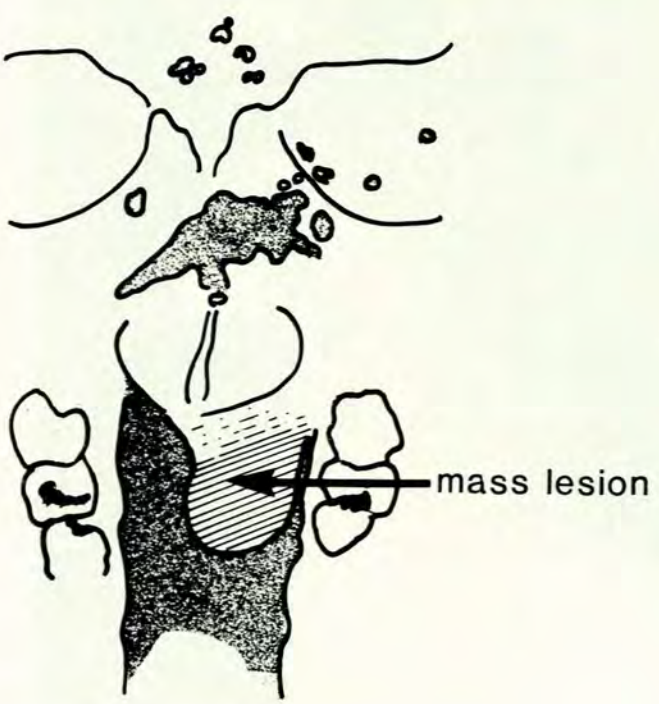

Fig. 1(a). Anteroposterior view of myelogram of intrathecal lesion of craniocervical junction extending from C2-3 to above the foramen magnum.

Fig. 1(b). Lateral view of myelogram of intrathecal lesion of craniocervical junction extending from C2-3 to above the foramen magnum.

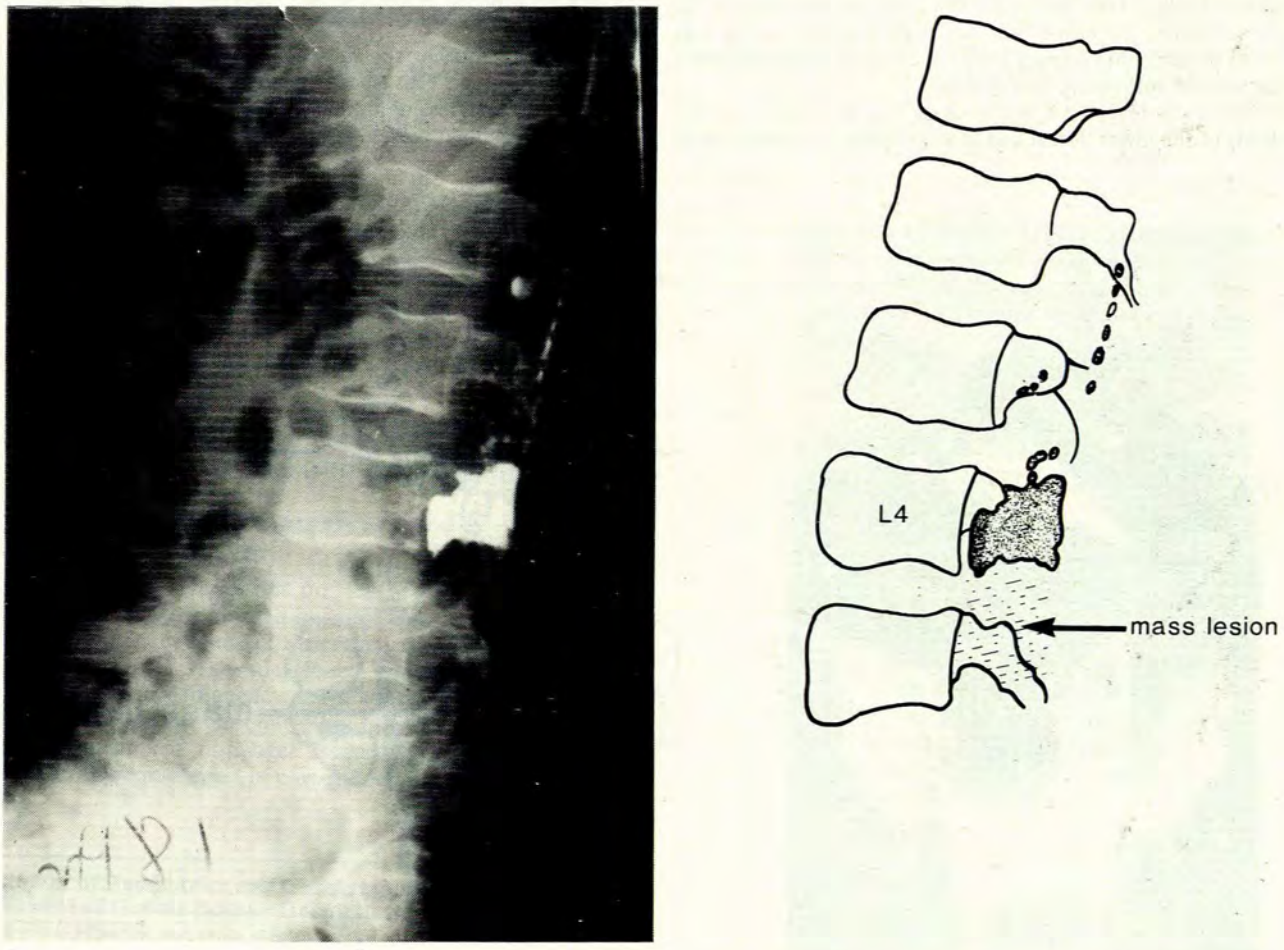

Fig. 2. Myelogram of intradural mass in cauda equina extending downwards from lower border to L4.

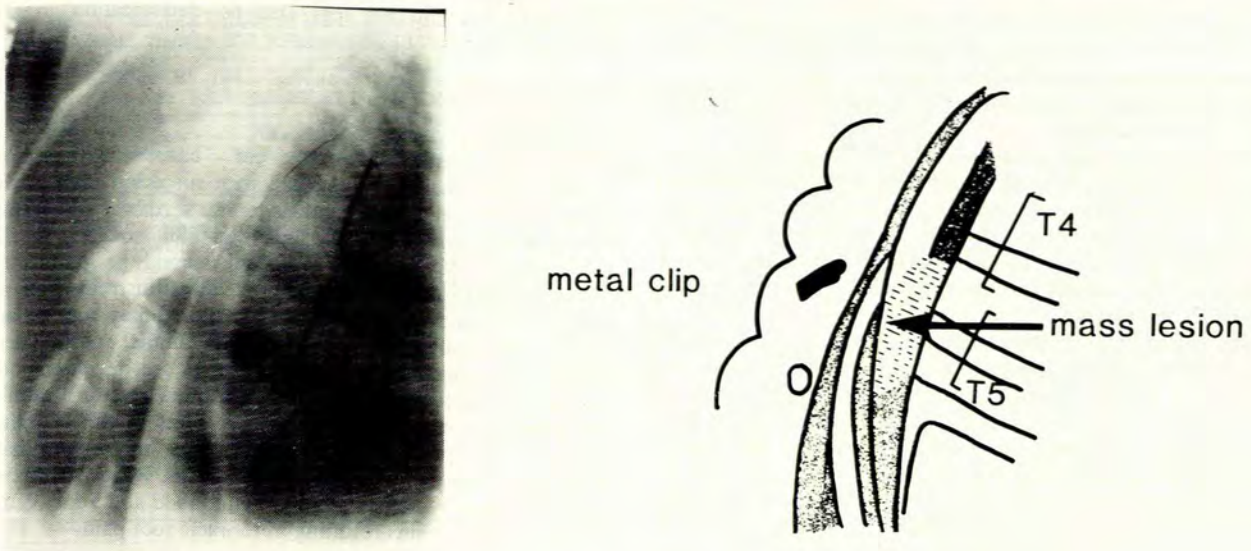

Fig. 3(a). Lateral view of myelogram of extradural mass at T5 level with anterior cord compression.

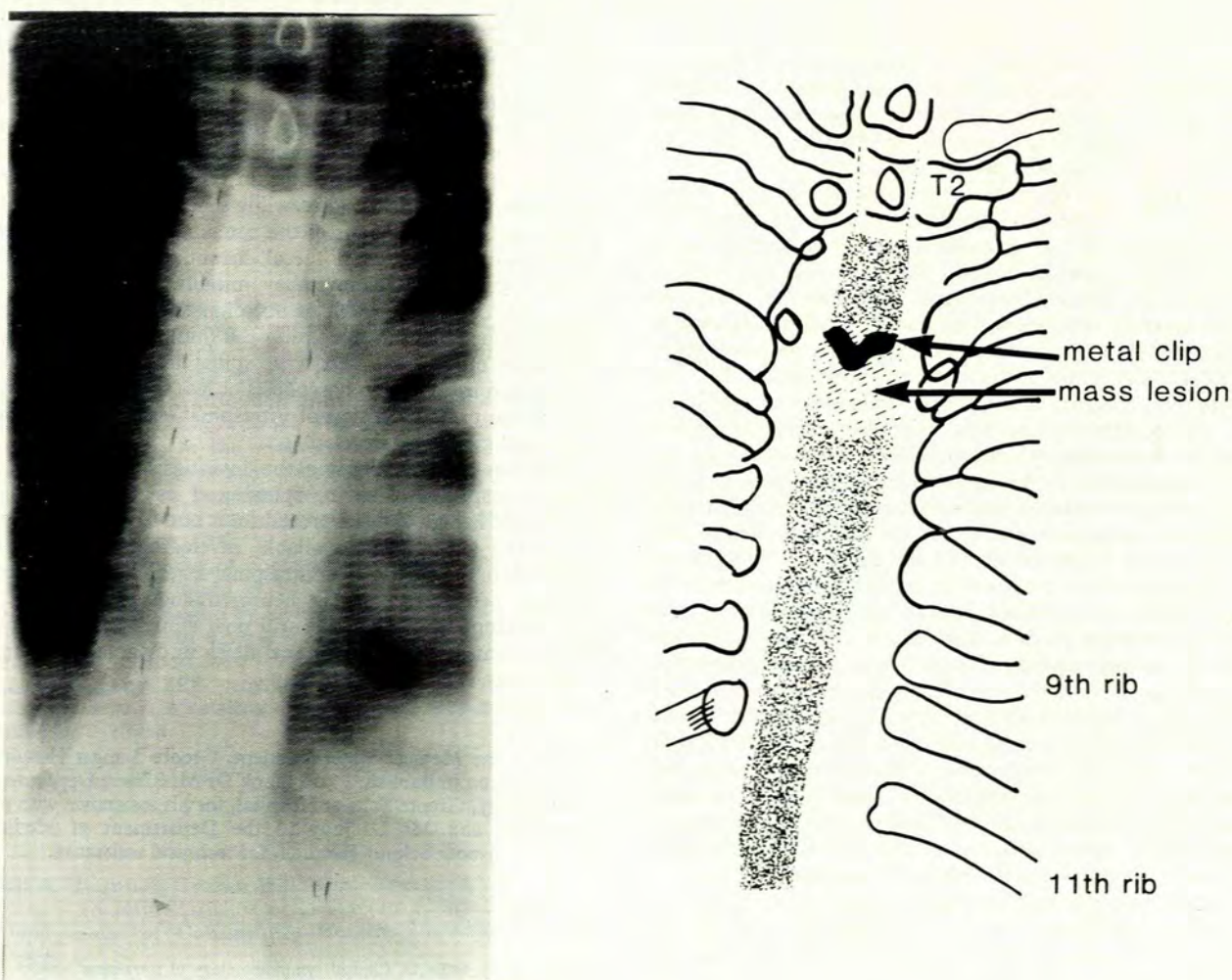

Fig. 3(b). Postero-anterior view of myelogram of extradural mass at T5 level with anterior cord compression.

and persistent severe backache (with or without root pain) not relieved by rest and worse at night is enough to warrant thorough investigation to exclude, among other causes, intraspinal mass lesions. To wait for the development of neurological signs is to wait too long.

In a child of this age a diagnosis of ankylosing spondylitis is highly improbable, particularly with such severe lumbar deformity in the absence of any preceding history of pauci-articular arthritis of the legs. Fuzzy, ill-defined sclerotic joint margins on radiography of the sacro-iliac joints during adolescence is a variation of the normal and does not allow for a diagnosis of ankylosing spondylitis.

After the myelogram the patient's neurological state deteriorated, a phenomenon known to occur in patients with intraspinal mass lesions. Neurosurgical consultation should always be obtained before myelography when a mass lesion is suspected, since urgent decompression after myelography may be required. If adequate magnetic resonance imaging (MRI) of the spinal cord is available, the need to perform a myelogram may be obviated unless specific cerebrospinal fluid tests, such as cytology, are considered necessary.

Case 3 — lesion of anterior middle thoracic cord

A 12-year-old boy, who had had a normal perinatal period, manifested some motor developmental delay, sitting alone only at 1 year and walking alone at 2 years. Arm function, speech, social and mental skills were acquired normally. By the age of 3 years his back was noted to be 'skew' and his gait clumsy. He developed attacks of abdominal pain diagnosed as abdominal epilepsy. Spinal radiography demonstrated a kyphoscoliosis with congenital fusion of the anterior aspect of T7-8-9 vertebrae. At the age of 6 years a spinal fusion was performed because of deterioration of the kyphoscoliosis. Since the patient was considered to be neurologically normal, myelography was not performed. He was assessed as having perceptive motor problems with clumsiness of his hands and feet for which he received occupational therapy and physiotherapy. From the age of 10 years the patient's gait began deteriorating and he walked with hunched back and flexed legs. He was admitted to a cerebral palsy school at the age of 12 years with the diagnosis of cerebral diplegia. Increasing gait difficulties and spasticity with marked flexion of his legs resulted in referral for myelography with the diagnosis of a possible spinal cord lesion.

On examination at the age of 12 years the patient was a well-looking co-operative boy with the only abnormal findings confined to his back deformity and the nervous system. There was a marked kyphoscoliosis of the thoracic spine with an overlying scar from the previous operation but no dermal or subcutaneous abnormalities were noted. His legs were flexed at the hips and the knees with marked bilateral spasticity. The power in the legs was reduced to 4/5 in all muscle groups. The knee and ankle reflexes were bilaterally brisk with bilateral ankle clonus. Plantar responses were bilaterally flexor. Abdominal reflexes were present and there was no disturbance of sphincter function. No sensory deficits were elicited and, although he complained of excessive sweating over the chest, no objective change could be detected over the chest and abdomen. The results of neurological examination of the arms, cranial nerves and mental function were normal.

A myelogram demonstrated an extradural mass at the level of the T5 vertebra with anterior spinal compression (Fig. 3). An anterior spinal cord decompression was performed and a mass was excised. Histological examination revealed a multilocular cyst lined by pseudostratified ciliated columnar epithelium, these being the features of an enterogenous cyst of the spinal cord. Five years later he remains well with normal power and tone in his legs and no deterioration in the kyphoscoliosis.

Comment

This case illustrates how the early onset of progressive kyphoscoliosis should be regarded with suspicion even in the absence of neurological signs. Congenital vertebral abnormalities raise the possibility of congenital abnormalities of the spinal cord.[12] Myelography or MRI should be performed before spinal fusion is undertaken. It also illustrates how an anterior midline mass lesion of the spinal cord may be misdiagnosed as cerebral palsy or, on occasion, familial spastic paraparesis.[1] Enterogenous (neurenteric) cysts are developmental foregut anomalies resulting in an intraspinal cyst or a dumbbell intra- and extraspinal cyst with defects in the vertebral bodies.[13] This produces slowly developing motor weakness preceding sensory or sphincter disturbances. Recurrent abdominal pain is extremely common in childhood and a rare cause is thoracic cord lesions resulting in thoracic root pain.[13]

Conclusion

The delay in diagnosis of spinal cord lesions in childhood alarms and frustrates those involved in the care of young patients. The incidence of spinal cord tumours in children with regard to age, location of tumour, tumour type, etc. varies among the series reported. However, the symptomatology is essentially the same in all the series. The older the child the more likely are the symptoms to resemble those of adults. The initial diagnostic feature almost always falls into one of two groups, being either early motor disturbances of gait (such as frequent stumbling and weakness or heaviness of limbs after exercise) or sensory abnormalities in the limbs (such as 'pins and needles', numbness and crawling sensation, and distortion of size and shape). Lesions of the conus medullaris, however, present with urinary and faecal incontinence. In younger children the clinical picture may initially be dominated by pain, with complaints such as occipital headache, neck pain and backache.[14] In this age group the finding of limitation of movement often associated with spinal deformity, such as torticollis/retrocollis, kyphoscoliosis or loss of lumbar lordosis, may be the presenting sign of intraspinal lesions long before the neurological signs develop.

MRI has recently become the investigation of choice in children suspected of having spinal cord lesions.[15] The diagnostic yield is high and the procedure is non-invasive and safe. Since MRI is not freely available, myelography still plays a major role in the diagnosis of intraspinal lesions and is preferred by some neurosurgeons and neuroradiologists. However, it must be emphasised that it is still poor clinical assessment of these patients that causes the real delay in diagnosis of spinal cord lesions.

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