Regrowth of diastematomyelic bone spur after extradural resection

Case report

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Complete regrowth of a diastematomyelic bone septum producing progressive neurological deficits was found in a 15-year-old boy who had a previous extradural resection of a similar bone spur with the dural sleeve left untouched. This case and two similar cases in the literature documenting continued neurological deterioration after extradural removal of septum emphasize the need for resection of the dural sleeve along with the bone septum if adequate relief of tethering is expected. The capability of the septum to regenerate may be due to the persistence of residual mesenchymal cells associated with the embryogenesis of the midline septum.

KEY WORDS • diastematomyelia • bone septum • embryology • spinal cord • dura

DIASTEATOMYELIA is an uncommon congenital anomaly of the neural axis and spinal column characterized by a division of the spinal cord or cauda equina into two distinct lateral compartments by a septum of bone, cartilage, or fibrous tissue enclosed in a dural sleeve. When diastematomyelia is associated with neurological deficits, treatment consists of surgical excision of the midline septum to release the tethering effect on the neural tissue. We recently treated a 15-year-old boy who had progressive neurological deficits due to complete regeneration of a midline bone septum in the lumbar region 11 years after a previous extradural resection of a bone spur from the same dural cleft.

Case Report

This 15-year-old boy was referred to us because of paresthesia and recurrent ulcerations in the lower extremities. He was born with a shorter left leg, bilateral clubfoot deformity, and bilateral dislocation of the hips. He was kept in a body cast for 10 months for treatment of the hip dislocation, and did not crawl until the age of 18 months. He began walking unaided at 3 years of age with a significant limp. Examination at the age of 4 years revealed planovalgus deformity of the feet and diffuse atrophy of the left leg. He had patchy bilateral S-1 sensory loss, absence of perianal sensation, and 3/5 power in the dorsiflexion of the left ankle. Plain films of the lumbosacral spine showed congenital fusion of the L-2 and L-3 vertebral bodies and laminae, and midline fusion defects in the dorsal arches of the L-4 to S-3 vertebrae, associated with a widened lumbosacral canal. Pantopaque myelography demonstrated a diastematomyelic bone spur opposite the body of L-3, spanning the sagittal plane between the posterior aspect of the body and the abnormally thickened laminae, and dividing the sacral thecal sac and the conus into halves. The neurological abnormality was attributed to the tethering effect of the bone spur, and at operation the anomalous neural arches of L-2 and L-3 were completely removed. The midline bone spur, measuring 1.5 × 0.9 cm, was dissected free from the dural investment on both sides, and avulsed from its anterior attachment in one piece. The dura mater was not opened and the dural cleft was left untouched.

Between the ages of 5 and 12 years, the patient underwent three procedures to lengthen the right Achilles tendon to try without success to arrest the progressive worsening of his now calcaneovalgus foot deformity. A right femoral epiphysiodesis was also performed to correct the increasing leg-length discrepancy.

At the age of 13 years, the boy began to notice increasing difficulty in walking and climbing stairs, with a tendency to trip on the left side. The numbness in his
left foot was increasing and involved progressively more proximal areas. There had been multiple instances of recurrent, nonhealing ulcers over the dorsum of the left foot and toes, and a susceptibility to chronic osteomyelitis which ultimately required amputation of the second and fifth toes. He also described the curious phenomenon of ascending paresthesia and dysesthesia in the left lower leg whenever he twisted his trunk to the right.

Examination. The patient’s legs were diffusely atrophic and spindle-shaped, with marked tapering toward the feet, which were in moderately severe and fixed calcaneovalgus positions despite previous surgical corrections. The left foot had obvious trophic changes in the dermis and nail beds, and large chronic ulcers over the dorsum and the tips of the third and fourth toes: the second and fifth toes were missing. Deep-tendon reflexes and plantar response were absent in both lower extremities. Sensorimotor abnormalities had obviously worsened since childhood. The left foot at this time had no intrinsic evertor or dorsiflexor function, and had but a flicker of plantar flexion. The right foot had absent intrinsic and evertor functions, and 3/5 power in dorsiflexion and plantar flexion. Both feet were anesthetic in the L-5 and S-1 dermatomes, and the ascending dysesthesia and hyperpathia over L-3 and L-4 on the left could be elicited readily by rotation of the trunk to the right and by straight-leg raising. Metrizamide myelography showed a persistence of the mid-sagittal filling defect at L-3 (Fig. 1). With computerized tomography imaging, the midline septum appeared to be of the density of bone (Fig. 2). The two half-cords were contained in separate thecal sacs above the septum and reunited around the lower border of the septum. The lower end of the conus appeared to reach the S-1 level and was tightly bowed against the dorsal aspect of the dural tube.

Operation. The laminae of L-2 and L-3 that had been previously removed when the patient was 4 years old had completely regenerated into a fused, hypertrophic, knobby mass. After the dura mater was carefully separated from the inner surface of the laminae, a midline bone spur was found plunging from the inner aspect of the malformed laminae into the dural sheath which enveloped the previously resected bone spur. The regrown spur, measuring $1.4 \times 0.7$ cm (Fig. 3), was subperiosteally dissected from the dura and lifted away from its anterior fibrocartilaginous attachment. The
Diastematomyelic bone spur

dura mater was opened over the two sleeves around the midline cleft, as well as above and below the cleft to expose the conjoined conus, the two half-cords, the cauda equina, and the filum. The two half-cords were adherent to the sides of the fibrous (dural) septum and tightly hugged its lower border at the point of reunion. It was at this point where the stress of transfixation of the cord was most severe, and also where the neural and fibrous tissues were most intimately adherent and required the utmost care for separation by lysis of adhesions. The midline septum was completely excised to release the tethering. A slightly thickened filum was also severed to eliminate all possibility of residual tension at the tip of the conus. A linear closure of the dorsal dural defect had in fact converted the double sacs into a single dural tube.

Postoperative Course. The patient did well postoperatively and immediately felt subjective improvement in the discomfort over the skin of the left leg. The ascending dysesthesia over the left leg could no longer be elicited. Two months later, he also had significant improvement in plantar reflexes and dorsiflexion of both ankles.

Discussion

This report illustrates two points concerning diastematomyelia. The first involves the surgical treatment. The surgical technique for the resection of the midline bone septum was thoroughly described in 1950 by Matson, et al.,7 and in 1963 by Moes and Hendrick.9 The sequential steps and various technical options have also been superbly illustrated by Meacham.8 Most authorities believe that the tethered spinal cord is adequately released only if the dural sleeve surrounding the septum is also resected flush with the anterior wall of the spinal canal, and the two halves of the cord allowed to reside within a single-chambered thecal sac.1-4,7-9,10 Despite strong statements to this effect in the literature, it is our impression that some neurosurgeons still question the necessity of radical resection of the dural sleeve following septum removal.

We were able to find only two documented examples of continued clinical deterioration following extradural removal of the bone spicule without dural sleeve resection. Matson, et al.,7 reported a child who became paraplegic after extradural resection of a bone septum without dural opening. This child subsequently required reoperation for more radical resection. Moes and Hendrick9 described a case very similar to ours. Their 2-year-old patient underwent a laminectomy and extradural removal of an L-3 bone septum because of foot deformity and inability to walk. The dural sleeve was left untouched. He slowly improved postoperatively, but was readmitted 9 years later with progressive leg weakness and recurrent cellulitis and osteomyelitis of the toes secondary to trophic changes in the lower extremities. Myelography showed persistent splitting of the contrast column at the original site of the diastematomyelic septum and, at operation, a new bone spicule was found to have reformcd within the previously spared dural cleft. There was significant neurological recovery after resection of both the bone septum and the dural sleeve.

These and the present report leave no doubt about the importance of dural sleeve resection. Anyone who has attempted to sweep the last vestige of the dural septum from its inferior adherence to the crotch of the rejoined conus will appreciate the potentially deleterious effect exerted by the stiff inferior edge of the dural sleeve on the tightly apposed neural tissue, even after the bone septum has been removed. Also, the dural sleeve itself often stretches as much as 1 cm cephalad to the bone septum, and is always adherent to the medial surface of the half-cords. It is inconceivable that adequate release of the conus could be achieved without complete elimination of this dural island, and that the latter could be safely accomplished without a wide opening of the dura. Moreover, intradural exposure affords the added advantage of incising the fibrous adhesions often seen to anchor the lateral surfaces of the half-cords to the dural wall, and of seeking and eliminating other coexisting lesions, such as thickened filums,5,6 dermal sinus tracts, dermoids, and lipomas,5,10 that might contribute to the tethering process.

The second point emphasized by this case involves the complete regrowth of the bone septum following extradural removal, and is of some embryological interest. During the early stages of the development of the human embryo, a dorsal canal connects the yolk sac with the amniotic cavity through the primitive knot (Henson’s node) of the embryo. This primary neurenteric canal persists only for a short time, but during its presence it establishes a passage between the primitive enteric cavity (yolk sac) and the amniotic sac through dorsal midline structures. With cranial migration of its

FIG. 3. Photograph of the recurrent bone spur. Measure is in centimeters.
cells to form the neuroectoderm and mesodermal somites, the primitive knot comes to lie at the tip of the coccyx in the fully developed embryo, and the closed dorsal opening of the primary neurenteric canal sometimes persists as a sacral dimple.

According to Bremer, diastematomyelia results from the presence of accessory neurenteric canals through the developing neural plate (primitive streak) in the young embryo at points cranial to the primitive knot. With lateral growth of the embryo, the yolk sac is compressed bilaterally, and a dorsal herniation of the entodermal lining travels up the accessory neurenteric canal and splits into halves the notocord, neural groove, and the medially migrating mesenchymal components that will eventually form the vertebrae. Even a temporary persistence of this dorsal fistula will result in the formation of two hemivertebrae bodies which, with later disappearance of the fistula, may come together in the midline. The cuff of mesenchymal cells surrounding the fistula may then converge to form a midline spur that projects into the spinal canal between the two half-cords. Such a spur, made of bone, cartilage, or fibrous tissue, can extend dorsal to the cord and posterior to dura to fuse with the posterior neural arches, depending on the original dorsal extent of the entodermal fistula. In the spinal cord there is little attempt to close the cleft, and mesenchymal tissues surround each half to give each a meningeal covering. Residual polyvalent mesenchymal cells may persist in the young child within this midline tract, and after simple extradural removal of the bone septum, these cells may be reactivated to cause complete regeneration of a new bone spicule seen in our case as well as in the child reported by Moes and Hendrick.

Corroborating evidence can be found in a case reported by Gilmor and Batnitzky. Their 10-year-old patient was found to have regeneration of a fibrous septum bisecting the conus at the exact location where a previous bone spicule was totally excised with its dural sleeve in the newborn period. Although total excision of the dural sleeve should minimize the possibility of septum regeneration, the case of Gilmor and Batnitzky suggests that the potential for recurrence may still exist, regardless of the extent of the original resection. This implies that long-term follow-up review should always be set up for those who have had operations for diastematomyelia to assure that regrowth of a sagittal septum does not occur.

References

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