SPONTANEOUS TRANSPOSITION OF THE SPINAL CORD

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Extensive posterior spinal defects are common in spina bifida. We report a patient whose severe scoliosis, in association with marked lordosis and rotation, allowed the spinal cord to herniate through the osseous deficit. The cord came to lie outside the spinal canal, bowstringing across the concavity of the curve. This rare abnormality was found at operation for posterior fusion and added considerable problems to an already demanding operative technique.

Spinal deformities are prevalent in patients with myelomeningocele; they result from paralysis, bony defects, and the adverse influence of gravity. Operative treatment is indicated when, as a direct result of spinal deformity, there is impending loss of stability while sitting, or loss of the ability to walk; this may occur at any time up to skeletal maturity (Hall and Poitras 1977). Anterior and posterior spinal fusion and instrumentation are both necessary because of the loss of muscular support for the trunk (O'Brien, Dwyer and Hodgson 1975). During the posterior approach to the spine, the dural sac, surrounded by scar tissue representing the closed meningocele, is commonly encountered in the region of the apex of the curve. Once this is dissected clear the remainder of the procedure is not usually hazardous. However, when the spinal cord is bowstrung from top to bottom of the spinal curve in a patient with intact neurological function in the lower limbs, extreme care must be taken. Indeed, failure to recognize the abnormally situated cord in our patient, who could walk, might have resulted in a significantly straighter spine but at the expense of a neurological catastrophe.

A case of spontaneous transposition has previously been described (Hamilton and Schmidt 1975) but it did not cause sufficient morbidity during life to be recognised and was demonstrated only at necropsy. The patient was not myelodysplastic and, since no cause could be identified, was labelled as a case of "idiopathic scoliosis".

CASE HISTORY

A girl aged 15 years 4 months had been born with a lumbosacral myelomeningocele which was closed at birth. Several days later a ventriculo-atrial shunt was inserted to relieve hydrocephalus. During the subsequent eight years she had several orthopaedic operations including transfer of the right iliopsoas for a subluxated right hip.

When she first presented to the spina bifida clinic in 1973, she was found to be intelligent with good daytime urinary control. Clinically the neurological lesion was at the level of L4, with power of the hip flexors recorded as Grade 4, the hip extensors Grade 0, the quadriceps Grade 3+, the hamstrings Grade 4, and the dorsiflexors and the evertors of the foot Grade 3. There was patchy sensation in both legs. She was able to walk with the help of bilateral long-leg calipers, a pelvic band, and crutches.

She was noted to have a collapsing type of lumbar scoliosis, convex to the left with marked pelvic obliquity.
(Fig. 1). Above this was a smaller, thoracic curve with a hemivertebra at T4. As a result of increasing severity of her lumbar scoliosis she was experiencing difficulty with walking and sitting; she was unable to sit without the support of one arm and could not regain her balance if pushed to one side.

It was therefore decided to perform anterior and posterior instrumentation and fusion. Radiographs taken before operation showed that the curve corrected from 125 degrees in the sitting position to 110 degrees in the supine position. Myelography showed no evidence of diastematomyelia, hydromyelia, or diplomyelia, although a large dural sac was present in the lumbar region (Fig. 2). Cardiorespiratory and urological examination provided no contra-indication for operation.

First, an anterior correction and fusion was performed using Dwyer instrumentation. Six weeks later she had a posterior fusion with Harrington instrumentation. During the second operation the normal upper part of the spinal column was identified and displayed by subperiosteal dissection. Approaching the curve it became apparent that the cord and dural sac were lying outside the protection of the bony canal in the concavity of the curve (Fig. 3). Careful dissection was necessary to avoid damaging intact neural structures. On two occasions the dura was inadvertently opened; each time the defect was immediately closed. At no time was the dura felt to be under tension, nor was the pressure of the cerebrospinal fluid felt to be unduly high. When the spinal cord and nerve roots had been safely identified and retracted, the sacrum was exposed and the lower distraction hook placed in position. The remainder of the operation and instrumentation was completed without further complication (Fig. 4). The patient made an uneventful recovery; she was fitted with a polythene jacket and after one month in a reclining wheelchair was allowed to sit up.

Six months later she developed acute hydrocephalus and died. Her parents kindly gave permission for the coroner’s pathologist to remove the spine en bloc and we received it for subsequent investigation.

Dissection of the spine revealed that the instrumentation was in position and the fusion solid. Detailed examination of the spinal cord and canal showed a wide cystic dilatation of the dura passing across the concavity of the curve. The nerve roots were lying on the concave bowl.
side of the dura being stretched from there to the convexity. It was apparent that the cord had been bow-
strung across the lordoscoliotic spine.

DISCUSSION

Lordosis is the most frequently found deformity of the paralytic spine (Kilfoyle, Foley and Norton 1965; Parsch and Goessens 1971). The lordosis is usually low in the spine and is frequently accompanied by scoliosis and by pelvic obliquity. As the neurological level of the lesion in these patients frequently extends to L3 and L4, the strong action of the flexors of the hips in the absence of effective extensors provides a forward tilting mechanism of the pelvis producing, and aggravating, the lordotic curve. The pelvic obliquity may be transpelvic (a stronger iliopsoas on one side than the other), or suprapelvic (inadequate muscular support for the trunk in the presence of gravity and frequently also of obesity).

The exact mechanism of the transposition of the spinal cord is difficult to define. It can be postulated that as the deformity progressed so the spinal cord became displaced from the bony canal, herniating through the posterior defect to lie in the concavity of the curve. The reason for the failure of myelography to demonstrate such an anatomical relationship may well be the marked rotation which is a constant feature of such curves.

At the second operation there was no doubt about the transposition of the cord, and the bowstringing was confirmed at necropsy. This condition has been reported previously by Hamilton and Schmidt (1975), but they could not account for the phenomenon, which appeared to occur in an idiopathic deformity. In their case, a radiograph taken several years earlier did not demon-
strate any bony defect, yet at necropsy they found absent pedicles throughout the concavity of the curve. They concluded that the loss of bone was due to pressure from the enclosed spinal cord until it burst free to lie in the concavity. That a taut spinal cord could destroy bone by such pressure without giving rise to any form of neurological symptoms is difficult to conceive.

Siriram, Bobechko and Hall (1972) drew attention to the danger of accidentally opening the dural sac during posterior fusion of these curves with the associated high incidence of deep infection and pseudarthrosis. Although in our case the sac was opened on two occasions, no complication resulted.

In conclusion, spontaneous transposition of the cord is an unusual occurrence that may not be detected by routine radiography. Consequently there is a risk of complications occurring both during and after operation. The most likely aetiology is herniation through a posterior defect leading to bowstringing across the concavity.

REFERENCES


