

# Adolescent Tethered Cord Syndrome : Type II Split Cord Malformation with Dermoid Cyst at Split Site

Anil Sharma, Vijay Raina.

## Abstract

Presented here is a case of fourteen year old female with diagnosis of Tethered Cord Syndrome (TCS) having type II Split Cord (Diplomyelia-D<sub>12</sub> to L<sub>2</sub> vertebral level) with dermoid cyst at the split cord site (L<sub>1</sub>-L<sub>2</sub> vertebral level). She presented with acute onset paraplegia with sphincteric involvement. She was having asymptomatic patent ductus arteriosus. After investigations, she was operated upon and has shown significant neurological recovery.

## Key Words

Tethering, Split-cord, Dermoid cyst, Paraplegia

## Introduction

Tethering of the spinal cord usually occurs when tip of the conus lies below the lower border of L<sub>1</sub> vertebral body [1]. Thickened fila and lipomas probably make up over 70% of all tethering lesions. Three other less common lesions are currently being critically reevaluated as to their embryogenesis, anatomic variations and clinical significance. Out of these three lesions viz. split-cord malformations (diastematomyelia, diplomyelia), myelomeningocele (spina bifida), and cervical myelomeningocele, the first one is being reported here.

## Case Report

A fourteen year old female presented with sudden weakness affecting lower limbs and loss of

bladder-bowel function of one week duration. Weakness started in her left leg followed, after two days, by total loss of motor function in lower limbs. The weakness was preceded by severe pain in the lower back which was non-radiating type. There was no history of fever, trauma or any strenuous physical activity in the past. She had a tuft of hair over her lower back since birth. On examination, she was a healthy young girl with normal higher mental functions and upper limbs. Lower limb examination showed complete flaccid paraplegia. There were equivocal lower abdominal reflexes. Loss of sensations was upto D<sub>10</sub>. She had no bladder-bowel sensation and was on indwelling catheter. Inspection of dorso-lumbar spine showed a tuft of hair at D<sub>11</sub>-L<sub>3</sub> level. No

From the Unit of Neurosurgery, Department of Surgery, Govt. Medical College, Jammu (J&K).

Correspondance to : Dr. Anil Sharma, Consultant Neurosurgeon, Department of Surgery, Govt. Medical College, Jammu (J&K).

scoliosis was seen. CVS examination showed continuous machinery murmur of Patent ductus arteriosus. Her haemogram and serum biochemistry was normal. CSF examination showed high proteins, normal sugar and no cells. Chest radiograph showed mild cardiomegaly. x-ray dorsal spine did not show any spur or split vertebral bodies. x-ray lumbar spine showed feature of spina bifida at L<sub>5</sub>-S<sub>1</sub> level.

**Myelogram** revealed split cord (two hemicords) seen from D<sub>12</sub> level downwards with a myelographic defect at L<sub>1</sub>-L<sub>2</sub> (Fig 1).

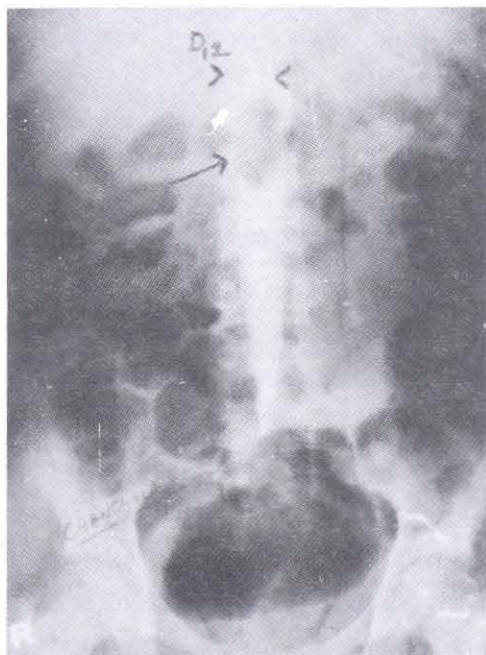


Fig. 1 Diplomyelia shown by two arrow heads and Dermoid cyst by a long arrow

**Myelo-CT** showed two hemicords having single dural investment and separate lateral sub-arachnoid spaces and a common midline sub-arachnoid space (Fig. 2). No bony spur could be demonstrated. At L<sub>1</sub> level, a mass was seen straddling both the cords and extending upto L<sub>2</sub> with obliteration of sub-arachnoid space (Fig. 3 & 4).



Fig. 2 Showing myelo-CT with diplomyelia and common midline subarachnoid space.



Fig. 3 Showing myelo-CT at lower level with intradural hypodense mass obliterating the subarachnoid space.

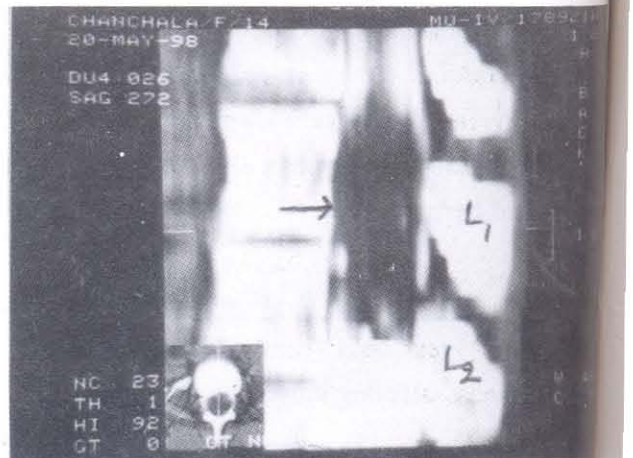


Fig. 4 Showing myelo-CT reconstruction with filling defect L<sub>1</sub>-L<sub>2</sub> level.



## Surgical procedure

The patient was taken up for surgery after preanaesthetic checkup and cardiology clearance. D<sub>12</sub>-L<sub>3</sub> laminectomy was done. From dura level onwards, whole procedure was done under magnification. Durotomy revealed two hemicords having no paramedian roots. The whole span of cord-split area (cranio-caudal) was occupied by a mass. Lower end of the mass was adherent to junction of two hemicords forming the conus. Aspiration of the mass revealed thick gelatinous fluid. Adequate care was taken to prevent spillage of fluid in the CSF. The separation of the cranial attachment of the mass revealed a significant sized artery in the axilla of cord bifurcation, which was coagulated. After the excision of the mass, the space between the two hemicords was seen to be covered with fibrous adhesions which were also lysed. No bony/cartilaginous spur was seen. Rootlets were normal in configuration and traversed their paths in normal direction. After achieving haemostasis, dura was closed in water tight fashion and incision closed in layers. Histopathological examination of the mass revealed features of dermoid cyst.

**Post operative period** was uneventful. Patient started showing neurological recovery in the first week. Stitches were removed after ten days and she was discharged.

**Follow up after three months** has shown that the girl has regained grade 5 power in her muscles at hip and knee joints and is able to sit independently. Her ankle dorsiflexors and plantar flexors are showing improvement. She has partly regained her bladder-bowel sensation and is on intermittent catheterization.

## Discussion

The normal flexion and extension motions of spine are associated with upward/downward movement of the cord. **Breig** has shown that neck flexion induces upward movement of the cord by as much as two cms. [2]. This associated mobility gets markedly restricted if the cord is tethered. Adult tethered cord syndrome (TCS) behaves differently from childhood TCS. Pain is almost always present and it is localized to perineum and back of legs [3]. It is dysesthetic, diffuse and bilateral [4]. Motor deficits in the form of frank leg weakness and sphincteric disturbances are common. Unlike childhood TCS, cutaneous stigmata of dysraphism are present only in less than 50 percent of affected adults. Symptoms get precipitated by many factors like trauma, manoeuvres associated with stretching of conus like extreme flexion-extension exercise, spinal stenosis etc. Type II split cord malformation is characterized by two hemicords with common dural covering. Medial surfaces of both cords are adherent to a midline fibrous partition which either inserts into dorsal dura or penetrates it to end in the tissues outside the spinal canal. This midline fibrous partition is the tethering agent.

Our case is unusual from many aspects. No bony/cartilaginous spur could be seen as an associated finding with cutaneous stigma of hypertrichosis. The main tethering agent was the dermoid cyst which because of its walls, being adherent to medial sides of both hemicords, was preventing the normal spinal cord movement during motion of the spine. Even minor accident or fall could result in sudden neurological deterioration, as happened in this case, because of relatively immobile split cords which were extremely vulnerable to forces of deformation when stretched. The pressure effect, because of voluminous



cyst, was maximum on the conus as evidenced by *slow recovery of sphincteric function* in follow up period. The artery coagulated in the axilla of bifurcation of the cord perhaps represented an anomalous vessel which persists as the only mesenchymal derivative between hemicords of Type II Lesion.

After going through the available English literature and to the best of our knowledge, this is the first case report of occult spinal dysraphic state in which a dermoid cyst is the main tethering lesion in a setting

of split-cord (diplomelia) leading to acute onset paraplegia.

#### References

1. Barson AJ. The vertebral level of termination of the spinal cord during normal and abnormal development. *J Anat* 1970; 106 : 489-497.
2. Breig A. Overstretching of and circumscribed pathological tension in the spinal cord : a basic cause of symptoms in cord disorders. *J Biomech* 1970; 3 : 7-9.
3. Pang D. Tethered cord syndrome in adults. In: Holtzman RNN, Stein BM (eds.) *The Tethered spinal cord* New York : Thieme-Stratton, 1985 ; pp 99-115.
4. Pang D., Wilberger JE Jr. Tethered cord syndrome in adults. *J Neurosurg* 1982 ; 57 : 32-47.

