Diastematomyelia and intramedullary epidermoid spinal cord tumor combined with extradural teratoma in an adult

Case report

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Diastematomyelia is rarely diagnosed in the adult. Only a few such cases have been reported in the literature. A 26-year-old Mexican man with lumbar diastematomyelia is reported who also harbored a T-12 intramedullary epidermoid tumor and an extradural teratoma located in the dorsal aspect of the dural sac opposite L-4. These three rare coincidental lesions were removed at surgery. The patient’s condition improved.

KEY WORDS - developmental anomaly □9 diastematomyelia □9 epidermoid cyst □9 intramedullary tumor spinal cord □9 teratoma

In 1962, Matson stated that “The extent of variability among congenital defects of the spinal axis is virtually infinite.” Diastematomyelia is a well recognized, although unusual, clinical syndrome in children, but is rarely reported in the adult. The case presented here is unique in that an adult man with diastematomyelia was found also to have two independent rare lesions: an intramedullary epidermoid tumor of the spinal cord, and an extradural teratoma.

Case Report

This 26-year-old Mexican man entered ISSSTE Hospital, Monterrey, on June 24, 1982, complaining of increasing lumbar pain radiating to the right lower extremity, and progressive weakness and paresthesias of that limb. These symptoms had been present for 1 year. This man was the eldest of five children, and had been the product of a normal pregnancy and delivery. An abnormal patch of lumbar hair was noticed at birth. The developmental milestones were normal. At first, the low-back pain was related only to exertion but 1 year before admission it became sharp and progressively worse. Two weeks before admission he developed weakness and then spasticity of the right lower extremity with impairment of gait. At the same time, numbness of this limb extended to the groin. Sphincter function and sexual performance were normal. Pulmonary tuberculosis had been diagnosed in October, 1980, for which he was treated medically until discharged as clinically healthy several months before his present admission. One of his four brothers had a lumbar meningocele.

Examination. The patient weighed 58 kg. He had a moderate lumbar lordosis, and a patch of abnormal hair on the back, extending over the L3–S1 vertebral levels. A nodular oval cutaneous scar, 20 × 10 mm in area, was observed immediately above the hypertrichosis and slightly to the left of the midline. However, no skin dimples were found, nor were any abnormal masses palpated in this area. There was exquisite tenderness and spasm of the lumbar paraspinal muscles, and marked spasticity, with increased deep-tendon reflexes, clonus, and Babinski sign of the right lower extremity. A sensory level, although faintly apparent in the left side, was suggested up to T-12. Position sense and vibratory sense were diminished in both legs. The anal reflex was weak.

Chest x-ray films showed apical fibrosis of the left lung. Roentgenographic study of the entire spine and
Congenital spinal anomalies in an adult

FIG. 1. Computerized tomography scan of the spine at L-2. An ossified midline septum divides the spinal canal into two segments. Notice the anomalous fusion of the laminae and spinous process.

FIG. 2. Lumbar myelogram showing an island-like midline filling defect caused by the septum of bone. A complete block is seen immediately above the septum. Notice thinning of the related pedicles.

linear tomography of the lumbar spine demonstrated a calcified L-2 midline septum. In addition, the films showed spina bifida from L-3 to S-3, widening of the interpedicular distance distal to T-12, defective L-2 and L-3 laminae, and abnormal fusion of aberrant spinous processes of L-2 and L-3. Bilateral thinning of the T-12 to L-2 pedicles, and dorsal scalloping of the T-12 and L-1 vertebral bodies were particularly noticed. A horizontal computerized tomography (CT) scan at L-2 showed the spinal canal to be divided in two halves by a septum of bone (Fig. 1). Metrizamide (Amipaque) myelography showed a thin longitudinal filling defect at the midline at L-2; the dye column separated into two lateral canals. A complete block, suggestive of an intradural tumor, was also identified (Fig. 2). The cerebrospinal fluid was tinged yellow, and contained 2 leukocytes/cu mm and a protein level of 680 mg/dl.

Operation. An elliptical incision was made, and a fibrous band about 5 mm wide was exposed at the midline, leading from the cutaneous scar to a nodular tumor which was about the size of a golf ball. The tumor was loosely connected to the extradural tissue opposite L-4, thence extending cephalad to enter the spinal canal between the anomalous laminae of L-1 and L-2 and terminating blindly in the dura. The lengthy spina bifida accommodated a widened dural sac. Laminectomy from T-10 to L-2 was then carried out. A midline septum of bone joined the aberrant L-2 spinous process and laminae and extended dorsoventrally, separating the dural sac into two lateral portions. There was a fusiform dilatation of the dural sac immediately above the septum. No dural pulsation was seen below T-10. The extradural tumor and the attached fibrous band were removed.

The cord was exposed through a longitudinal dural incision encircling the spur of bone, and a 4-cm cleft, traversed by the septum, was observed at the midline. The cord reunited immediately below the cleft, with the conus ending opposite the L-3 level. Proximal to the cleft was a localized area of cord expansion, with the pia arachnoid showing scattered specks of cholesterol crystals and sebaceous material. The septum was dissected free extradurally, and amputated at its basal attachment in the ventral aspect of the spinal canal. Subsequently, using microsurgical techniques, an intramedullary tumor of the spinal cord extending from T-12 to L-1 was completely removed, except for tiny portions of adherent capsule. The tumor contained soft, waxy, greenish, sebaceous material. It did not contain hair. A diagram summarizing the operative findings is shown in Fig. 3.

Postoperative Course. The pain and the spasticity disappeared. The patient was soon ambulatory, with slight right foot drop. He observed that he had a more forceful urine stream by the time of hospital discharge on the 7th postoperative day.

Pathological Examination. Microscopic examination of the intramedullary lesion showed keratin debris; the wall was formed by a smooth lining of stratified epithelium supported by a layer of collagenous tissue, characteristic of an epidermoid cyst. The extradural tumor was classified as a mature teratoma, containing a mixture of respiratory, digestive, mucinous, fatty, cartilaginous, and bony tissues. No sex
chromatin studies were carried out on this lesion. The attached band was fibrous, with no lumen or epithelial lining, so it did not qualify as a dermal sinus.

**Discussion**

Diastematomyelia has been defined as a form of spinal dysraphism in which the spinal cord is divided longitudinally into two discrete portions, usually by a septum of bone, cartilage, or fiber. Concomitant duplication of the investing dural sac may or may not occur: the septum is usually absent when the dural sac is single. Diplomyelia, on the other hand, is a true duplication of the spinal cord. The septum is thought to result from an incomplete endeavor of the mesodermal vertebral centers to form pedicles, or from aberrant mesodermal cells protruding in the ventral aspect of the neural plate instead of developing around it. The embryological, clinical, and radiographic features of diastematomyelia, as well as the surgical indications and technique, are amply discussed in the literature. Positive contrast myelography and particularly horizontal CT scans will clearly demonstrate the lesion, as in the current case.

Table 1 summarizes 12 cases of diastematomyelia in the adult collected from the literature, including the current case. Ages ranged from 20 to 59 years, with a median age of 33 years 5 months. The female: male ratio was proportionally similar to the ratio for children; there were eight females and three males.

### TABLE 1

**Clinical data in 12 adults with diastematomyelia**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors, Year</th>
<th>Age (yrs), Sex</th>
<th>Septum Level</th>
<th>Onset of Symptoms</th>
<th>Observations</th>
<th>Local Cutaneous Changes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Hamby, 1936</td>
<td>20, F</td>
<td>L-3, L-4</td>
<td>delayed</td>
<td>blow to the back</td>
<td>hypertrichosis</td>
</tr>
<tr>
<td>2</td>
<td>Herren &amp; Edwards, 1940</td>
<td>23, F</td>
<td>T-11</td>
<td>childhood</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Seaman &amp; Schwartz, 1958</td>
<td>39, M</td>
<td>T-5</td>
<td>childhood</td>
<td>kyphoscoliosis</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Freeman, 1961</td>
<td>20, F</td>
<td>L-2, L-3</td>
<td>delayed</td>
<td>small it calf</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td></td>
<td>43, F</td>
<td>L-3</td>
<td>delayed</td>
<td>no myelography</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>English &amp; Maltby, 1967</td>
<td>48, M</td>
<td>L-5</td>
<td>delayed</td>
<td>50-mile truck ride, L-1-2</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td></td>
<td>32, F</td>
<td>L-3</td>
<td>delayed</td>
<td>ridge</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Bertrand, 1967</td>
<td>20, ?</td>
<td>T-11</td>
<td>childhood</td>
<td>saddle block, no operation</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Dale, 1969</td>
<td>59, F</td>
<td>T-5, T-6</td>
<td>childhood</td>
<td>low cerebellar tonsils, arachnoidal cyst, scoliosis</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Sheptak, 1978</td>
<td>36, F</td>
<td>no septum, sacral cord cleft</td>
<td>childhood</td>
<td>automobile accident, single dural sac, fibrous band</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>Garcia, et al., 1980</td>
<td>37, F</td>
<td>L-2, L-3</td>
<td>delayed</td>
<td>rotoscoliosis, herniated disc at L-4</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>Garza-Mercado, 1983</td>
<td>26, M</td>
<td>L-2</td>
<td>delayed</td>
<td>T-12 intramedullary epidermoid cyst, L-4 extradural teratoma, fibrous band</td>
<td></td>
</tr>
</tbody>
</table>

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in the group (in one case the sex was not specified). The septa were located at T-5 in two patients, at T-11 in two more, and in the lumbar area in seven; one patient with a sacral cord cleft had no septum.\textsuperscript{34} Seven patients had delayed symptomatology. The remaining five had symptoms arising at childhood, including urinary abnormalities, vertebral anomalies, or limb deformities. Local cutaneous changes were present in six patients, in most cases hypertrichosis. Trauma was an apparent precipitating factor in four patients: a blow to the back in one patient (Case 1),\textsuperscript{13} a 50-mile ride in a pick-up truck in another (Case 6),\textsuperscript{6} a lumbar puncture in another (Case 7),\textsuperscript{6} and an automobile accident with acute flexion injury in the fourth (Case 10).\textsuperscript{24} In one patient (Case 6) the diastematomyelia was associated with a proximal ridge of bone,\textsuperscript{6} and with a distal herniated disc in another (Case 11).\textsuperscript{9} One patient (Case 10) had a meningocele repaired in infancy, and years later surgery revealed a subcutaneous fibrous band containing nerve-like elements penetrating the dural canal to terminate in the divided cord.\textsuperscript{34} This is the only patient without a septum who had a single dural sac. Eleven patients were submitted to surgery, one (Case 5) without preoperative myelography.\textsuperscript{8} One patient (Case 6) deteriorated postoperatively, and the rest improved. One patient (Case 9) had an associated arachnoidal cyst, and lowered cerbellar tonsils.\textsuperscript{5}

Epidermoid cysts and teratomas are rare intracranial tumors (0.2\% to 1\%, and 0.5\% respectively).\textsuperscript{30} They are even more unusual among intraspinal lesions.\textsuperscript{11,21,35} Sloof, \textit{et al.},\textsuperscript{35} found them to constitute nearly 1.4\% of intraspinal growths, including dermoid cysts. Epidermoid cysts result from imperfect closure of the medullary folds, or from retained heterotopic ectodermal implants, between the 3rd and the 5th gestational weeks.\textsuperscript{7,30,33} They may have an iatrogenic origin, following single or multiple lumbar punctures,\textsuperscript{34} meningocele repairs,\textsuperscript{10} or inadequate suturing of scalp lacerations,\textsuperscript{36} and they have been produced experimentally in immature albino rats.\textsuperscript{37} A dermal sinus ending blindly in the dura has rarely been reported in association with an intramedullary epidermoid cyst in an adult.\textsuperscript{31} The origin of the teratomas remains controversial. They may result from faulty germ-cell migration from the endoderm to the primitive gonads;\textsuperscript{10} nuclear sex determinations also suggest this etiology.\textsuperscript{28} The pluripotential heterotopic embryonic tissue grows haphazardly after escaping the influence of the primary organizer during early development.\textsuperscript{29}

Epidermoid tumors concentrate in the thoracic spine and more commonly affect young adults.\textsuperscript{29} About one-third to one-half of intraspinal epidermoid tumors are intramedullary. Of the 90 intraspinal epidermoid cysts compiled in 1962 by Manno, \textit{et al.},\textsuperscript{21} 51 were congenital, 18 of which were intramedullary, and one was associated with diastematomyelia (Case 42). This patient was an 8-year-old girl originally reported by Black and German.\textsuperscript{2} Intraspinal teratomas are much rarer lesions. Up to 1978 only 46 cases had been reported,\textsuperscript{29} and by 1980 only nine intramedullary teratomas had been described.\textsuperscript{10} Occasionally a teratoma may be associated also with a dermal sinus.\textsuperscript{32}

As shown by this group of adult patients, diastematomyelia does not necessarily preclude a long asymptomatic life. The harmonic interdependence between the cord and the traversing septum, however, may be disrupted by external or internal environmental conditions, and symptoms may then appear on account of local cord injury.\textsuperscript{5,12,34} The patient reported here had a wide spectrum of developmental anomalies (Fig. 3) representing part of the so-called “dysraphic state of Lichtenstein;”\textsuperscript{19} he had diastematomyelia with a septum of bone, spina bifida, expanding congenital tumors, a fibrous band, and integumentary lesions. All these anomalies must have resulted from a single embryological alteration early in life. What Matson wrote 20 years ago\textsuperscript{22} remains as valid now as it was then.

\section*{Addendum}

Since the submission of this manuscript, a new case of adult diastematomyelia has been reported by Maroun FB, Jacob JC, Mangan MA, \textit{et al.}: Adult diastematomyelia: a complex dysraphic state. \textit{Surg Neurol} 18:289-294, 1982. The patient was a 29-year-old female nursing assistant with an L3-4 diastematomyelia defect seen at myelography, associated with a lumbar megasac, fibrous bands, and high termination of the theca, for which the patient was operated on. She required a second operation 16 months later, at which time a large coccygeal cyst tethered to a lipoma containing filum terminale and aberrant extradural nerve roots were observed.

\section*{Acknowledgment}

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\section*{References}

1. Bertrand G: Cited in Reference 6
2. Black SPW, German WJ: Four congenital tumors found at operation within the vertebral canal. With observations on their incidences. \textit{J Neurosurg} 7:49-61, 1950
7. Fleming JFR, Botterell EH: Cranial dermoid and epi-

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