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Clinical Case Series

Neurological deterioration years after closure of myelomeningocoele – 'the second lesion'

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Case presentation

MP is a 36-year-old man who had an upper lumbar myelomeningocoele closed on the first day of life. He had a degree of hydrocephalus but never required any treatment for this. As a child there was a marked disparity in lower limb function – with the right leg near normal but with the left leg spastic, very weak and shortened. He had undergone left adductor tenotomy and psoas transplant aged 10 years and had a left femoral fracture at 16 years but remained well motivated and was mobile with elbow crutches and calipers. He had a neurogenic bowel, a neuropathic bladder with a suprapubic catheter, and preserved sexual function. Mobility and function otherwise were generally static over the next 20 or so years.

At the age of 35 years he presented to his GP with a year's history of worsening back pain, especially severe at night, disturbing sleep. This was associated with loss of sensation of the previously normal right lower limb. He was referred to a Spinal Orthopaedic Clinic and the referral was prioritised as 'routine' with an expected waiting time of 53 weeks. Twelve months later his pain and sensory loss had worsened and he was having difficulty weightbearing on the right leg: his outpatient appointment was expedited. In the interim he had a fall fracturing his left tibia. By chance, he was seen for this at the Fracture Clinic by an Orthopaedic Surgeon with a Paediatric interest; he reported an abrupt marked deterioration in power in the right leg some 3 weeks earlier such that he could no longer walk and was admitted for investigation and a neurosurgical opinion.

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By now he had no power or sensation in the left leg, and in the right leg had Grade 2 hip flexion, Grade 3 knee extension, no power at the ankle and a suspended sensory loss from L3 to S1. MR of the neuraxis demonstrated a moderate supratentorial hydrocephalus and a Chiari II malformation with the tonsils down to C2 though with no compression at the foramen magnum. In the spine (Figures 1 and 2) there was an extensive intradural mass lesion lying posterior to the neural tissue at the level of the myelomeningocoele repair and a moderate size thoracolumbar syrinx above this. The differential diagnosis of this lesion was felt to be either a dermoid cyst or a tumour.

Comments by participants

What is your differential diagnosis?

M Spencer (General Practitioner) points out that he sees very few cases of spina bifida at any age and finds this difficult, but highlights possibilities such as a problem arising as a direct late consequence of his myelomeningocoele or secondary mechanical or musculoskeletal consequences due to abnormal posture and use of the lower body. As such he would consider disc prolapse as well as 'non-back' causes of back pain on the differential including urinary tract infection and constipation.

DL Douglas (Orthopaedic Surgeon) also pays heed to incidental pathologies adding meningioma or AV malformation to the list. He mentions that patients with spina bifida and asymmetric neurology frequently have back deformities such as scoliosis. He would also consider the possibility of a symptomatic syringomyelia but ultimately feels spinal cord tethering to be the

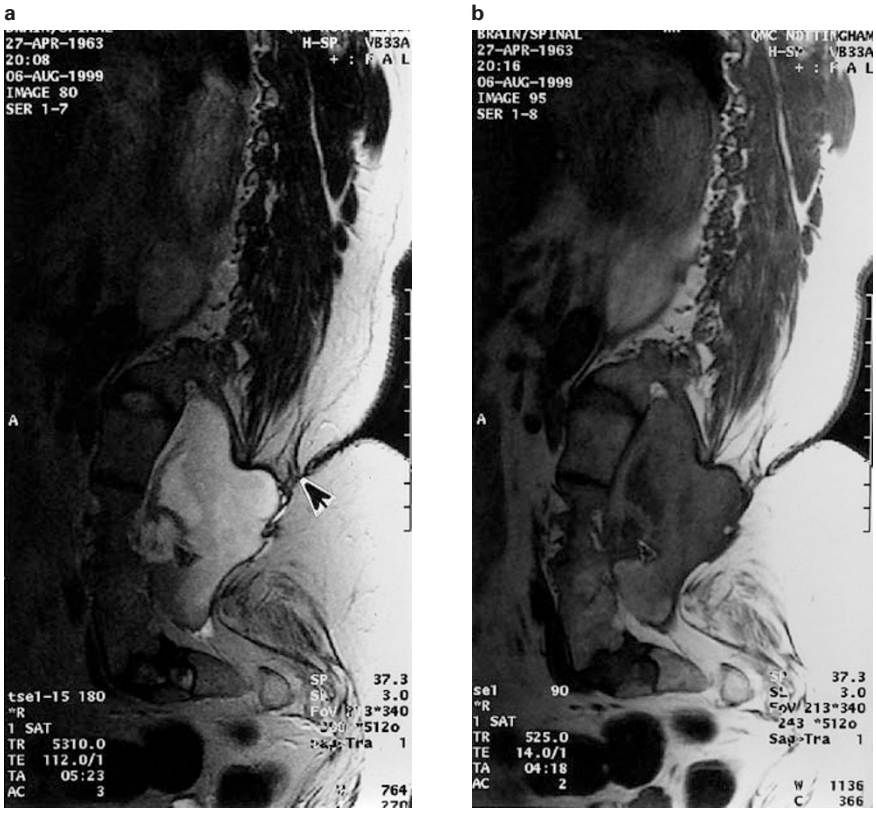

Figure 1 Sagittal MR images through the lower lumbar lesion with T2 (**a**) and T1 (**b**) weighting showing the complex kyphoscoliosis and repaired lumbar myelomeningocoele. A track (arrowed) extends from the skin surface to merge with a very large (95×55 mm) heterogeneous soft tissue lesion which fills and expands the lumbosacral spinal canal

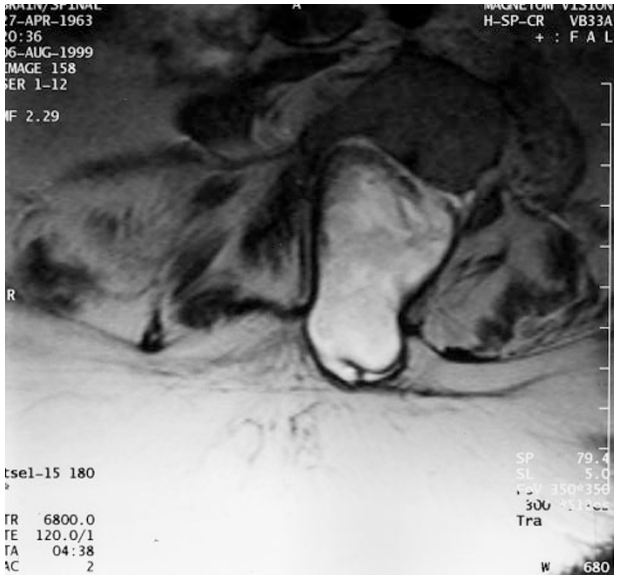

Figure 2 Axial T2 weighted MR image approximately at the midpoint of the lesion

most likely cause having seen several such patients present at this age.

Is the nocturnal pain relevant?

M Spencer feels that the lack of relief at rest would make a mechanical problem less likely, and DL Douglas emphasises that he considers this a sinister symptom of spinal compression. BD White (Neurosurgeon) agrees saying that the nocturnal pain suggests an organic pathology other than a degenerative cause but does not in itself constitute an emergency. He would certainly hope to be informed however of the development of an objective and progressive neurological deficit while a patient with these symptoms was waiting for an appointment.

Who should he be referred to, and what should happen at clinic?

M Spencer feels that as the initial condition is neurological, a neurosurgical opinion and investigation would be most appropriate and would make the referral to a hospital specialist by letter. Recognising his lack of experience of the problem and unaware of exact potential waiting times for clinic he would avoid a Central Appointments system but direct the letter via a specific consultant to allow him or her to make their own decision on urgency or perhaps back the letter up with a phone call to say that the referral is on its way.

At clinic DL Douglas would carry out a thorough examination, arrange plain X rays of the thoracolumbar spine, possibly including bending views and would expect the single most useful investigation to be MR, though an EMG might be considered.

IM Holland (Neuroradiology) would hope to perform MR in 4–6 weeks if he was referred with back pain alone, 3–4 weeks if the nocturnal pain was mentioned, and 1–2 weeks in the presence of the progressive loss of power.

Having seen the imaging he would have reported the differential diagnosis thus – 'This is a complex spinal dysraphic syndrome with a very complicated cord tethering. The appearances could represent:

(1) complex tethered cord with lipomyelomeningocoele
(2) lipomyelomeningocoele with some associated intradural tumour
(3) inclusion dermoid in view of previous history
(4) possible organised chronic infected collection
(5) PNET (primitive neuroectodermal tumour) type tumour'.

Would you be keen to operate by the time he was admitted? What would you do? Do you foresee any particular difficulties?

BD White if presented with this patient at admission, with grade 3 weakness at best in his right leg but with the prospect of rapid deterioration to complete paraplegia, would offer him exploration, decompression and untethering of the lumbosacral cord in the hope of retaining residual function, with some possibility of recovery but no expectation of improvement to his previous best. He feels that the hydrocephalous is not symptomatic and although there is a Chiari II malformation this is without tonsillar impaction in a capacious foramen magnum and upper spinal canal. He would however be wary that treatment of the low lumbar meningocoele might risk decompensation of the supratenorial hydrocephalus which could be addressed by insertion of a ventriculoperitoneal shunt if required.

Actual management

An operation to establish the nature of the lesion and decompress the neural tissue was discussed with the patient and his wife. They understood that no improvement could be guaranteed and that indeed there was a risk of deterioration – in particular the possibility of decompensating the hydrocephalus was discussed with them. At operation a huge intramedullary dermoid was encountered containing typical keratinous material and hairs and a macroscopic excision of this was performed. No attempt was made to reconstitute the spinal cord and the dural closure was protected with a deep extradural drain brought out at a distance from the incision.

Histology confirmed the diagnosis of dermoid cyst. There was some immediate improvement in right lower limb power to Grade 4 by the 5th postoperative day but his postoperative course was then complicated by the development of a wound infection. He had a pyrexia, headache and neck stiffness and altered mental state. Coliforms were cultured from drain fluid and, without formal CSF sampling, a presumptive diagnosis of bacterial meningitis was made. With appropriate intravenous antibiotics these problems settled and he was able to be discharged some 4 weeks postoperatively, free of back pain although sadly with the loss of the power which had initially appeared to be improving in the right leg.

He was readmitted a month later having developed a frank CSF fistula at the site of the original myelomeningocoele repair and more recent operation. He was apyrexial and otherwise well. Following discussion of the options ('wait and see' *versus* CSF diversion) with the patient and his wife a right ventriculoperitoneal shunt was inserted once it had been confirmed that the CSF discharging from the wound was sterile. This resulted in rapid resolution of the fistula. He remains well and has regained considerable power in the right lower limb.

Analysis

Approximately 85% of live born infants with neural tube defects now survive into adulthood and current provision of care for this group is often less than ideal. There remains a misconception that surgical closure of a myelomeningocoele creates a static situation when in fact up to 15% of such patients can expect to have a secondary deterioration due to tethering of the spinal cord, anything from a few years to decades later.[1,2] Magnetic resonance imaging can differentiate a tethered cord from a more complex situation as found here. The differential diagnosis of 'the second lesion' includes hydrocephalus, hindbrain herniation with or without hydromyelia, tethering of the placode, and intraspinal arachnoid cyst, dermoid cyst, or tumour.[3]

Clinical presentation of such lesions is rather variable including progressive scoliosis, back pain and other deficits which may in some cases be difficult to extract from the patients' background neurological impairments and tragically may often be accepted by the patients themselves as inevitable consequences of their underlying condition. Curiously in many cases there is an abrupt deterioration in power or in sphincter function, suggesting that ischaemia perhaps from repeated trauma of head, neck and trunk movements has some role in the pathophysiology. Surgical untethering can prevent further deterioration and alleviate symptoms and it should certainly always be considered.[3,4]

The situation is complicated in the presence of hydrocephalus which will usually be concomitant. If the hydrocephalus has been treated by shunting it is very important to take a 'top down' investigative

approach and consider whether the secondary deterioration is really a manifestation of shunt dysfunction.[5] Unfortunately where there has never been a need for CSF diversion, as in this case, any intradural procedure runs the risk of destabilising a compensated situation with the development of symptomatic hydrocephalus or CSF fistula. Here there was the additional disappointment that neuroendoscopic third ventriculostomy, which is becoming the procedure of choice in management of many cases of hydrocephalus,[6,7] would not solve the problem of the lumbar wound CSF fistula although he has done very well following the insertion of a ventriculoperitoneal shunt.

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