Adult Diastematomyelia

Neville A. Russell, Brien G. Benoit, Andrew J. Joaquin and Nayef Al Fayez

Abstract: With modern imaging techniques, various types of spinal dysraphism are being diagnosed in adults with increasing frequency. We report a 42-year-old woman with diastematomyelia whose symptoms characteristically were precipated by trauma. She exhibited other typical features such as a posterior midline hair patch and vetebral malformations. Metrizamide myelography with computerized tomography showed a bony septum at T11 with a cleft at T9-T11 separating two dural tubes each of which contained a single spinal hemicord. The septum was excised with complete relief of symptoms.

Résumé: Diastématomyélie adulte. Depuis l'avènement des techniques modernes d'imagerie, différents types de dysraphie médullaire sont diagnostiqués de plus enplus souvent chez l'aulte. Nous rapportons le cas d'une femme de 42 ans, porteuse d'une diastématomyélie, dont les symptômes ont été précipités typiquement par un traumatisme. Elle présentait d'autres manifestations caracteeristiques, somme une touffe de poils en position médiane postérieure et des malformations vertébrales. Une myélographie au meetrizamide avec tomographie assistée par ordinateur a montree un septum osseux à D11 avec une fente à D9-D11 séparant deux tubes duraux dont chacun contenait une demi-moelle épinière. Le sptum a été excisé entraînant une disparition complète des symptômes.

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Diastematomyelia (DM) is a relatively uncommon form of spinal dysraphism, in which the spinal cord or conus is split in the sagittal plane, over a number of segments, into two separate parts or hemicords. The cleft usually contains an osseocartilagenous septum that projects backwards from the posterior surface of the vetebral body to create two distinct arachnoidal-dural sheaths, each containing a separate hemicord.[1] In another type of DM, both hemicords are enclosed within a single meningeal sheath without a septum, but usually with fibrous bands attaching the neural elements to the posterior dura or to the laminae.[2] Theories of the embryogenesis of DM are usually based upon a primary abnormality or a secondary defect in the neural ectoderm which causes it to split and results in the formation of two hemicords. The gap between them becomes filled with mesodermal tissue which creates the septum.[1] DM is associated with a well-recognized syndrome or lower spinal cord or conus medullaris dysfunction, the features of which include weakness and atrophy of the distal lower limb musculature, impairment of sphincter function and sacral sensory loss.[3] Associated cutaneous abnormalities, especially a mid-line hair patch and vertebral malformations are common.[1,3] Although adult cases have been reported[4,5] DM has been considered to be almost exclusively a condition of childhood. Now, with the modern imaging techniques, it is being diagnosed in older patients with increasing frequency.[6-8]

In this report we describe the case of a 41-year-old woman with DM who became symptomatic after an accident. Computerized tomographic (CT) scanning after metrizamide myelography provided a precise diagnosis and surgery brought about prompt relief. We review the diagnostic features of adult DM and discuss the mechanisms that underlie the development of symptoms in this condition.

Case Report

History

Two years previously this 41-year-old woman sustained an extension injury to her neck and upper torso in a motor vehicle accident. This resulted in neck and right shoulder pain, and a peculiar dull backache localized to the lower thoracic spine with radiation around her right side in a hemi-girdle fashion. The neck and shoulder pain abated but the backache worsened and was aggravated by all forms of exertion. It was not relieved by rest and interfered with her sleep. She had no complaints of bowel or bladder dysfunction.

Examination

There was a large hairy patch overlying the lower thoracic spine. Scoliosis, convex to the left, extended from T6 to T12. Pin prick sensation was diminished in a band-like distribution over the right side from approximately T6 to T12. There were no other neurological or musculoskeletal abnormalities

Radiological Investigation

Conventional radiographs of the thoracic spine confirmed the scoliosis. In addition, they demonstrated a bony septum at T11, spina bifida of T11-T12, and widening of the interpedicular distance from T9 to L1. A computerized tomographic (CT) myelogram demonstrated splitting of the spinal cord by the bony septum from T9 to T11 (Figure 1). The conus was normally positioned and the filium terminale not thickened.

Operative Findings

The laminar defect to T11 was identified and a 3-level laminectomy from T10 to T12 was carried out. Two dural tubes separated by the mid-line bony spur were identified. The spur was partially removed using a high speed drill. The dura was then opened in a longitudinal direction leaving a small ellipse attached to the remnant of the bone spur

From the Division of Neurosurgery, Ottawa Civic Hospital, the University of Ottawa, Ottawa, Ontario, Canada and the King Fahad National Guard Hospital, Riyadh, Saudi Arabia.

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Reprint requests to: Dr. Brien G. Benoit, 606 - 1081 Carling Avenue, Ottawa, Ontario, Canada K1Y 4G2

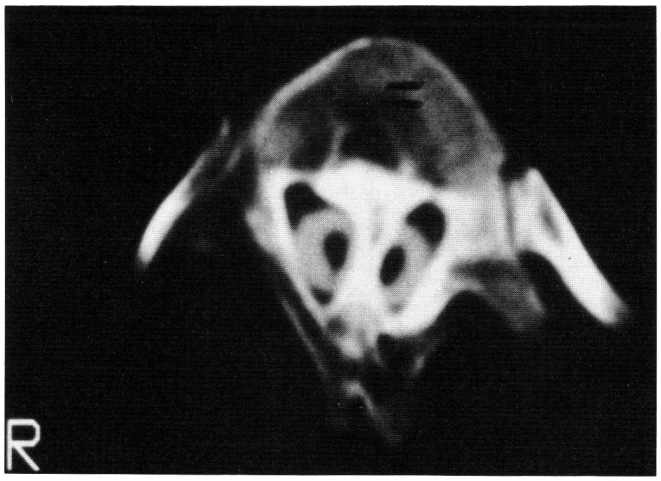

Figure 1: *Metrizamide (CT) myelogram showing two distinct spinal hemicords separated by a bony septum.*

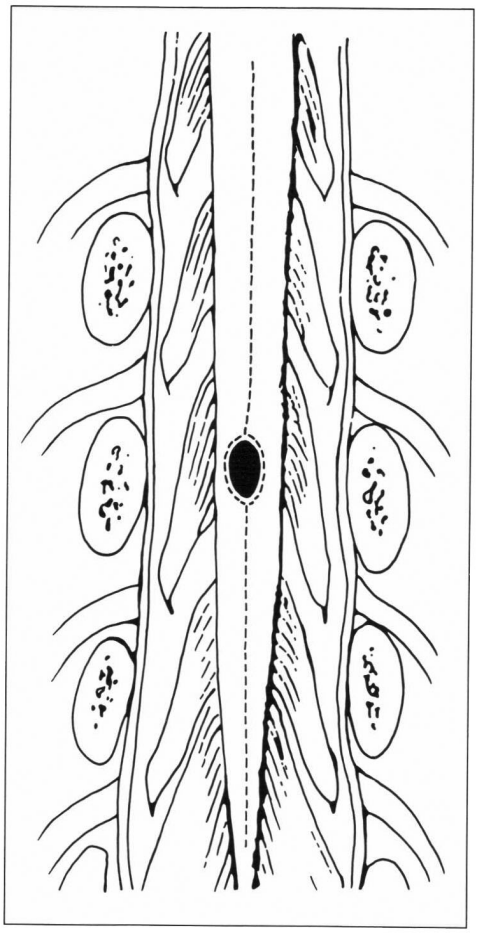

Figure 2: *Sketch depicting the operative site after laminectomy. The broken line represents the dural incision which exposes each hemicord and the normal spinal cord cephalad and caudad to the bone spur.*

(Figure 2). This gave access to both hemicords as well as to the normal spinal cord cephalad and caudad to the cleft. the neural elements were found to be separated by the spur and attached to it by fine adhesive bands. With the aid of the operating microscope these adhesions were lysed and the remainder of the bone spur removed. The anterior dura was densely adherent to the vertebral body and to the periphery of the base of the bone spur. The posterior dura were closed in a "water tight" fashion, to recreate a single dural tube.

Postoperative Course

The patient was discharged home on the 8th postoperative day. Her pain abated and the area of numbness on her right trunk gradually disappeared. Seven years post surgery she is asymptomatic.

Discussion

Pathophysiology

The theory explaining symptom development in DM, on the basis of differential growth between the transfixed spinal cord and the vertebral column has mainly been discarded.[9] Instead it is believed that neural damage results from sustained traction by the bone spur, fibrous bands and adhesions, or by other abnormalities, such as tight filum terminale. This may cause ischemic injury or prejudice the blood supply to the cord, making it vulnerable to further insult. Thus traction, plus the cumulative effect of repetitive trauma associated with everyday spinal movement, may tip a delicate balance, resulting in new or progressive neurological deficits.[9,10] The age at which symptoms appear may determine by the degree of traction and the severity of stretching of the the neural tissue.[10] With severe traction, symptoms appear in infancy or childhood and are related to growth spurts. Lesser degrees of traction may result in minor or stable childhood deficits, or remain subclinical, until adult life, when influenced by other factors such as trauma or spondylosis. Our patient was asymptomatic until an injury, and other authors have identified definite events or incidents that precipitated the development of symptoms in adult DM.[6,7]

Diagnosis

Although the symptoms in our patient and in those reported by others were "adult onset", there are often abnormalities from childhood that should call attention to the diagnosis.[5] These are mainly non-progressive, often minor, neurological or musculoskeletal anomalies or various cutaneous stigmata. In fact congenital anomalies of the vertebral column are so common in DM that when seen on plain radiographs in patients with appropriate clinical findings, they may be pathogonomic. Metrizamide myelography with CT scanning seems to be the most accurate radiologic method of evaluating DM.[6] It provides a precise display of the intrathecal structures; not only can the septum be identified, but density measurements can determine whether it is made up of bone or fibrous material. It also provides accurate information as to the relationship of the neural tissues and meninges to the cleft, and can identify associated lesions such as cysts or lipomas. It will also determine the position of the conus medullaris, which was normal in our patient but is often found to be low-lying in this condition. The role of magnetic resonance imaging (MRI) in adult DM has not been clarified. In one case when used as an adjunct to conventional investigations, it displayed the two hemicords, though with no greater precision than the metrizamide CT study.[7] In another, a 3-dimensional technique was necessary because scoliosis made the sagittal images unreliable and evaluation in the coronal plane was limited by the spinal curvatures.[11]

Treatment

Surgical decompression of the neural structures, removal of the bone spur, and lysing adhesions, is indicated in patients with DM who have new or progressive neurologic deficits.[6,8,12] The majority of cases will benefit from surgery, though a fixed deficit, such as a small hypoplastic limb, will not improve. The indications for surgery in asymptomatic or stable patients remain controversial. There are proponents of prophylactic surgery to prevent further neurologic deterioration[9] and others who resort to operation only if deterioration occurs.[12]

A greater awareness of the minor stigmata, as well as modern imaging techniques, is leading to the diagnosis of various types of occult spinal dysraphism in increasing numbers of older patients. The operating microscope and intraoperative spinal cord monitoring have made spinal surgery safer than ever before. Given this information, and knowing that minor trauma may precipitate devastating neurologic disability, we suggest that in adult patients with DM it may be prudent to err on the side of aggressive treatment and recommend appropriate prophylactic surgery once the diagnosis has been established.

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