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A Type I Split Cord Malformation is a Significant Pathomechanism of Spina Bifida Aperta in Neonates

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Abstract：Spina bifida（SB）is a well known spinal cord malformation which can result in serious neurological consequences in neonates. A new classification of SB based on the embryological pathomechanism of spinal dysraphism has been recently proposed. According to this classification, some SB may result from diastematomyelia in embryos and such SB is referred to as split cord malformations（SCM）and classified into Type Ⅰ and Ⅱ based on the presence and absence of an osseocartilaginous median septum, respectively. However, SCM are thought to be rare pathomechanisms of SB. We retrospectively reclassified 23 cases of apparent SB found in neonates（12 of SB aperta and 11 of SB occulta）according to the new classification. We found 3 cases of Type Ⅰ SCM in the 23 newborns（13%）or 12 newborns with SB aperta（23%）. All cases of Type Ⅰ SCM presented SB aperta associated with Chiari Ⅱ malformation with myelomeningocele. They all had paralysis in the lower extremities. Hypoplasia of the feet and clubfeet were seen in all patients while unilateral defects of the ribs were observed in a patient with thoracic SB. Found in 23%, Type Ⅰ SCM therefore appears to be a significant pathomechanism of SB aperta in neonates.

Key words：**Spina bifida, Split cord malformations, Myelomeningocele, Chiari Ⅱ malformation, Diastematomyelia, Notochord**

Introduction

Spina bifida（SB）is a well known spinal cord malformation which can result in serious neurological consequences in neonates. A new classification of SB, both aperta and occulta, based on an embryological and anatomical understanding of spinal dysraphism has recently been proposed. According to this new classification, some SB cases may result from spinal dysraphism in embryos and these SB cases are called split cord malformations（SCM）or diastematomyelia and they are classified into two types；Type Ⅰ and Ⅱ based on the presence and absence an osseocartilaginous median septum, respectively.[1)2)] However, SCM are thought to be rare pathomechanisms of SB.

In this study, we reclassified apparent SB found in neonates who had been admitted to Fukuoka University Hospital according to the new classification. The clinical presentations of diastematomyelia are herein described and we also evaluated the significance of diastematomyelia in the pathomechanisms of SB in neonates.

Subjects and Methods

A total of 23 newborn infants with apparent SB were admitted to the neonatal intensive care nursery of Fukuoka University Hospital from January 1985 to December 2006. Fifteen were males and eight females. The gestational ages were 32 to 40

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weeks with a mean of 38.79 weeks, 2 were preterm and 21 term newborns. The birth weights were 1,713 to 4,031 g with a mean of 3,007.7 g and 5 were low birth weight babies (<2,500 g).

The 23 babies were admitted to the hospital because of apparent SB identified at birth while 12 had SB aperta and 11 SB occulta. All 12 patients with SB aperta had Chiari II malformation, among whom 7 were associated with myelocele and 5 were with myelomeningocele. In patients with SB occulta, 4 had lipomyelomeningocele 2 congenital dermal sinuses, 2 spinal lipomas, 1 lipomyelocele, and 1 terminal myelocystcele. (Table 1.)

For the reclassification, the diagnoses of Type I and II SCM were made based on the findings of either neonatal CT or magnetic resonance imaging (MRI) of the spine and/or on the findings of postmortem examinations when available. Differentiation between Type I and II was done based on the presence and absence of the septum, respectively. The information required for the reclassification was available from all the patients.

Results

In this study, a diagnosis of Type I SCM was made in three patients (2 boys and 1 girl) with SB aperta. All cases with Chiari II malformation were also associated with myelomeningocele. No Type II SCM was identified. The clinical presentations of the three patients with Type I SCM are summarized below.

Case 1. (*Male*) : The patient was born at 35 weeks of gestation. His fetal ultrasonography showed hydrocephalus and fetal magnetic resonance imaging identified myelomeningocele. He was delivered by cesarean section at 37 weeks and 4 days of gestation. His height was 38.5 cm (−6.0 SD), and body weight was 2,427 g (−1.2 SD). The fontanel was 3 cm×2 cm without distention. A naked neural placode(6.0 cm×7.0 cm)was observed in the thoracic region of the spine. Hypoplasia and clubfeet were observed in the lower extremities and complete paralysis in the lower limbs were noted. He

Table 1. 23 cases with spina bifida

anatomical classification	split cord malformation	sex	level of dysfunction	gastational age (weeks)	birth weight (g)	Apgar score (1 min/5 min)	prenatal diagnosis (hydrocephalus)	Chiari II malformation
SBA, MMC	+	female	Th10	32	1713	4/5	+	+
SBA, MMC	+	male	Th10	37	2427	6/8	+	+
SBA, MMC	+	male	L2	37	2330	7/8	+	+
SBA, MMC		female	L3	38	2947	8/9	+	+
SBA, MMC		male	L5	37	2765	6/8	+	+
SBA, MC		female	L3	38	2731	8/8	+	+
SBA, MC		female	L5	37	2247	7/8	+	+
SBA, MC		male	L5	37	3655	8/9	+	+
SBA, MC		male	L5	39	3125	9/9	+	+
SBA, MC		male	S1	38	2844	10/10	−	+
SBA, MC		female	S2	38	2537	9/9	+	+
SBA, MC		male	S2	40	2973	9/9	−	+
SBO, LMMC		male	S1	39	3300	9/10	−	−
SBO, LMMC		male	S3	37	2592	9/9	−	−
SBO, LMMC		male	S3	40	3338	9/9	−	−
SBO, LMMC		male	−	39	2688	9/10	−	−
SBO, LMMC		female	−	40	2976	9/10	−	−
SBO, LMC		male	S3	39	3540	10/10	−	−
SBO, SL		male	S1	40	3245	9/10	−	−
SBO, SL		female	S3	36	2220	8/10	−	−
SBO, DDS		male	−	37	2824	2/9	−	−
SBO, DDS		female	−	40	3000	8/9	−	−
SBO, TMCC		male	L4	40	4031	5/10	−	−

SBA, spina bifida aperta; SBO, spina bifida occulta; MMC, myelomeningocele; MC, myelocele; LMMC, lipomyelomeningocele ; LMC, lipomyelocele ; SL, spinal lipoma ; DDS, dorsal dermal sinus ; TMCC, terminal myelocystcele

underwent an emergency operation for spinal repair on day 0. Ventricular drainage was placed for hydrocephalus due to Chiari II malformation on day 9. MRI after the operations revealed spinal dysraphism and an osseous spur in the thoracic region (Fig. 1).

Case 2. (*Female*) : The patient was born at 32 weeks of gestation. Her fetal ultrasonography findings showed hydrocephalus. The patient was delivered by an elective cesarean section at 32 weeks and 5 days of gestation. Her height was 33.0 cm and body weight was 1,713 g. The fontanel is 4.5 cm×4.5 cm and had distention. The size of myelomeningocele was 4 cm×4 cm×2 cm. Conse-

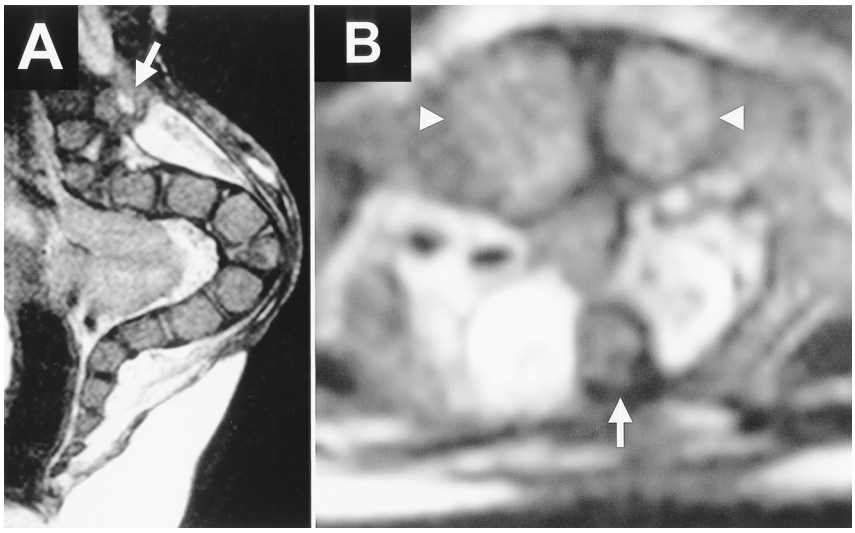

Fig. 1. A spinal MRI image demonstrated an osseus spur on the Th8 vertebral column coexisting butterfly vertebrae at Th8. (A) Sagittal T2-wighted MR image shows a bony spur (arrow). (B) Horizontal T2-weighted MR imaging demonstrate butterfly vertebrae (arrowheads) and a bone spur (arrow).

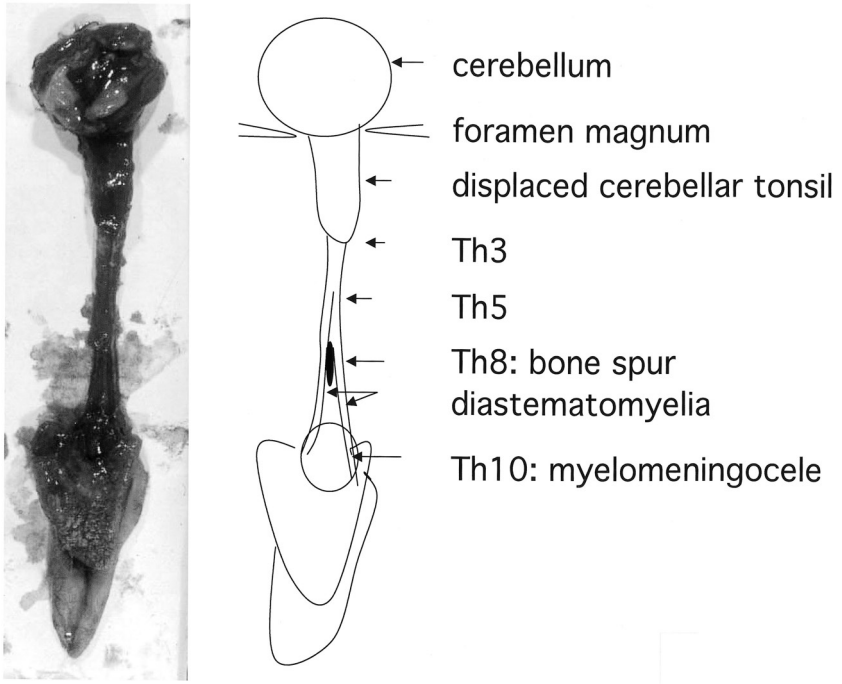

Fig. 2. A displaced cerebellar tonsil is seen. Diastematomyelia from Th5 and a bony spur at Th8. Myelomeningocele was found at Th10.

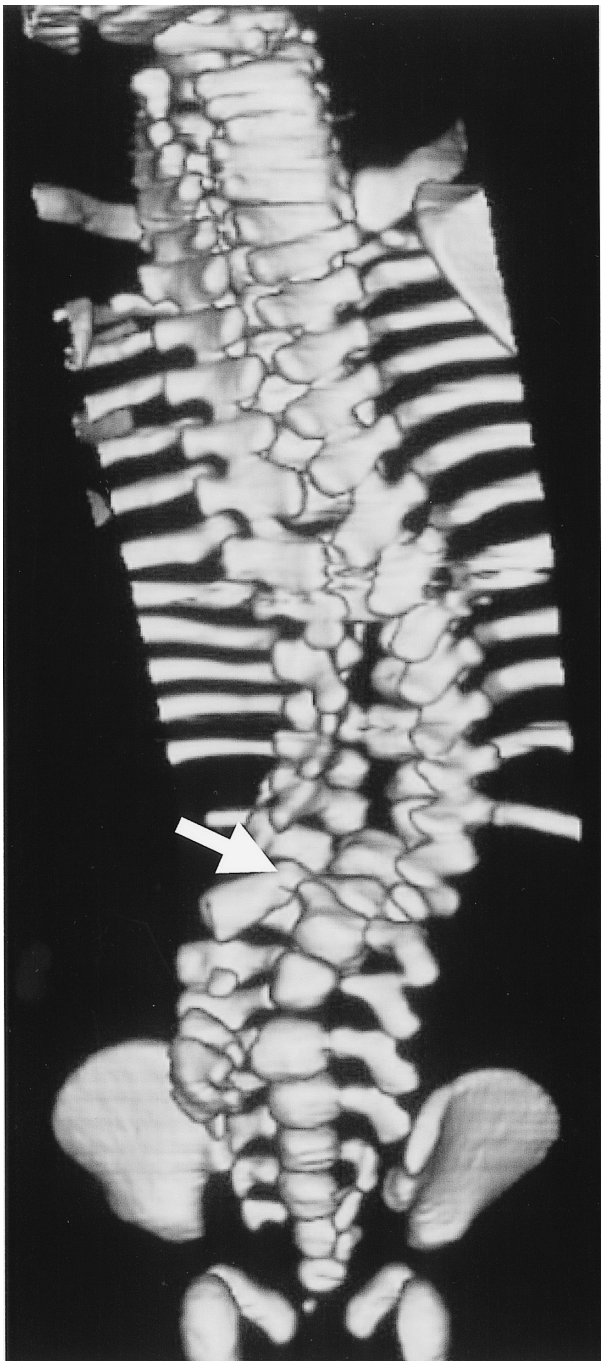

Fig. 3. Spinal 3 Dimmensional CT image demonstrated an osseous spur on the L3 vertebral column (arrow) coexisting with hemi vertebrae at Th11, 12.

cutive defects of the left fifth to twelfth ribs were observed on chest X-rays. She also had clubfeet and complete paralysis in the lower extremities. The patient died eight hours after admission. No surgical intervention was performed at the parents' request. A postmortem examination confirmed diastematomyelia with a bony spur and myelomeningocele (Fig. 2).

Case 3. (*Male*) : The patient was born at 37 weeks of gestation. His fetal ultrasonography showed hydrocephalus and fetal MRI showed multiple myelomeningoceles. He was delivered by cesarean section at 37 weeks and 0 day of gestation. His body weight was 2,330 g. His Apgar scores were 7 and 8 at 1 and 5 minutes, respectively, whereas mechanical ventilation and the artificial surfactant replacement therapy were required for respiratory distress syndrome. A naked neural placode was observed in the lumbar region of the spine. Hypoplasia and clubfeet of the lower extremities were observed. He could only move his left thigh. Complete paralysis was noted in the right lower limbs. An emergency operation for spinal repair was performed on day 0. A ventriculo-peritoneal shunt operation was performed for a Chiari II malformation on day 8. MRI after the operations demonstrated spinal dysraphism with an osseous spur in the lumbar region (Fig. 3).

Dicussion

In this study, we found 3 cases of diastematomyelia with a septum or Type I SCM in 23 newborns with apparent SB (13%) or 12 newborns with SB aperta (23%). All cases of Type I SCM presented SB aperta with Chiari II malformation associated with myelomeningocele. They all had paralysis in the lower extremities. Hypoplasia and clubfeet were seen in the patients with SB in the lumbar and thoracic region while unilateral defects of the ribs were seen in the patients with SB in the thoracic region in one case.

SB can result from spinal dysraphism, which indicates a defective closure of the neural tube in fetal life, which thus encompasses heterogeneous pathomechanisms. Among spinal dysraphism, SCM is a more specific term to describe the spinal dysraphism resulting from deranged embryogenesis of the notochord and vertebrae.[1)-4)] There is a synonym, diastematomyelia, which originated from two Greek words ; *diastemato* meaning a cleft and *myelos* meaning the cord.[5)]

The unified theory of embryogenesis now suggests that all SCM originate from a single ontogenetic defect which occurs around the time when the primitive neurenteric canal closes. The defect

may form an' accessory neurenteric canal' through the midline embryonic disc. Therefore, this dimorphic feature allows a communication between the yolk sac and the amnion, thus leading to contact between ectoderm and endoderm in the canal. This abnormal fistula consequently causes regional ' splitting' of the notochord and the overlying neural plate.[1] Experimental SCM produced by the surgical induction of a dorsal midline fistula supports the theory underlying SCM[6] .

Recently a new classification of SB, both aperta and occulta, based on an embryological and anatomical understanding of spinal dysraphism, as described above, has been proposed. According to this new classification, the term SCM is recommended to be used for all double spinal cords and two types of SCM have been identified. Type I consists of two hemicords, each of which is unsheathed by its own dural tube and separated by a dura－sheathed, rigid, osseocartilaginous median septum. Type II is characterized by two hemicords housed in a single dural tube separated by a non－rigid, fibrous median septum.[1]

Regarding the prevalence of SCM in spinal dysraphism, several studies have been carried out focusing on myelomeningocele whereas few have dealt with the newborns in relation to apparent SB according to the new classification including the differentiation for Type I and II SCM. For example, diastematomyelia is seen in from 31% to 46% of patients with myelomeningocele.[3] Kumar R and colleagues[7] found 46 cases of SCM out of 138 cases of spinal dysraphism. They found 19 cases of SCM with myelomeningocele and 15 cases of type I SCM. Therefore, SCM with myelomeningocele was found in 41 % of total SCM cases. Pang D and colleagues[2] found 13 cases of surgically treated myelomeningoceles in 31 cases of SCM (42%). Reigel DH and collogues[8] found 13 cases of diastematomyelia (11%) in 123 cases of SB. Taken together, though no precise information regarding the prevalence of SMC in newborns with apparent SB is yet available, the prevalence is assumed to be low.[9]

Since we have shown that SCM type I contribute to 23% of the pathogenesis of SB aperta in this study, SCM type I thus seems to be a significant cause of SB aperta in newborns. The finding that SCM type II was not found may be consistent with that fact that SCM type II is found in individuals with subtle spinal neurological signs. Open spinal dysraphisms are usually operated on at birth, without MRI and hence the pathomechanism according to the new classification might not have been considered previously. In these days a prenatal diagnosis of diastematomyelia is possible as early as the midtrimester by ultrasonography, and fetal MRI can also be applied to accurately diagnose the type of diastematomyelia.[10][11] SCM hence may be diagnosed more often than before in SB, and it is considered to be a significant embryological pathomechanism of SB in neonates.

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