Diastematomyelia in Adults*

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The term "diastematomyelia" in its strict sense refers to a developmental defect in which the spinal cord, or its intraspinal derivatives, is divided longitudinally into lateral halves. Ollivier is credited with the origination of the term in 1837, deriving it from the Greek "diastema," meaning cleft, and "myelos," meaning "marrow" and subsequently spinal cord or medulla. Clinical usage of the term, however, has tended to designate those cases of cord-splitting in which the cord is transfixed by a septum. Most authors differentiate this type of malformation from diplomyelia (double medulla) which is a true duplication of the cord.

Diastematomyelia is rarely diagnosed in adults; only four cases have been described in which the diagnosis was established during the lifetime of the patient. A fifth case is known to us through personal correspondence. Since clinical awareness of this entity was aroused in 1950 through articles by Matson and Neuhauser, more than 100 clinical cases have been described, mostly in children, in whom disturbance of gait, failure of bladder control, and congenital cutaneous defects are the most common features.

We are reporting two cases of diastematomyelia in adults.

Case Reports

Case 1. A 48-year-old man entered the hospital with spasmodic pain, weakness, and tremor in the right leg of 3 months' duration. The symptoms began after a 50-mile ride in a pickup truck.

Examination. Neurological examination showed a spastic paraparesis, loss of pain and temperature sensation over the sacrum, and an unusual patch of hair in the lumbosacral region. Low back films showed a circular calcified shadow at L-5 in the upper portion of a spina bifida occulta (Fig. 1). Lumbar myelography (Fig. 2) demonstrated widening of the spinal canal in the lumbosacral region, a bizarre termination of the dural sac, a filling defect in the center of the oil column at L-5, and a partial block at the L1-2 interspace. It was felt that the partial block was due to a transverse bony ridge secondary to lumbar spondylosis.

Operation. A laminectomy was performed from T-9 to L-5. The spinal cord was greatly lengthened and was divided by a bony spur projecting dorsally from the body of L-5. The bony spur was surrounded by a sleeve of dura mater. The nerve roots in the low lumbar region left the spinal cord at right angles. Several adhesions contributed to traction on the cord. A transverse bony ridge was present at L1-2. The spur and dural sleeve were removed, and the adhesions were lysed.

Postoperative Course. The patient was temporarily worse, developing a paraplegia and urinary and fecal incontinence. He received intensive physiotherapy, and 20 months after surgery had better bladder control and was walking with mechanical aids, though he was still spastic.

Case 2. A 32-year-old woman developed a flaccid paresis of her left leg after a saddle block anesthesia for her fourth delivery. No bowel or bladder difficulty developed. She had had three uncomplicated saddle blocks in the past.

Examination. There was a flaccid paresis of the muscle groups of the left leg and sensory losses in the saddle area and along the medial thigh and calf. Lumbar spine films disclosed a spina bifida occulta at L-3 (Fig. 3). Myelography was carried out at the L1-2 interspace, revealing diastematomyelia at L-3 (Fig. 4). It was elected to follow the patient conservatively, reserving surgical intervention until the symptoms became worse.

Discussion

Although first described in 1837, diastematomyelia had not been identified outside the
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Fig. 1. Case 1. Plain spine film showing calcified nodule in spina bifida defect at L-5.

Fig. 2. Case 1. Myelogram showing widening of spinal canal, filling defect at L-5, and partial block at L1-2 interspace.
autopsy room until 1940, when Herren and Edwards\(^4\) tabulated 42 cases of diplomyelia, only two of which had been discovered during life. Fifteen of the 42 cases were associated with a spur dividing the cord. Other sporadic case reports followed; then, in 1950, 10 cases were reported by Neuhauser and associates and nine by Matson,\(^4\) which were all diagnosed preoperatively and confirmed at operation.\(^4,6\)

In the majority of cases, diastematomyelia has been caused by a spur of bone, fibrous tissue or fibrocartilage which protrudes into the sagittal plane from the posterior aspect of a vertebral body and divides the neural canal. The spur is invariably surrounded by a sleeve of dura mater, and may even be attached dorsally to the dura or to the neural arch. The spur and its dural sleeve form a septum which, by piercing through the spinal cord or by passing between the fibers of the cauda equina, can fix the cord in a low anatomical position, preventing its normal ascent during growth of the spinal column. This fixation of the cord causes traction on the cord which becomes manifest in the form of progressive neurological dysfunction as the individual matures. In some cases, Arnold-Chiari malformation has also developed. The septum usually occurs in the low thoracic or lumbar regions, but it has been described as high as T-2.\(^5\)

The neurological disturbances produced by deformity of the cord vary greatly from individual to individual. Most frequently there is difficulty in walking or in learning to walk, caused by a weakness or paralysis of the muscles in the legs. Deep tendon reflexes
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TABLE 1

Diastematomyelia in seven adults

<table>
<thead>
<tr>
<th>Case</th>
<th>Author</th>
<th>Age</th>
<th>Level of Lesion</th>
<th>Onset of Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Hamby</td>
<td>20</td>
<td>L3-4</td>
<td>delayed</td>
</tr>
<tr>
<td>2</td>
<td>Seaman &amp; Schwartz</td>
<td>20</td>
<td>L2-3</td>
<td>delayed</td>
</tr>
<tr>
<td>3</td>
<td>English &amp; Maltby</td>
<td>48</td>
<td>L-5</td>
<td>delayed</td>
</tr>
<tr>
<td>4</td>
<td>English &amp; Maltby</td>
<td>32</td>
<td>L2-3</td>
<td>delayed</td>
</tr>
<tr>
<td>5</td>
<td>Herren &amp; Edwards</td>
<td>23</td>
<td>T-11</td>
<td>childhood</td>
</tr>
<tr>
<td>6</td>
<td>Seaman &amp; Schwartz</td>
<td>39</td>
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<tr>
<td>7</td>
<td>Bertrand</td>
<td>20</td>
<td>T-11</td>
<td>childhood</td>
</tr>
</tbody>
</table>

will vary with the level of the lesion, but they are usually not hyperactive in children. Pathological reflexes have not often been described. Sphincter disturbances of the bladder and rectum are present in about 50% of the cases. Cutaneous sensation may be diminished in some areas below the waist.

Many associated abnormalities have been reported in the integumentary and skeletal systems. A hairy patch over the affected portion of the spine is probably the most frequently noted cutaneous abnormality, but nevi, hemangiomas, and pilonidal dimples have also been seen. Foot deformities, inequality of the size of the feet or legs, and spina bifida are common skeletal abnormalities. Diastematomyelia may even occur in conjunction with a meningocele.

The presence of one or more of the above features in a child with gait disturbance should raise the suspicion of diastematomyelia. The diagnostic x-ray features, first described by Neuhauser, et al.,6 include a widening of the spinal canal at the level of the lesion, shown by an increase in the interpedicular distance, and in certain cases the visualization of the spur. In cases in which the spur is composed of bone, it may be seen as an oval-shaped density in the approximate center of the widened canal in the anteroposterior view. Usually the spur is not visible in lateral views. When in doubt, the diagnosis can be confirmed by myelography, since there is always a characteristic filling defect in the center of the oil column at the level of the lesion. Lumbar puncture alone does not help the diagnosis.

Surgical removal of the spur and dural sleeve and lysis of the adhesions may halt the progress of the neurological deficit, but marked improvement occurs in only a small number of cases.

The neurological manifestations resulting from cord traction, appearing as they do during growth of the individual, have led to the recognition of the abnormality in infancy or childhood in the great majority of clinical cases described to date.

Five adult cases, other than the two presented in this report, have been reported and are summarized in Table 1. Cases 1 through 4 we believe to be true examples of the onset of symptoms and signs in adult life without any previous suggestion during childhood. It is important to note the low level of the congenital abnormality in each of these four cases.

It is interesting to speculate why symptoms were delayed in these cases of diastematomyelia discovered in adults. In the case of Herren and Edwards8 and in that of the 39-year-old man of Seaman and Schwartz9 as well as in Bertrand's case, there clearly was evidence of neurological malfunction dating back to well before medical attention was sought, perhaps to childhood. It might be supposed that these three cases had either been overlooked, not brought to medical attention, or misdiagnosed in earlier years. However, in the second case of Seaman and Schwartz,9 Hamby's case,2 and our two cases, the patients were asymptomatic until the relatively rapid onset of neurological difficulty during adult life. In these latter
four cases, the spur was located in a low lumbar position so that it either divided the cauda equina or divided the conus medullaris at such a low level that the cord did not re-unite below the spur. Although traction on the cord can occur in such cases via adhesions from the spur to the meninges of the cord or to the cauda equina, this relatively small amount of traction might be sufficient to elongate the spinal cord during growth, although not sufficient to produce early symptoms. The delicate balance attained in such an individual might easily be upset in later years by a stressing event of some sort, such as the blow to the back in Hamby’s case. In our first case, the stressing event seemed to be the formation of a bony ridge at L1–2 secondary to spondylosis over which the cord was sufficiently stretched to de-compensate the delicate neurological balance the patient had maintained for years. In our second case, the precipitating event may have been intragenic at the time of the saddle block.

Summary

Diastematomyelia is rarely diagnosed during adult life. We have summarized five previously-reported adult cases and added two new ones. We have emphasized the low level of the lesions and the fact that the spinal cord did not re-unite below the spur as factors which we believe caused the delay in onset of symptoms. The final acute onset seemed to follow a stressing event which in some unexplained way dec compensated a delicate neurological balance.

References

1. Bertrand, G. Personal communication.
7. Ollivier. Trie de des maladies de la moelle epiniere. Cited by Perret (see ref. 8).