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Spinal Congenital Dermal Sinus

— Experience of 5 cases over a period of 10 years —

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Spinal congenital dermal sinus (CDS) is a rare entity which supposedly results from the failure of the neuroectoderm to separate from the cutaneous ectoderm during the process of neurulation. The lesions are most frequent at the lumbosacral followed by the occipital region. CDS of the thoracic region is very rare. The patients with spinal CDS present with meningitis and/or mass effect from the associated inclusion tumor. They are usually dermoid or epidermoid cysts. Teratoma is rarely associated.

The authors experienced 5 cases of spinal CDS over a 10 year period. Of the 5 cases, 2 were at thoracic and 3 were at lumbosacral levels. Dermoid cyst, epidermoid cyst and teratoma were associated in one case each. Two cases presented with neurological deficit and meningitis while an additional case presented with neurological deficit and a history of probable meningitis. Pain was present in 2 cases. Magnetic resonance imaging played an important role in the diagnosis of the lesion and planning of surgery. All the cases showed a good response to surgery even though one patient had persistent neurological deficit.

Key Words: *Spinal congenital dermal sinus, Thoracic spine, Teratoma, Magnetic Resonance Imaging*

INTRODUCTION

Congenital dermal sinus (CDS) is a lesion connecting skin to deeper structures including neural tissue. It supposedly results from the failure of the neuroectoderm to separate from the cutaneous ectoderm at the third to fifth week of gestation. CDS tracts extending to the spinal canal are rare, the incidence is approximately 1/2,500 live births (Powell et al., 1975;Ceddia et al., 1990). Lumbosacral and occipital regions are frequently involved while only 6 to 10% of lesions are at the thoracic area (Powell et al., 1975; French, 1989; Venger et al., 1989). CDS is commonly associated with inclusion cysts, however, association with teratoma is very rare (French, 1990).

In Korea, only one case of spinal CDS has been described in the literature (Chung et al., 1975).

The authors have experienced 5 cases of spinal CDS since October 1982. The rarity of the lesion prompted us to study the clinical features and surgical results of the cases retrospectively.

CLINICAL MATERIALS AND METHODS

From October 1982 to December 1992, 7 cases of congenital dermal sinus with intracranial or intraspinal extension were operated on at the Department of Neurosurgery, Seoul National University Hospital. Of those lesions, 2 were at the occipital area. The fifth case of spinal CDS was operated on in January 1989. To analyze the location of the lesions, clinical manifestations, associated pathol-

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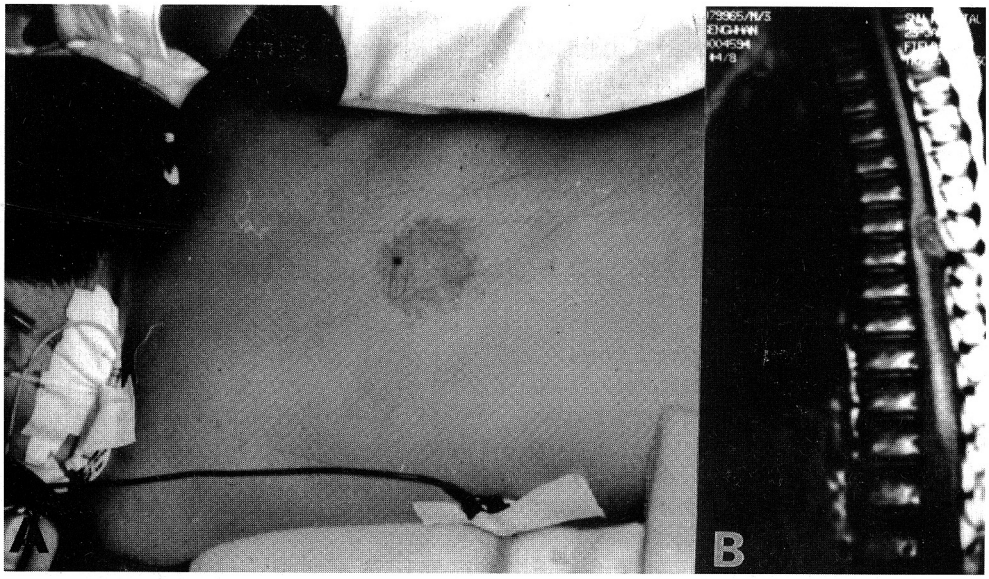

Fig. 1. A: Skin stigmata of case 1. Hemangioma, dimple with focal hypertrichosis were noted. A paperclip was applied and removed at the level of the dimple resulting in the clip mark on the skin. B: Thoracic spine MRI (T1 weighted, sagittal) showing the CDS tract and a mass compressing the spinal cord from the posterior aspect.

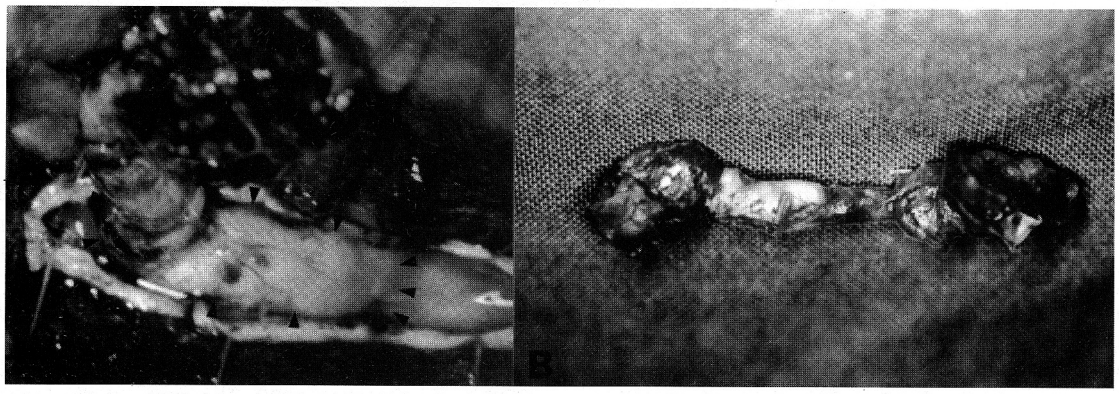

Fig. 2. A: Operative photograph of case 1 showing the CDS tract attached to the intramedullary mass (arrow heads) which was located at the posterior aspect of the spinal cord. B: Pathological specimen. The right side is the skin while the left side is the intramedullary dermoid cyst.

ogy and operative results, medical records, related photographs and radiological data were reviewed. The associated mass was diagnosed histologically with a light microscope. In this study CDS's combined with other spinal dysraphism (for example, lumbosacral lipomatous lesions) were excluded.

ILLUSTRATIVE CASES

Case 1

A 2-year-old boy was admitted due to paraparesis. He had appeared healthy until 5 days before admission when progressive paraparesis appeared. On the thoracic back there was a small dimple with hypertrichosis and surrounding focal

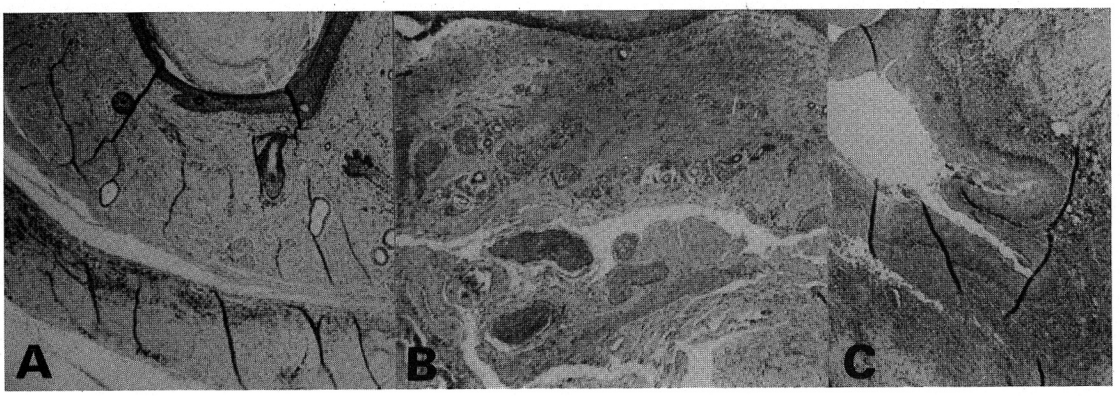

Fig. 3. Histological section of case 1 demonstrating stratified squamous epithelium with keratin material and many inflammatory cells. Dermal appendages were noted. (hematoxylin and eosin, x40).

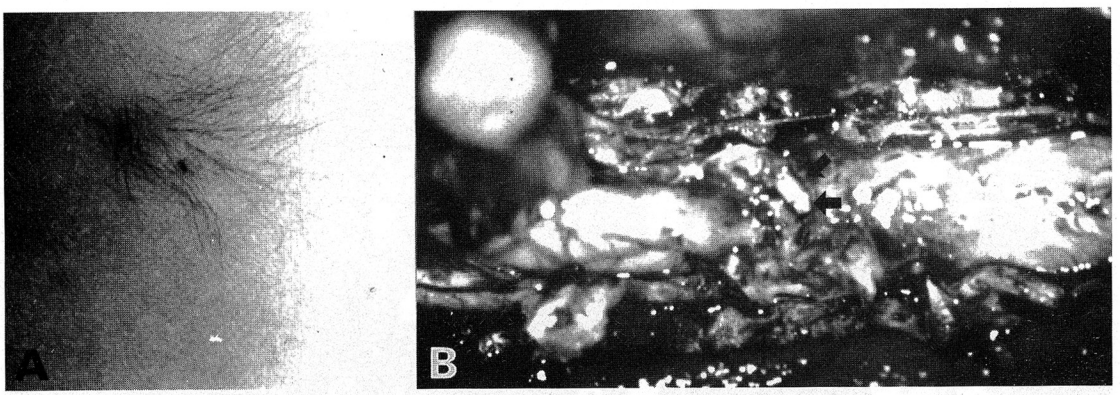

Fig. 4. A: Skin lesion of case 2. Hemangioma, dimple and hypertrichosis were noted. B: Operative photograph showing the CDS tract from the skin surface to the spinal cord. At the junction (arrows), there was a dense fibrous adhesion.

hemangioma (Fig. 1, A). Physical examination revealed fever, nuchal rigidity, urinary incontinence, paraparesis of grade II/V and bilateral extensor plantar response. No abnormality was found on plain spine radiographs. On magnetic resonance imaging (MRI) an oval mass compressing the spinal cord from the posterior aspect was noted at the level of T6 and T7. A tract connected the mass to the skin through the space between the spinous processes of T7 and T8 (Fig. 1, B). The sinus tract and the dermoid cyst were *en bloc* removed (Fig. 2). Histologically the tract consisted of stratified squamous epithelium with keratin material (Fig. 3, A). Dermal appendages and many inflammatory cells were noted (Fig. 3, B-C). Though the spinal curvature became mildly kyphotic after surgery, the urinary incontinence and paraparesis improved and motor power became normal one month later.

Case 2

A 3-year-old boy was admitted with the chief complaint of intermittent paraparesis. For the six months preceeding admission he had suffered from fever and paraparesis. On admission signs of infection were absent. Physical examination revealed a skin dimple with hypertrichosis and hemangioma on the thoracic back (Fig. 4, A), paraparesis of grade III/V with bilateral extensor plantar response. Plain spine radiographs were normal. Myelography showed a complete block of the subarachnoid space at the level of the upper end of T12 vertebral body. The tract of CDS entered the spinal canal through the space between the spinous processes of T11 and T12. It was attached to the posterior surface of the spinal cord

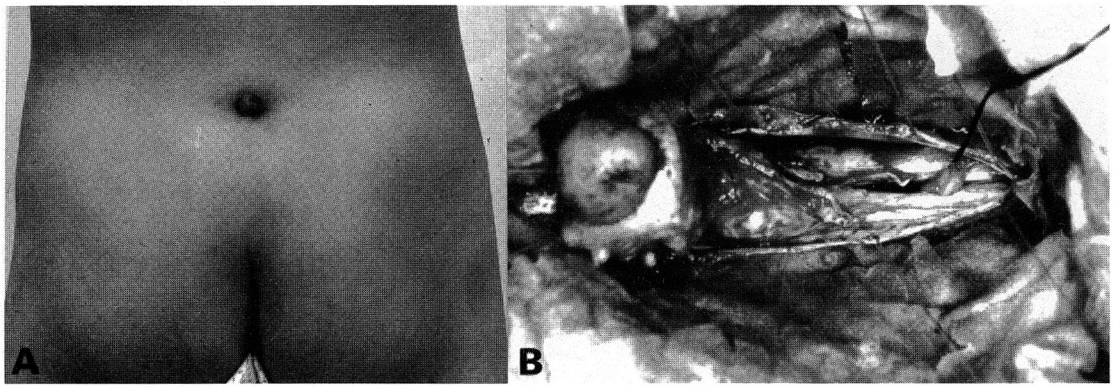

Fig. 5. A: In case 3 there was a pedunculated bean-sized mass on his lumbosacral back. B: Operative photograph demonstrating the CDS tract connecting the cutaneous lesion to the dorsal surface of the spinal cord. The proximal part of the spinal cord was split.

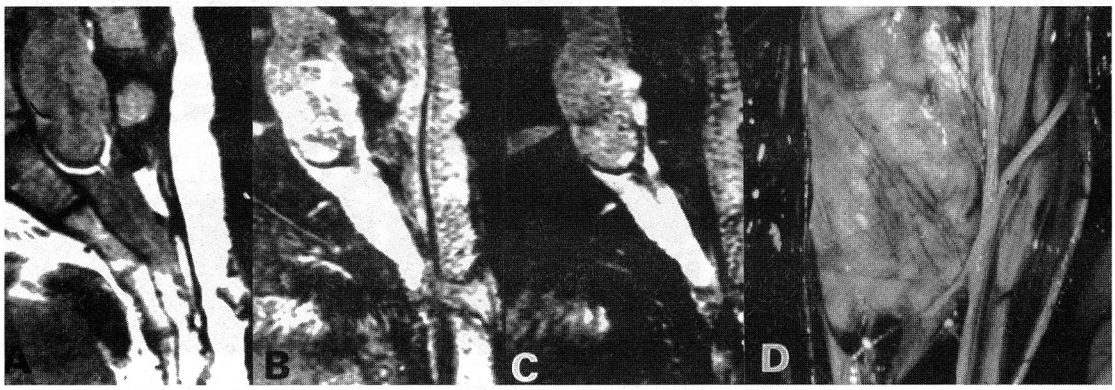

Fig. 6. A: Lumbosacral spine MRI (T1 weighted image, sagittal) of case 4 showing an intraspinal mass and the CDS tract from the lower end of the mass to the skin. B and C: Gradient echo showing the skin lesion and part of the tract more clearly. D: Operative photograph demonstrating an intraspinal mass and laterally displaced cauda equina.

with the surrounding fibrous adhesion (Fig. 4, B). For removal, the tract was cut at the fibrous junction with the spinal cord. Postoperatively he regained power in the lower extremities and began to walk one month later.

Case 3

A 16-year-old boy was admitted after suffering from left leg pain for 2 months. On his lumbosacral back, there was a bean-sized pedunculated mass with whitish discharge (Fig. 5, A). Knee jerk, ankle jerk and the dorsiflexion of the big toe were weak on the left side. Though the body temperature was normal, the white cell count in the cerebrospinal fluid was increased to 268 cells/mm^3 (polymorphonuclear cells 17%, lymphocytes 81%, protein 85 mg/dl, glucose 53 mg/dl) suggesting meningitis. *S. epidermidis* and *S. aureus* were cultured from the discharge of the sinus tract. The CDS tract passed through the interspinous process space between L4 and L5. The small cutaneous teratoma was also removed. Conus medullaris was located at the level of L4-5 and the spinal cord was split at the level of L3-4 (Fig. 5, B). After surgery the pain disappeared. Left leg deep tendon reflexes and motor power became normal one month later.

RESULTS

The 5 cases of spinal CDS are summarized in Table 1 and Fig. 7. There are four boys and one woman. The ages ranged from 1 month to 24 years. The chief complaint was paraparesis in the

Table 1. Summary of 5 cases with spinal CDS

	Case 1	Case 2	Case 3	Case 4	Case 5
age/sex	2/M	3/M	16/M	24/F	1 mo/M
chief complaint	paraparesis	paraparesis	leg pain	lumbago	skin dimple
infection	+	+ (history)	+	–	–
location of skin dimple	T8-9	T12-L1	L5-S1	S3	S3
entry into the spinal canal	T7-8	T11-12	L4-5	S2	S2
associated mass	dermoid	–	teratoma	epidermoid	–
result	normal life mild kyphosis	normal life	normal life pain relief	static ND* pain relief	normal life dimple removed

ND=neurological deficit: urinary incontinence, saddle anesthesia

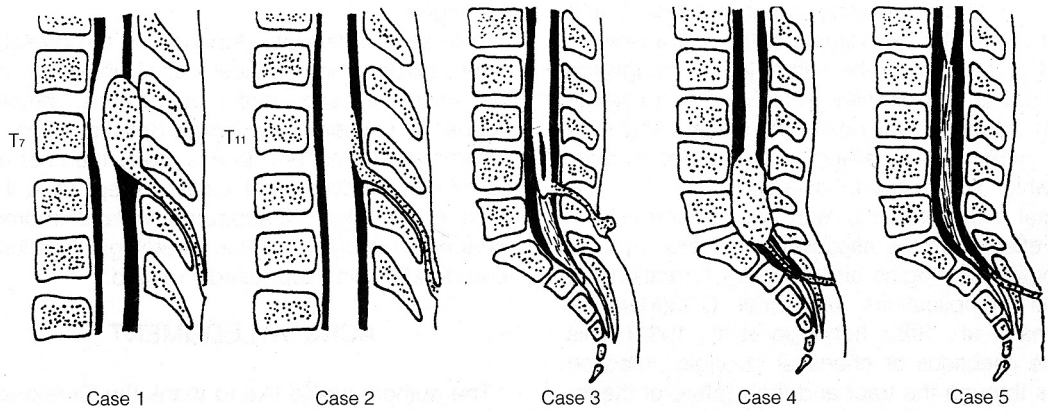

Fig. 7. Schematic drawing of 5 cases.

two young children (2 and 3 years of age), pain in 2 cases (one adolescent and one young woman) and skin dimple in 1 new born case. All the cases had skin dimples located about 1 vertebral segment lower than the entry level into the spinal canal. Two cases (case 1 and 3) showed signs of meningitis with negative culture and an additional case (case 2) had a history of probable meningitis. Dermoid cyst, epidermoid cyst and teratoma were associated in one case each. In 2 cases (case 1 and 4) MRIs were taken. The MRI delineated the pathological anatomy clearly (Fig. 1-B and 6). The associated masses were removed totally without additional neurological deficits. All the cases, except one (case 4), showed good postoperative course with normal daily activities. In case 4, even though the pain was relieved, the preexisting urinary incontinence and saddle anesthesia did not improve.

DISCUSSION

Spinal CDS was first described by Moise in an 18-year-old boy who presented with staphylococcal meningitis (Moise, 1926). Walker and Bucy in 1934 coined the term "congenital spinal dermal sinus" to describe defective separation of the neuroectoderm from the surface ectoderm resulting in an epithelial tract penetrating a variable distance into the deeper tissue, including the spinal cord (French, 1990). Along the tract, inclusion cyst, in most cases dermoid or epidermoid cyst, can occur at any site. Teratoma is associated on rare occasions, in approximately 4% of spinal CDS cases (French, 1990). Case 3 of the present series had a cutaneous teratoma.

The usual sites of the lesion are the lumbosacral and occipital areas. Only 6 to 10% of spinal CDS's were located at the thoracic region (Cheek and Laurent, 1985;Venger et al., 1989;Ceddia et al.,

1990; French, 1990). In this study 2 out of 5 spinal CDS's were located at the thoracic area. The high proportion of thoracic CDS's in this series may be due to the small number of cases.

Because of the "ascent" of the spinal cord compared to the vertebral bony structures, the CDS tract courses cephalad as it enters into the spinal canal. In this study there was about 1 vertebral segment difference between the levels of the skin dimples and the entry level into the spinal canal. Congenital sacrococcygeal dermal sinus is another entity because the embryology of the region is different from CDS's of the area above it. Sacrococcygeal CDS rarely extends into the spinal canal, if ever (French, 1990). In this series, one CDS located at S3 level extended to the epidermoid cyst at the low-lying conus medullaris and the other with the skin dimple at the same location ended at the dura of the sacral level. Though the levels of the skin dimples were S3, the cranial direction of tracts as they enter the spinal canal suggests 'lumbosacral' rather than 'sacrococcygeal' CDS which shows caudal direction.

Spinal CDS presents with skin abnormalities, mass effect from the associated inclusion cyst, or meningitis. Meningitis and abscess formation are serious complications of spinal CDS(Maurice-Williams et al., 1980; Schnegg et al., 1981). This can be infectious or chemical in origin. Infection occurs through the tract and the rupture of the inclusion cyst causes chemical meningitis. In this series case 1 and 3 had meningitis. Though the culture of microorganisms isolated no causative organism they were thought to be infectious because there was no evidence of rupture of the associated masses at surgery. Case 2 seemed to have a history of infection with aggravation of neurological deficit which suggested meningitis or intraspinal abscess formation. Dense fibrous adhesion at the junction between the CDS and spinal cord supported the suspicion of history of meningeal inflammation.

Diagnosis of spinal CDS largely depends on the high index of suspicion. Skin stigmata and repeated meningitis of unknown origin are important clues. Radiological diagnosis is advanced by MRI which clearly demonstrates the extraspinal portion of the CDS tract and associated mass lesions (Rindahl et al., 1989; Venger et al., 1989). The intraspinal portion of the CDS tract is not always well delineated by MRI (Barkovich et al., 1991). In young infants sonography has been tried. However, the tracts ascending toward the spinal cord are not clearly demonstrated (Naidich et al., 1986). In this series Case 1 and 4, who were operated on after 1987, were diagnosed by MRI. The tracts of CDS (even the intraspinal portion of case 4) and the associated masses were well visualized by MRI. The good delineation of pathological anatomy in those cases conformed to the idea that MRI is the diagnostic procedure of choice.

The treatment of CDS is prophylactic surgical removal of the lesion for easier surgery and better outcome (Matson, 1969; Cheek and Laurent, 1985; Venger et al., 1989; French, 1990). In cases with infection, surgery is recommended after control with antibiotics. However, due to rapidly progressing neurological deficit, case 1 of this series was operated on urgently in spite of evident meningitis.

The result of surgery depends on the presence of pre-existing neurological deficit, infection, and the nature of associated mass lesions. Infection frequently causes neurological deficits which are sometimes permanent. In this study the final outcome was excellent in all cases except case 4. In case 4, lumbago disappeared without improvement of neurological deficit. Neurological deficit of long duration was supposedly related.

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In Korea, only one case of spinal CDS has been described in the literature (Chung et al., 1975).

The authors have experienced 5 cases of spinal CDS since October 1982. The rarity of the lesion prompted us to study the clinical features and surgical results of the cases retrospectively.

CLINICAL MATERIALS AND METHODS

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