ELSEVIER

The Spine Journal 7 (2007) 622–626

THE SPINE JOURNAL

Adult presentation of spinal dysraphism and tandem diastematomyelia

Paul Porensky, BA*, Kenji Muro, MD, Aruna Ganju, MD

Department of Neurological Surgery, Feinberg School of Medicine, Northwestern University, 244 E. Pearson St. #1705, Chicago, Illinois 60611

Received 12 April 2006; accepted 9 August 2006

Abstract

BACKGROUND CONTEXT: Diastematomyelia is a split-cord malformation often accompanied by other cord or column anomalies.

PURPOSE: To report on an adult patient with diastematomyelia and discuss the embryological basis and related developmental sequelae of this split-cord malformation.

STUDY DESIGN: Case report.

METHODS: A summary of the management of a 54-year-old woman with recent clinical symptomatology related to an undiagnosed split-cord malformation is presented with accompanying literature review.

RESULTS: A rare adult presentation of diastematomyelia with accompanying intradural extramedullary epidermoid tumor was repaired with resection of the soft-tissue mass and excision of the fibro-osseous septum.

CONCLUSION: Initial presentation of diastematomyelia is rarely seen in adults; accompanying pathology includes scoliosis, tethered cord, and intradural tumors. Effective treatment involves identification of the primary pathology.

Keywords: Diastematomyelia; Type I split-cord malformation; Scoliosis; Tethered cord; Adult presentation; Intradural extramedullary epidermoid tumor

Introduction

Diastematomyelia is a spinal dysraphism characterized by the presence of an osseous, cartilaginous, or fibrous septum that splits the spinal cord into two distinct hemicords. These split-cord malformations (SCMs) can be further classified as to whether each hemicord is contained within its own dural sac (type I) as opposed to containment of both hemicords within a single common dural sheath (type II) [1].

SCMs are often accompanied by other cord or column anomalies, including tethered cord, dermoid or epidermoid tumors, syringomyelia, and scoliosis. The presence of these occult intraspinal lesions is suggested by cutaneous stigmata such as hypertrichosis or dimpling [1,2]. Both diastematomyelia and these other associated abnormalities are rare within the general population; it is even more uncommon for initial clinical presentation to occur in adulthood [3–5]. We report on the initial presentation of a tandem type I SCM, tethered cord, and intradural extramedullary epidermoid tumor in an adult.

HPI

A 54-year-old woman presented with a 4-year history of progressive left lower-extremity paresis and back pain that radiated into the left leg. At age 13, she was diagnosed with dextroscoliosis, which was treated with a body cast. The patient was asymptomatic for the next 4 decades except for a subjective history of progressive scoliosis. At age 50, she noted new onset of left leg paresis; this was accompanied by radicular thoracic pain localized to the T8 dermatome. Urinary urgency began within the past year, although bowel function was normal. There was no history of recent or remote trauma. She denied neurosurgical or orthopedic investigation of her symptoms before this presentation and had not sought evaluation for the progressive scoliosis

FDA device/drug status: not applicable.

Nothing of value received from a commercial entity related to this manuscript.

* Corresponding author. 244 E. Pearson St. #1705, Chicago, IL 60611. Tel.: (312) 515-6148; fax: (312) 695-4075.

E-mail address: p-poresnky@md.northwestern.edu (P. Porensky)

1529-9430/07/$ – see front matter
doi:10.1016/j.spinee.2006.08.006

subsequent to her diagnosis in adolescence. Spine films from age 13 were not available for comparison.

On physical examination, the patient was noted to have a truncal shift with elevation of the left hemipelvis and right shoulder. Formal leg length measurements were not performed, although a discrepancy was clinically evident. There was decreased strength in the left lower extremity: iliopsoas 3/5, quadriceps 4/5, hamstrings 4/5, anterior tibialis 1/5, and gastrocnemius 1/5. Lower-extremity reflexes were 2+ and symmetric. Clonus was not present; Babinski was present in the left toe. Sensory examination revealed a T7 sensory level to pinprick; the left side appeared hypesthetic to the right. The patient was ataxic and required a cane for ambulation. There were no neurocutaneous stigmata noted.

Presurgical imaging

Magnetic resonance imaging (MRI) and computed tomography (CT) myelography revealed multiple abnormalities. The thoracic dextroconvexity measured 64° from the top of T5 to the bottom of L2; there was a left-sided coronal shift and positive sagittal balance (Figs. 1A, B). An intradural extramedullary mass, mostly consistent with an epidermoid tumor, compressed the cord at the T7–T8 level (Figs. 2, 3). Diastematomyelia was seen in two discrete areas, with a cephalad split from T4–T5 to T8–T9, followed by cord rejoining and a more caudal diastematomyelia from T11 to L2–L3 (Fig. 4). There was a short segment bony septum at T8 (Fig. 5); the conus was low lying, ending at L4.

Intraoperative course

A midline thoracolumbar exposure was followed by T4–T9 laminectomy. At approximately T8, dura was in continuity with the spinous process dorsally. Arising from the spinous process was a bony spicule that appeared to course ventrally within the canal. The dura was taut around this intervening structure and was opened in an elliptical fashion, thereby leaving a central island of dura adherent to the spicule. Once the dura was opened, we noted underlying arachnoid adhesions and a large pearly white mass (Fig. 6). The mass was grossly consistent with epidermoid tumor; this was debulked and dissected away from both the fibro-osseous septum and adherent nerve roots. The septum divided the spinal cord and was resected down to its base on the dorsal aspect of the vertebral body. The cord was no longer tethered and appeared to reapproximate in the midline. The dura was repaired with a bovine pericardium patch. Neurophysiologic monitoring was stable throughout the procedure.

Postsurgical imaging

A CT scan and MRI show T4–T9 laminectomy, excision of the mass from T6–T8, and resection of the diastematomyelia (Fig. 7). The thoracolumbar diastematomyelia and low-lying conus remain stable.

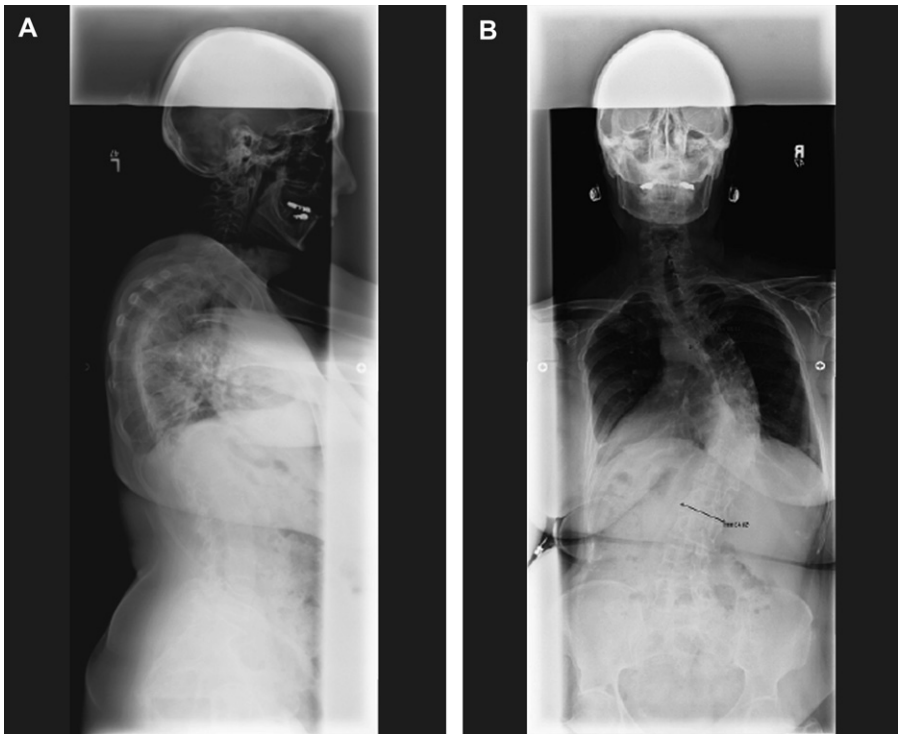

Fig. 1. (Top) Preoperative lateral scoliosis X-ray. (Bottom) Preoperative anteroposterior scoliosis X-ray.