The Turkish Journal of Pediatrics 31:89-94, 1989

DIASTEMATOMYELIA: A REPORT OF TWO CASES*

*Sabahattin Çobanoğlu MD***

Key words: diastematomyelia, diplomyelia, intramedullary tumor, scoliosis, spinal dysraphism

Diastematomyelia is a variety of spinal dysraphism in which there is a congenital splitting of a part of the spinal cord or, rarely, of more than one part[1]. The condition is sometimes called diplomyelia because the medial aspect of the gray matter of the two hemicords often displays small ventral and dorsal horns, suggesting duplication. However, there is no double innervation of the lower extremities[1]. It should be stressed that diastematomyelia refers to the split neural tissue and not to the septum[2].

In many cases the two spinal cords are contained within a single dural tube without an intervening septum. However, when each spinal cord has its own dural sheath there always is a septum of bone, cartilage or fibrous tissue that can prevent ascent within the vertebral canal as the vertebral column grows in length or it may be the cause of pressure laterally on one or the other spinal cord[3]. Therefore, in diastematomyelia a low conus medullaris or tethered cord is almost always present[1]. The results of septal pressure are, therefore, more likely to be seen in childhood, although adult cases have been reported rarely in the literature[3,4].

Diastematomyelia is usually accompanied by a number of other malformations including cutaneous changes, skeletal–especially vertebral-anomalies, hydromyelia, meningocele or myelomeningocele, the Klippel-Feil syndrome, hydrocephalus and the Arnold-Chiari malformation[1,5].

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. Even though excision is not followed by the anatomical restitution of normal structure, it brings about clinical improvement[5,6].

In this paper, we present two cases of a rather rare congenital anomaly, diastematomyelia, with hope that they will be of interest to pediatricians since they might encounter this type of case in their practice.

---

* From the Department of Neurosurgery, Trakya University Faculty of Medicine, Edirne.

** Assistant Professor of Neurosurgery, Trakya University Faculty of Medicine.

Case Reports

Case 1

A five-year-old girl presenting with right Sprengel's shoulder, mild congenital high thoracic scoliosis and diastematomyelia at the 4th lumbar vertebra (LV4) was referred to the Department of Surgical B Neurology from the Orthopedic Department of Edinburgh University. Neurological examination revealed that the patient's left leg was shorter than the right and the right foot smaller than the left. There was no abnormality of the cranial nerves or upper limbs. Systemic findings, power, tone, sensation, coordination and gait were all normal. Anal sphincter tone was also normal, as was sensation. Direct plain x-rays confirmed a right thoracic scoliosis which extended from LV4 into the sacrum with a possible diastematomyelic spur at LV4. Accordingly, the patient underwent a total myelography which illustrated an ovoid filling defect at the LV4 level with no other abnormality (Fig. 1).

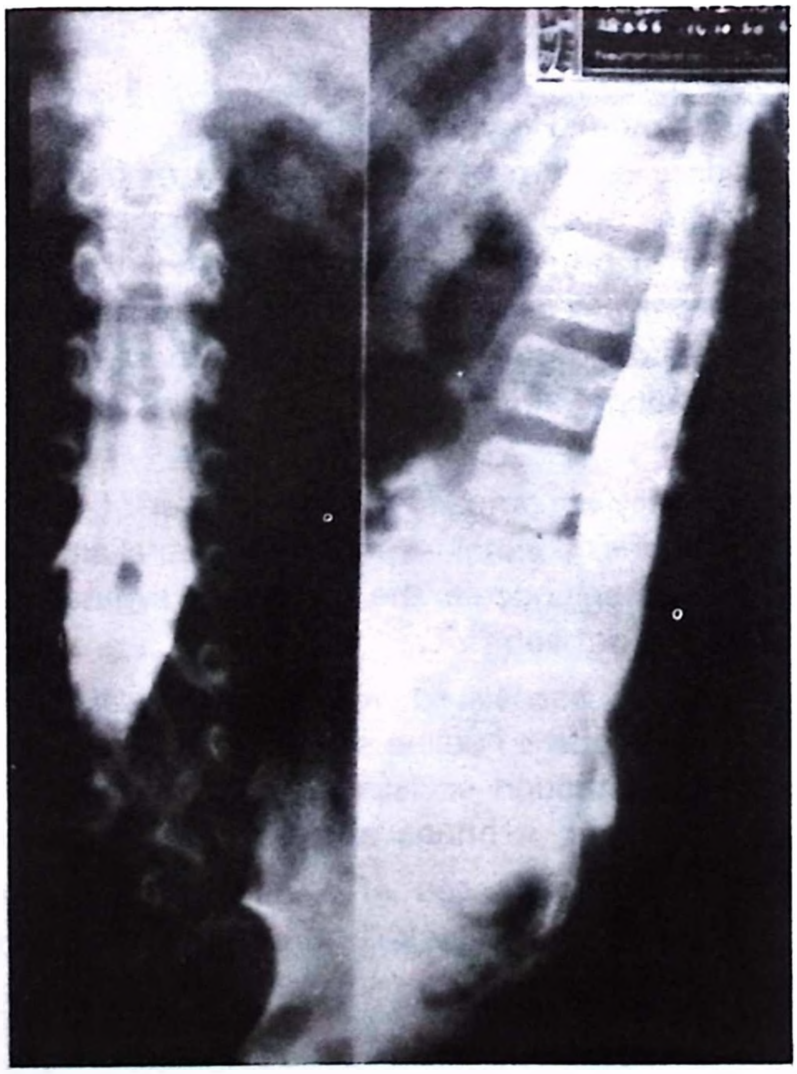

Fig. 1: Lumbar radiculography (Case 1) antero-posterior (AP) lateral views show an ovoid filling defect at the LV4 level.

Laminectomies were performed from LV3 to LV5. At the level of the 4th lumbar vertebra a bony spur was defined and two dural tubes surrounding it were exposed, which immediately divided above and reunited below the spur. After trimming away some of the posterior extremity of the spur, the dura mater was opened at the midline vertically above and below the spur. These midline incisions were then joined together. The bony spur was excised after which the dura mater was sutured posteriorly and anteriorly. The bony spur appeared to be keel-shaped and lightly adherent to the dorsal aspect of the vertebral body having a small rudimentary articular surface. During the operation it was also noticed that similar to the dura, a low cord had split just above the spur, encircled it and reunited immediately below, the left neural tube being the smaller of the two. Roots left the lateral aspect of these two halves of the cord. However, no roots left medially.

Fig. 2: Lumbar radiculography (Case 2). AP view shows an ovoid filling defect at the IV4 level.

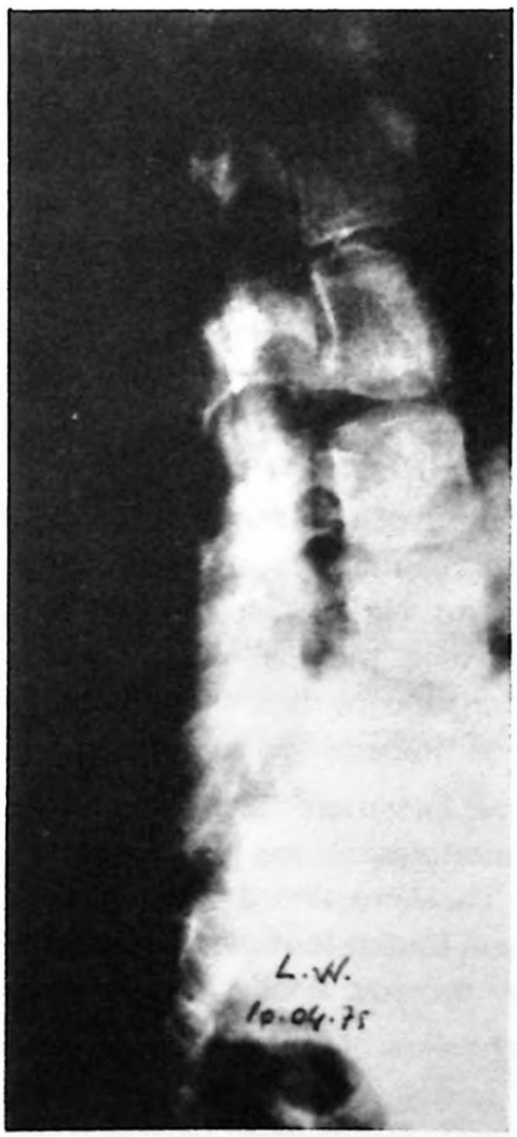

Fig. 3: Lateral view of lumbar radiculography (Case 2).

Case 2

A six-year-old girl was referred to the same neurosurgery department by an orthopedist because of congenital skeletal anomalies. Physical examination revealed that she had a limp and an elevated left shoulder (Sprengel's shoulder). Her thoracic spine was scoliotic and there was a hairy patch of skin over the apex of the scoliosis. Bilateral supinator jerks, ankle jerks, and a right knee jerk were all absent. Power, tone, sensation, and coordination in all limbs were normal.

Cervical spine x-rays showed a hypoplastic odontoid process and fusion of CV2-3 vertebrae. Thoracic spine x-rays revealed severe left thoracic scoliosis extending from CV7 to TV12, measured at 72°. Additionally, there were multiple congenital anomalies with a unilateral unsegmented bar on the concavity of the scoliosis extending from TV1 to TV9. Lumbosacral X-rays showed a possible diastematomyelic bony spur at the LV4 level. A total myelography confirmed that there was an ovoid central defect (Figs. 2, 3). The patient was operated on and laminectomies were performed from the LV4 to SV1 levels. As in the first case, the same surgical procedure was applied to excise the bony spur. In this case, however, there was additional pathology regarding the bony spur and low conus medullaris, that is, spina bifida from the LV4 to SV1. An epidermoid tumor was attached to the right halves of the conus medullaris. The bony spur and tumor were carefully removed. The patient's post-operative condition was satisfactory. At a later date, she underwent orthopedic intervention for the sciolosis.

Discussion

Diastematomyelia is more common in females than in males. Although the average presentation age is 4.3 years, there have been reports of adult cases in the literature. No specific genetic disease was present in our two cases, which coincides with the data in the literature[1,2,4,7-9]. Since diastematomyelia was reported in two female sibships by Kapsalakis[7] in 1964, it is possible that his anomaly is transmitted in an autosomal recessive mode of inheritance.

James and Lassman[3] have reported that the most common presenting features of diastematomyelia are lumbrosacral hypertrichosis, foot deformities and a limp. Our first case presented with right Sprengel's shoulder, high thoracic scoliosis, and leg and foot deformities while our second case presented with left Sprengel's shoulder, thoracic scoliosis and hypertrichosis over the scoliosis.

The mechanism of neural damage whereby the septum and its dural attachments lead to damage within the split cord have been much discussed but poorly understood[2]. James and Lassman[3] found extrinsic bands in eight of eleven cases who were operated on for diastematomyelia. Microscopic examination of the bands revealed aberrant and atrophied nerve roots which were considered to be a

cause of neurological damage because of traction of the spinal cord, conus medullaris, filum terminale or cauda equina[2,3]. In Case 1 no extrinsic pathology was seen during the operation apart from a diastematomyelic bony spur, however, in Case 2, there was also an epidermoid tumor. It is not known how epidermoid tumors or teratomas are related to spinal cord malformations[5,10].

In one series the level of split was frequently seen at LV2 while in other reported series it occurred at LV4. This data is similar to the cases reported by us, although other levels of split have been reported rarely in the literature[2,8,9].

Both of our cases were operated on in order to remove the diastematomyelic bony spur, believed to have a considerable tethering effect on the neural system, thereby preventing the progression of neurological damage. The operation resulted in an entirely free and mobile segment of the low spinal cord, yielding a good result.

Summary

Two cases of diastematomyelia are presented and the related literature is reviewed. Diastematomyelia, a complete or incomplete sagittal division of the neural axis into two halves, is usually accompanied by a number of other malformations. One of the cases in this paper appeared to have an epidermoid tumor which is rarely associated with this seldom seen congenital anomaly. Diastematomyelia, which becomes symptomatic in childhood, requires early surgical intervention to relieve the tethering effect on the cord by removal of the splitting pathology which is thought to be the main cause of neurological deterioration in the ensuing years.

Acknowledgement

I wish to thank Prof. J. Douglas Miller, MD, Chairman of the Department of Clinical Neurosciences, Edinburgh University, United Kingdom, and Dr. J. Shaw, MD who taught me the technique involved in this type of neurosurgery and kindly permitted the publication of the Department's cases.

REFERENCES

1. Gunthkelch AN. Diastematomyelia. In Wilkins RH, Rengachary SS (eds). *Neurosurgery*. New York: McGraw, 1985, pp 2058-2060.
2. Kennedy PR. New data on diastematomyelia. *J Neurosurg* **51**:355, 1979.
3. James CC, Lassman LP. Diastematomyelia. A critical survey of 24 cases submitted to laminectomy. *Arch Dis Child* **39**:125, 1964.
4. English WJ, Maltby GL. Diastematomyelia in adults. *J Neurosurg* **27**:260, 1967.
5. Ugarte N, Gonzalez-Crussi F, Sotelo-Avila C. Diastematomyelia associated with teratomas. Report of two cases. *J Neurosurg* **53**:720, 1980.

6. Pang D, Parrish RG. Regrowth of diastematomyelic bone spur after extradural resection. Case report. *J Neurosurg* 59:887, 1983.
7. Kapsalakis Z. Diastematomyelia in two sisters. *J Neurosurg* 21:66, 1964.
8. Beyerl BD, Ojemann RG, Davis KR, et al. Cervical diastematomyelia presenting in adulthood. Case report. *J Neurosurg* 62:449, 1985.
9. Kuchner EF, Anand AK, Kaufman BM. Cervical diastematomyelia: a case report with operative management. *Neurosurgery* 16:538, 1985.
10. Garza-Mercado R. Diastematomyelia and intermedullary epidermoid spinal cord tumor combined with extradural teratoma in an adult. Case report. *J Neurosurg* 58:954, 1983.
11. Chehrazi B, Haldeman S. Adult onset of tethered spinal cord syndrome due to fibrous diastematomyelia: case report. *Neurosurgery* 16:681, 1985.