DIASTEMATOMYELIA ASSOCIATED WITH DORSAL KYPHOSIS PRODUCING PARAPLEGIA

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In the course of the last two decades scattered reports have appeared in the literature regarding the two separate conditions of diastematomyelia and kyphoscoliosis with paraplegia. In all, 62 cases of diastematomyelia were found in the literature.2-4,6-13 A review of the reports of kyphoscoliosis with marked neurological symptoms revealed a total of 43 well-proven cases.5,9 As far as is known this is the first case to be reported in which the two conditions existed in the same patient. It is presented as an unusual entity and it may suggest a possible reason for the development of neurological symptoms in some cases of kyphoscoliosis. The co-existence of two lesions in the bony spinal column and the presence of a single neurological abnormality are of interest in the problem of the etiology of diastematomyelia.

CASE REPORT

#75293 A.U.H. M.M., aged 12, was referred with the complaint of gradual paresis of both lower legs of 7 months’ duration. With the exception of kyphosis and possibly scoliosis, which had been noticed at birth, the patient’s early development was allegedly normal. He walked at the age of 18 months. There was a history of consanguinity at the level of his great-grandparents. There was no history or evidence of tuberculosis in his family.

The patient had been well and able to work on his father’s land until 7 months before admission. At that time his family noticed that his gait had become unsteady and that he was beginning to stumble. This progressed rather quickly, so that within 1 month he was unable to walk without support. Two months after the onset of his symptoms he was taken to a hospital and put on a hard bed for 2 months. This produced no improvement. He was then put in a plaster cast to include all of both legs and back for 3 months. When the cast was removed, the patient found that he could not move his legs at all and that his legs had become numb. He was then brought to the American University Hospital in Beirut.

Examination. He was a well-built, well-nourished boy in no distress. At T6 there was a marked kyphosis with slight left scoliosis (Fig. 1). Over the spine at the level of L1 to L3 there was an area of pigmentation and a prominent tuft of hair (Fig. 2).

Neurological examination revealed a level of hypaesthesia at T10, and a loss of light touch sensation at T6. Deep touch sensation was slightly decreased from T6 to L1, moderately decreased from L1 to L3 and was absent below L4. Position and vibratory sensations were lost below T10. There was spastic paralysis of both lower legs. Abdominal reflexes were absent. Knee and ankle jerks were markedly increased with prolonged clonus at both ankles. Babinski and Gordon reflexes were positive. There was no abnormality of sphincter control. There was no difference in the size or length of the legs.

The clinical impression was compression of the cord at T6 with fixation of the cord, probably by a meningocele between L2 and L3. Roentgenograms revealed that there was a sharp angulation of the thoracic spine to about 90° at the level of T6 to T7. T6 was shown to be a posterior hemivertebra wedged in between the two contiguous vertebrae, but

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markedly shifted dorsally. In the upper dorsal spine there was furthermore a moderate left concave scoliosis. All the lumbar vertebrae showed an extensive schisis of their arches; L3 and L4 showed schisis of the body as well. No spinal process of the lumbar vertebrae was seen in the frontal views except for L2 which showed an oblong ring-like shadow which had the general appearance of the cross section of spinous process. The disc space between L2 and L3 was markedly narrowed.

A myelogram was performed which revealed a complete block of the column of dye at the level of the sharp angulation in the upper thoracic spine (Fig. 3). The spinal canal in the lumbar spine was uniformly wider than normal. At the level of L2 the spinal canal showed a constant central filling defect, about 2 by 1 cm., by which the spinal canal seemed to be split into two halves. At the center of this filling defect, the above-mentioned bony ring shadow was seen.

1st Operation. On March 24, 1953 a laminectomy from L1 to L4 was performed, with disclosure of the defect shown in Fig. 4. This was removed. This spicule of bone was firmly attached to the under surface of the fused lamina of L2 and L3, which were markedly thickened and vascular. The attachment of the spicule of bone to the dorsal surface of L2 was thinner but still bony. The dura mater was separated from the spicule easily and when the bone was removed the cord was seen to move upward. The dura mater was not opened. No nerve roots could be seen coming off from the medial aspects of the two cords. The wound was closed without drainage.

Course. There was gradual loss of most of his spasticity in the following 10 days.

2nd Operation. On April 9, 1953 an exploratory laminectomy was performed at T5 to T7. There was moderate pressure on the cord at T5-T6 and T6-T7 by the laminae of T5 and T7. There were no pulsations seen in the cord. The cord did not seem to be stretched tightly over the gibbus at the time of the operation. However, because of the backward displacement of the T6 hemivertebra, the spinal canal was narrowed in an anteroposterior diameter and it