A Case of Tethered Cord Syndrome Manifested with a Neurotrophic Ulcer

Dae Young Kim, M.D., Seung Chul Lee, M.D., Young Ho Won, M.D., Inn Ki Chun, M.D.

Department of Dermatology, Chonnam University Medical College, Kwangju, Korea

We report herein a case of tethered cord syndrome with recurrent ulcerations of the skin in a 17-year-old girl. The patient presented an asymptomatic punched-out deep ulcer on the right buttock and right heel. The lesions were noticed as bullae on the sites, which developed into ulceration. At birth, a reddish nodule was noted on the lower back, which was excised when she was 3 years old at a private clinic. Neurological examination revealed hypoesthesia below the L₅ dermatome. We suspected a simple neurotrophic ulcer which was treated with zinc oxide dressing and oral antibiotics. 1 month later, the ulcers had healed, but tense bullae had developed on the same right buttock. Spina bifida with intraspinal lipoma was revealed by plain X-ray film and magnetic resonance imaging. Tethered cord syndrome was diagnosed, and treated with a surgical excision of the lipoma, and repair of the dura.

This is a very rare etiology of a neurotrophic ulcer due to a central nervous system disease, and is the first reported case in the Korean dermatologic literature.

(Ann Dermatol 7:(4)346–349, 1995)

Key Words: Neurotrophic ulcer, Tethered cord syndrome

Neurotrophic ulcers are classic chronic ulcers that appear over the areas of pressure and trauma in the skin that has lost sensation. These are classically seen in diabetes and leprosy but also occur in a large number of other forms of neuropathic and neurologic disease.

One of the etiology of a neurotrophic ulcer is tethered cord syndrome, which is a condition where tight filum terminale or intraspinal lipoma causes overstretching or compression of the caudal part of the spinal cord. Sensory deficits are present in two thirds of all patients, but ulcer are rarely observed.

As far as we know, no case of tethered cord syndrome with complicating chronic bulla and ulcers has been reported in the Korean literature.

REPORT OF A CASE

A 17-year-old girl visited our hospital with recurrent ulcers on her right buttock and heel in February, 1993 (Fig. 1). The ulcers on her buttock were first noticed about 2 years ago and recurred 3 times during the last 2 years. On past medical history, she had a reddish nodule on her lower back in her early childhood, which was removed without histopathologic examination 15 years ago. She has been suffering from urinary incontinence for the last 8 years, especially severe in the night. The family history was not contributory.

On physical examination, there were a round, deep punched-out ulcer with darkish or yellowish crust on the right buttock. A diffuse elevated palm-sized soft mass with a linear scar from the previous operation on the overlying skin was observed and palpated on the lower back. Hyper-
Neurotrophic scars were also found on the left extremity and buttock. She has a normal growth and general appearance.

Neurological examination revealed hyposthesia below the L₅ dermatome, but this was normal in motor function. The normal deep tendon reflex of the patellar and achilles was lost, and the pathologic reflex was absent. Motor nerve conduction test was within normal ranges, and needle electromyography had no abnormal spontaneous activities. However the sensory nerve conduction test revealed mild decrease in the amplitude and latency on the right lower extremity compared with that of lower extremity.

Routine laboratory findings including complete blood cell count, urinalysis, stool examination, liver function test, renal function test, VDRL test and blood sugar were within normal limits or negative. The plain X-ray of the lumbo-sacral spine revealed a fusion failure of L₅ and sacrum. On the MRI examination, there was an extensive bony defect on the posterior neural arch of L₅ and sacrum, which was replaced with an abundant fatty mass extended from the overlying subcutaneous mass (Fig. 2). The fatty mass anchored the thickened filum terminale.

The patient was diagnosed as having a neurotrophic ulcer by tethered cord syndrome and re-
ferred to the Department of Neurosurgery for the surgical correction. After the spinal cord was un-
tethered microsurgically, the fatty mass attached to spinal cord was carefully removed as much as possible and the dura was repaired. Gross apper-
ance of the removed large mass was yellowish color similiar to fat tissue. The histopathologic findings was characteristics of lipoma with a mature fatty cell(Fig. 3). Three months after the operation, hypo-
esthesia was not changed, but her urinary in-
continence had improved slightly and no other neurological complications and skin ulcers had developed(Fig. 4).

DISCUSSION

Neurotrophic ulcer is a chronic ulceration that de-
velops in the anesthetic skin. Characteristically it is
painless, persistent, and uninflamed, and appears in
areas subject to trauma or pressure. The most
common site is the foot at its pressure points, par-
ticularly on the sole over the metatarsal heads.
The lesion becomes soft, moist, and malodorous, and
later exudes a thin purulent discharge. Deeper
perforation and secondary infection often lead to os-
teomyelitis'.

Two broad categories of neurotrophic ulcer are rec-
ognized according to their origin. The first category
is a central nervous system origin. Dementia, cere-
brovascular accidents, or anoxic enccephalopathy
cause pressure sores in many elderly patients. The
spinal cord injuries in traffic accidents such as a
penetrating wound, dislocation or fracture of the
vertebral column are significant in causing a neu-
rotrophic ulcer'. Syringomyelia caused by the in-
complete closure and cavitation of the spinal
cord, and tabes dorsalis of neurosyphilis also causes
a neurotrophic ulcer. Tethered cord syndrome is a
rare etiology of chronic neurotrophic ulcer origin-
ating from the central nervous system.

The second category is a peripheral nervous system
origin. Diabetes and leprosy complicating peripher-
al neuropathy are the most common causes of
neurotrophic ulcer, especially of the foot'. Rarely,
hereditary neurological disorders including domi-
nant multilating sensory polyneuropathy in adults,
recessive multilatory sensory polyneuropathy in
childhood, and familial dysautonomia cause the
neurotrophic ulcer'.

The incidence of neurotrophic ulcer in this syn-
drome is unknown, but it is very rare. The mecha-
nism of neurotrophic ulcer in this syndrome is
based either on the tight film terminale or on in-
traspinal lipoma which causes overstretching and
compression of the spinal cord. So the area in-
volving the dermatome is anesthesia, and the fric-
tion bullae is followed by a rupture of tense bullae.
That area develops a neurotrophic ulcer due to
pressure necrosis, the mean level of cord termina-
in the full-term infant is just above the L1-L3, disc
space, with a range of L1 to L3. The conus reaches at
the mature adult level, where it designates as L2-L3,
approximately 2 months after full-term gestation'.
Although the term, "tethered cord syndrome", was first described by Johnson in 1857, it had been
used in other reports such as tethered conus, tight
filum terminale, and lumbosacral lipoma'. The
ture incidence of the tethered cord syndrome in
the overall picture of occult spinal dysraphism is
unknown. The sex ratio of tethered cord syn-
drome appears to be approximately 2 to 1 for fe-
males'. Tethered cord syndrome consists of an
abnormal low conus medullaris tethered by one or
more forms of intradural abnormality, which is
caused by a short, thickened filum terminale, fi-
brous bands or adhesions, or a totally intradural
lipoma'.

We would stress the presence of skin lesions ac-
companying this syndrome, because early detec-
tion by skin lesions may prevent the permenant
loss of the nerve function. Skin lesions occur in
half of the patients with tethered cord syndrome.
Hypertrichosis is the most common cutaneous
sign evident at birth. Particularly if it is located on
the lumbosacral lesion, careful examination is es-
sential to detect this particular stigma'. The pre-
sence of subcutaneous lipoma is a clear indication of
probable cord involvement, and exploration has
been recomended, even in the absence of neuro-
logic sign. Congenital lipoma are not encapsulated
and are finely lobulated, much like subcutaneous
fat. Hemangiomatus discoloration, capillary
hemangioma of the portwine, and flat type he-
mangioma in lumbosacral lesion have also been
detected. It is an unusual location for an ordinary
hemangioma and should be considered as part of occult
spina bifida until proved otherwise'. Congenital
dermal sinus does not occur commonly, however
may complicate some forms of infection such as
meningitis, and abscess that called attention to
the lesion. An epidermoid or dermoid cyst may form at any point along the dermal sinus. The speed of the patient's worsening condition varies. Micturition may first occur in the adult without evidence of abnormality in the leg. The clinical manifestation is progressively worsens with time. Sensory deficit and bladder dysfunction have sometimes been observed. Pain in the back or leg or in the arches of the feet have also been observed. A relatively high rate of occurrence of scoliosis or kyphosis was noted in the presentations. Radiologic examination was essential for the diagnosis of the disorder. Plain roentgenograms showed posterior or spina bifida, widening interpedicular distance, vertebral body abnormality, and ossified septum. Ultrasonography can be performed on newborns in incubators and infants. Prone and supine water-soluble contrast myelography shows the low conus. Most cases show abnormalities in the myelogram but a normal view does not absolutely exclude this syndrome. Nowadays, computed tomography and magnetic resonance image are used for high diagnostic accuracy. Since Bassett, in 1950, it has been reported that untreated tethered cord syndrome aggravated the neurological signs, and many authors demonstrated the importance of the early detection of the disease and a preventive operation before the neurologic signs. So the importance is given to an earlier diagnosis when deficits are less severe and improvement is more likely.

This case demonstrated that a neurotrophic ulcer may occur rare occasions be caused by tethered cord syndrome. A dermatologist or pediatrician may be the first physician to see the cutaneous manifestation of tethered cord syndrome in infant and early childhood. We therefore stress that this syndrome is an important clinical entity closely related to neurologic deficit, of which the dermatologist should be aware. Also, neurologist, urologist and neurosurgeon should follow up and treat these patients to prevent any future neurologic deficits.

REFERENCES