A split cord malformation (SCM), also called a diastematomyelia, is a rare spinal anomaly and refers to a sagittal division of the spinal cord into 2 symmetrical or asymmetrical hemicords. A variant of this malformation associated with a split of the spinal column, spinal bony spurs, myelocoeles, myelomeningoceles, lipomas, and dermal sinuses has been previously reported in the literature. We report an unusual case of SCM with segmental vertebral anomalies.

Case Report
A 5-year-old girl was born as a low-birth-weight baby of 1190 g at 36 gestational weeks. Pregnancy and family history were unremarkable, but the short and thin left lower limb was noticed by her parents soon after discharge. She was diagnosed with SCM by MR imaging at that time, though clinical detail and MR imaging data were not available. She presented with a 5-year history of worsening neurologic dysfunctions but the short and thin left lower limb was noticed by her parents soon after discharge. She was diagnosed with SCM by MR imaging at that time, though clinical detail and MR imaging data were not available. She presented with a 5-year history of worsening neurologic dysfunctions. Pregnancy and family history were unremarkable, but the short and thin left lower limb was noticed by her parents soon after discharge. She was diagnosed with SCM by MR imaging at that time, though clinical detail and MR imaging data were not available. She presented with a 5-year history of worsening neurologic dysfunctions. There was a small dermal sinus rostral to the anus, without communication to the intradural space. CT of the spine revealed a spina bifida at the L2 to S2 levels, and there were congenital vertebral anomalies associated with hypoplasia of T12 and L1. MR imaging revealed the presence of a split cord beginning at the L2 vertebral body level in a single dural sac, and the distal thecal sac was widened (Fig 2A). A tethered cord was seen at the L5 level (Fig 2B). The right hemicord had an intradural connection to the upper cord and a relatively normal configuration. On the other hand, the proximal part of the left hemicord, with an aberrant course anterolaterally in the spinal canal, was tapered to a point of complete absence of the dural sac (Fig 2C–E) and had no apparent intradural connection to the upper cord. The distal part of the left hemicord resumed a normal appearance and presented the lateral set of nerve roots arising from the left hemicord (Fig 2F). CT myelography showed the left hemicord with the small remnant of subarachnoid space running through the intravertebral cleft at the level of spinal anomalies (T12–L1). CT myelography showed the left hemicord with the small remnant of subarachnoid space running through the intravertebral cleft at the level of spinal anomalies (T12–L1). CT myelography showed the left hemicord with the small remnant of subarachnoid space running through the intravertebral cleft at the level of spinal anomalies (T12–L1). On the other hand, the proximal part of the left hemicord, with an aberrant course anterolaterally in the spinal canal, was tapered to a point of complete absence of the dural sac (Fig 2C–E) and had no apparent intradural connection to the upper cord. The distal part of the left hemicord resumed a normal appearance and presented the lateral set of nerve roots arising from the left hemicord (Fig 2F). CT myelography showed the left hemicord with the small remnant of subarachnoid space running through the intravertebral cleft at the level of spinal anomalies (T12–L1). CT myelography showed the left hemicord with the small remnant of subarachnoid space running through the intravertebral cleft at the level of spinal anomalies (T12–L1). CT myelography showed the left hemicord with the small remnant of subarachnoid space running through the intravertebral cleft at the level of spinal anomalies (T12–L1). The spinal column in patients with SCMs has nearly always been reported to be abnormal. The lamina is often thick and fused with the ipsilateral or contralateral lamina of adjacent vertebrae, and spine bifida is almost always present. A spinal bifida at the L2 to S2 levels in our case was in keeping with previous descriptions. The lamina is often thick and fused with the ipsilateral or contralateral lamina of adjacent vertebrae, and spine bifida is almost always present. A spinal bifida at the L2 to S2 levels in our case was in keeping with previous descriptions. The lamina is often thick and fused with the ipsilateral or contralateral lamina of adjacent vertebrae, and spine bifida is almost always present.
SCMs in that the vertebral body was dysplastic at the level of T12 and L1, in which a hypoplasia of the subarachnoid space and hemicord on the left was also observed. These findings radiographically similar to ours have been described as segmental spinal dysgenesis (SSD).3-5

The skeletal system develops from mesoderm.6 By about embryonic day 16, the primitive streak begins to regress and cells at the rostral lip of the primitive knot migrate between the epiblast and hypoblast, forming the notochordal process. The
notochord, which develops from the notochordal process, induces surrounding mesoderm (the paraxial mesoderm, derived from the primitive streak) to condense into paired blocks of somites. Each somite becomes differentiated into a ventromedial part called the sclerotome, which will form the cartilage, bones, and ligaments of the vertebral column, and a dorsolateral part called the dermomyotome, which will form the paraspinal muscles and overlying skin. Paolo et al. surmised that the causal insult in SSD results from chorda-mesodermal derangement during gastrulation. SSD has been reported as an autonomous entity with characteristic clinical and neuroradiologic features and a rare congenital spinal anomaly characterized by localized agenesis or dysgenesis of the lumbar or thoracolumbar spine and focal abnormalities of the underlying thecal sac, spinal cord, and nerve roots. Below the segmental agenesis, the bony spinal canal, thecal sac, and spinal cord resume a normal appearance. 

Previous studies of imaging in patients with SSD have reported that CT myelography shows marked bony narrowing of the spinal canal with the small remnant of subarachnoid space, and MR shows tapering of the spinal cord to a point of marked narrowing or complete focal absence. The radiologic findings of the left hemicorn in our case were in keeping with these previous descriptions. Therefore, we concluded that our case was the rare variant of SCM with coexisting SSD.

With the assumption by Paolo et al., we thought that these unusual findings could be caused by an isolated developmental aberration and propose the following mechanism (Fig 6). Under the unified theory of Pang et al., the abnormal communication between ectoderm and endoderm causes “regional” splitting of the notochord. It would be possible that, when each separated notochord is induced surrounding the paraxial mesoderm, an embryologic derangement of the paraxial mesoderm may occur on only the left notochord. This may have stimulated the dysgenesis of left hemivertebra and hemicorn. Dias et al. showed that in complex dysraphic malformations the paired mesodermal anlagen remain separate and develop independently over variable portions of their length. Such a mechanism further supports the validity of our hypothesis. This seems to be in agreement with the very low incidence of the rare coexisting anomalies (SCM and SSD). As another mechanism, we thought that a variation in Pang’s hypothesis also might explain this unusual finding. According to the unified theory of Pang et al., in type I SCMs, the cells of the meninx primitiva pass between the split notochords and migrate around them. The sclerogenic potential of these cells leads to the formation of the midline bony spur and the hypertrophied vertebral body, and the arachnoid develops from the inner lining of those cells. Hence, the bony spur is excluded from the CSF space. During this process, it would be possible that a larger cell population accumulates along the left notochord than the right. Their sclerogenic potential might have stimulated the formation of a marked bony narrowing of left spinal canal with the small remnant of subarachnoid space.

References