BMH MEDICAL JOURNAL

BMH Med. J. 2020;7(3):59-74. **Review Article**

Tethered Cord Syndrome

Suresh S Pillai, Jayafar K, Sunil Paul Pathrose, Hari Sankar

Division Of Spine Surgery - Dept. Of Orthopedics, Baby Memorial Hospital
Calicut, Kerala, India

Address for Correspondence: Dr. Suresh S Pillai, MBBS, D. Ortho, MS (Ortho), DNB (Ortho), MNAMS, Spine Fellow (Chang-Gung University, Taiwan), AO Spine Fellow (Nottingham, UK), MCh (Ortho), Consultant Spine Surgeon, Baby Memorial Hospital, Kozhikode, Kerala, India. PIN: 673004. E- mail: sureshorth@googlemail.com

Abstract

Tethered cord syndrome can be due to various reasons. It is seen in patients with spina bifida occulta. The inelastic tight filum can cause neurological lesions like urinary and bowel incontinence, claudication, and persistent back pain varying with posture etc. Other lesions like ependymoma of the conus, epidermoid cysts, neureneteric cysts, lipoma of the filum, meningomyelocele, lipomeningomyelocele and even teratoma of the sacrococcygeal region can cause tethered cord syndrome. Timely identification and prompt correction give excellent results.

Keywords: Tethered cord syndrome, Intradural epidermoid, myelomeningocele, lipomyelomeningocele, ependymoma of the conus, fatty filum, lipoma of the filum

Introduction

A stretch induced functional disorder of the spinal cord with its caudal portion anchored by an inelastic structure is called tethered cord syndrome (TCS) [1]. The spinal cord is a soft structure with interlacing fibers of neurons glia and vasculature, supported by the pia matter prevents separation between these structures. Also, the dendate ligaments and dura mater in the cervical and thoracic cord level resist cephalad or caudad and traction exerted to the spinal cord [2,3]. When the spine grows rapidly with an inelastic filum no protection is offered by the dura and dendate ligaments, but the visco elasticity of the filum terminal prevents dysfunction of the spinal cord elements [4]. So the visco elasticity of the filum prevent the stretching of the cord during normal movements [5,6]. If the cord is stretched beyond certain limits lasting derangements in the spinal cord may ensue. Cord tethering can result in metabolic derangement and further histological changes, if stretched further. Various degrees of reduced state of cytochrome a, a3 implies impaired oxidative metabolism in these patients. A decrease in blood flow and glucose metabolic impairment is correlated with cytochrome reduction. Early untethering, when mild symptoms are present offers best treatment for TCS. Yamada et al have demonstrated improvement in neurological status and metabolism of the cells of the cord after untethering surgery. Irreversible neurological

damage is possible in chronically tethered cord when sudden stretch occurs. The current understanding is that TCS is caused by steady excessive tension in the lumbosacral cord accentuated by repeated stretching of the spinal cord by flexion or extension movements [7].

In the early stage of life, the bony spine grows faster than the spinal cord causing the spinal cord to effectively "ascend." The process is very rapid during 8-25 weeks of gestation. At birth the conus is located opposite to L2 [8]. It reaches the adult level at age two months post-natally [4,9]. There is evidence that the conus does not ascend further during childhood [10] and the conus terminates near the L1-2 disc space in the majority of normal individuals. Conus termination normally vary from the mid T12 to mid L3 level. Although 94-97.8% terminate above the L2-3 disc [11,12]. So it is wiser to consider termination of conus at L2-3 as normal at any age. A conus terminating at or below L3-4 is abnormal (except in premature infants and a full-term new borns). A fatty filum can be incidental finding and is not diagnostic of tethered cord [13] because it is present in 5.8% of normal individuals on postmortem examination [14]. Thickness of filum terminal is controversial. Some considers filum thickness more than 2 mm [15] as abnormal and some others consider filum diameter greater than 1 mm [16] as abnormal. In CT the fat appears as dark and as bright signal in T1 weighted MR sequences. Spina bifida occulta is seen more in patients with cord tethering [17]. Different studies show different association of spina bifida occulta and tethered cord syndrome. Patients with occult spinal dysraphism may have associated lesions such as congenital dermal sinus, lipomyelomeningocele, fibrous traction bands, and abnormal filum terminale, but it is not imperative to evaluate these patients for tethered cord syndrome unless they have neurological deficit [18]. So, classic neurological deficit suggestive of tethered cord syndrome needs modern imaging irrespective of the presence of spina bifida occulta in plain x-ray films.

Cord tethering associated with spinal bifida occulta

Low Cord Termination

Spinal cord exhibits antero-posterior and craniocaudal movement when the patient changes positions, unlike tethered spinal cord, which exhibits reduced or no movement [19]. Dynamic MRI can be used to assess this craniocaudal movement.

Thick Filum

Thickened and shortened filum may cause cord tethering. Fatty infiltration of the filum is a clue to cord tethering, being present in 29% of patients with thickened filum [15]. The filum takes the shortest course, tenting the dura posteriorly giving a triangular appearance to the thecal sac. MRI will show the posterior placement of the conus and the filum [20]. The filum appears as a small round filling defect extending through multiple sections in the MRI. The thick filum has brighter signal than CSF on T1 or proton density weighted MRI scans. Distal cord syrinx and caudal regression syndromes may be associated with cord tethering. An associated dermoid, lipoma, or rarely a sacral teratoma also may be present.

Dermal Sinus Tract

Dermal sinus tract seen in MRI as a subcutaneous line of decreased signal (relative to fat) on T1 weighted images. It may be associated with sacral and intraspinal lipomas [21].

Diastematomyelia

A congenital split in the spinal cord is termed diastematomyelia. There may be a fibrous, bony, or cartilagenous septum separating the two hemicords which are united above and

below the septum. This would cause tethering and traction of the cord. Two types of diastematomyelia are identified, one with single arachnoid and dural coverage of both hemicords and second one with separate dural and arachnoid covering of the hemicords, which unites above and below the septum. Each hemicord has dorsal and ventral nerve roots on its corresponding side. This may be associated with lipomyelomeningocele. It is called diplomyelia when there is complete duplication of the spinal cord, each having a central canal, two dorsal and two ventral horns and four segmental nerve roots at each level [15].

Lipomas

Lipomas associated with myelodysplasia are composed of fatty and fibrous elements. These lipomas are usually extradural, but occasionally can extend through a dorsal myeloschisis into the central canal and become intradural [22].

Dermoids

Skin and skin appendages may get entrapped in the spinal canal during lumbar puncture with unstyletted needles [23] or during embryonic development forming epidermoids or dermoid inclusion cysts.

Cord Tethering With Spina Bifida Cystica

Spina bifida cystica includes lesions like meningoceles, myeloceles, meningomyeloceles, lipomyelomeningoceles, and hemi myeloceles. All these lesions are known to cause cord tethering, but not in the purview of this article.

Signs & symptoms

Complaints of constant aching in the back and legs or in one leg, localized deep in the muscles. Pain increased by flexing and extending the back, movements that lengthen the spinal canal, i.e. any movement that decreases the lumbar lordosis can cause stretch in the cord by the inelastic filum.

Examples:
- Sitting cross legged in a Buddha position
- Bending over the dish or sink
- Holding baby at waist level
- Lying supine
- Slouching as in a chair

The back and leg pain will be expressed as a feeling of tearing, cramping, or pulling. The pain may be described to extend to the thoracic back muscles and rarely to the nuchal musculature. This is due to reflex contraction of the back muscles. Also, patients complain of numbness of the lower limbs while walking, bony pain in the legs and feet, difficulty in urination (80%), rectal incontinence (30%), difficulty in standing still. Maximum walking distance that has decreased over months to years. Walking that can cause numbness and urinary urgency. There wont be radicular pain down the legs and no aggravation of symptoms by coughing/sneezing, unlike patients with disc prolapse. In tethered cord syndrome (TCS) there may not be paravertebral muscle spasm and local tenderness over the lumbar spine. They may have foot or leg deformities.

Imaging studies

MRI shows cord elongation and filum thickening or posterior displacement of the filum, which signifies that filum travels along the concave side of the lumbar lordosis to minimize the spinal cord tension. Filum and conus touching the posterior arachnoid membrane is an important finding. The thickness of the filum is an inaccurate method of diagnosis. Diagnosis of TCS must rely primarily on the symptomatology and the specific imaging feature of posterior displacement of the conus and filum that attach to the posterior arachnoid membrane. A fibro-adipose filum, elongated spinal cord, lipoma attached to the filum tip thickened filum, capacious sacral sac [24] (in more than 50% of TCS patients). Tethered cord syndrome can occur in intradural epidermoid tumors, ependymoma of the conus, diastamatomylia, inelastic filum, myelomeningocele, lipomyelocele etc. Associated syrinx may be found in the spinal cord in these patients.

Differential diagnosis of tethered cord syndrome

		TCS	IVDP
1	Radiating Pain	-	+
2	Aggravated by coughing	-	+
3	Myotomal deficit	-	+
4	Dermatomal deficit	-	+
5	Paravertebral Spasm	-	+
6	Pain on Straight Leg raising Test	-	+
7	Pain Inguinal Or Genito-Rectal Area	+	-
8	Effect of 3B(Body) posture	+	-
9	Pain worse by slouching than sitting straight	+	-
10	Pain worse on lying supine	+	-
11	Incontinence	+	-
12	Musculoskeletal deformities	+	-

Indication for surgeries in adults with TCS

1. Pain aggravated by repeated flexion and extension of lumbar spine
2. Physical exercise that accentuate spinal curvature
3. Progressive motor and sensory change [25] with skipped myotomal, dermatomal deficit
4. Increasing lack of bowel or bladder control
5. Increasing lordosis or scoliosis or foot and toe deformities

Surgery

While positioning the patient keep adequate lumbar lordosis to avoid stretching of the cord.

Do not allow lumbar kyphosis while positioning to avoid post-operative neurological deficit. After laminectomy at L2/L3 intra-operative stretch test is an important step to confirm lack of filum elasticity before the filum is sectioned. Intra-operatively the filum is seen tight from the surrounding cauda equina fibers. Hold the filum with two pairs of forceps and pulling them in opposite directions. A fibrous or fibro-adipose filum barely stretches (less than 10%) in TCS patients whereas a normal filum stretches at least 80% of its original length. The filum is usually rotated as much as 90 degree and the anterior spinal vein is visible laterally. The filum is resected 1 cm apart at two points. (Electrical stimulation of nerve root and filum is advocated to identify the filum physiologically).

All the patients who underwent surgery for TCS were relieved of leg pain on awakening from anesthesia. These patients report increased ambulation and improved bowel and bladder function.

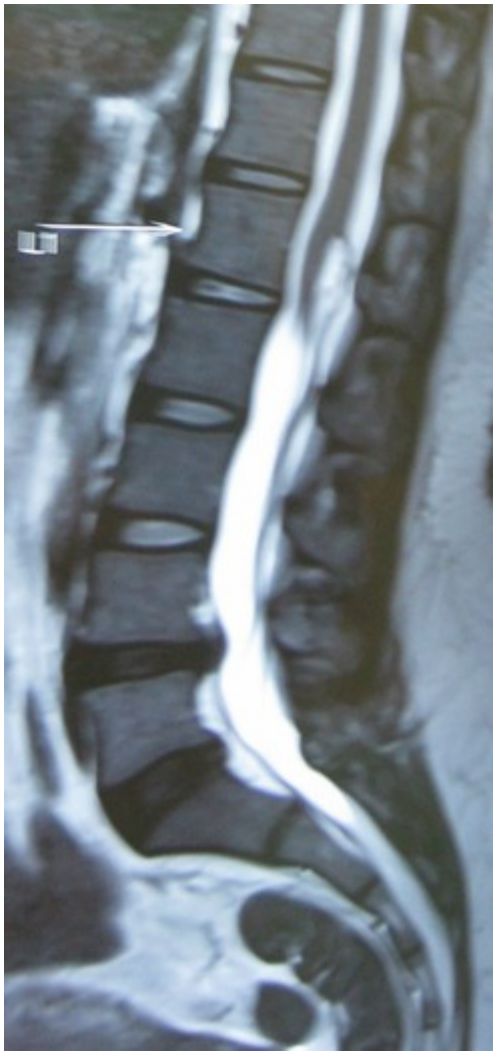

Figure 1: Young female with lipoma of the filum terminale

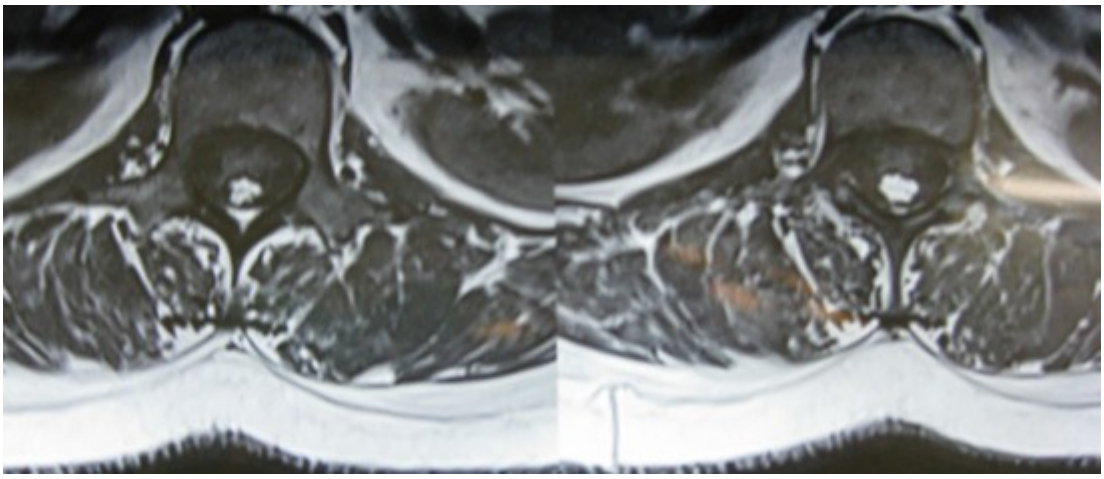

Figure 2: Young female with lipoma of the filum terminale - MRI-axial section

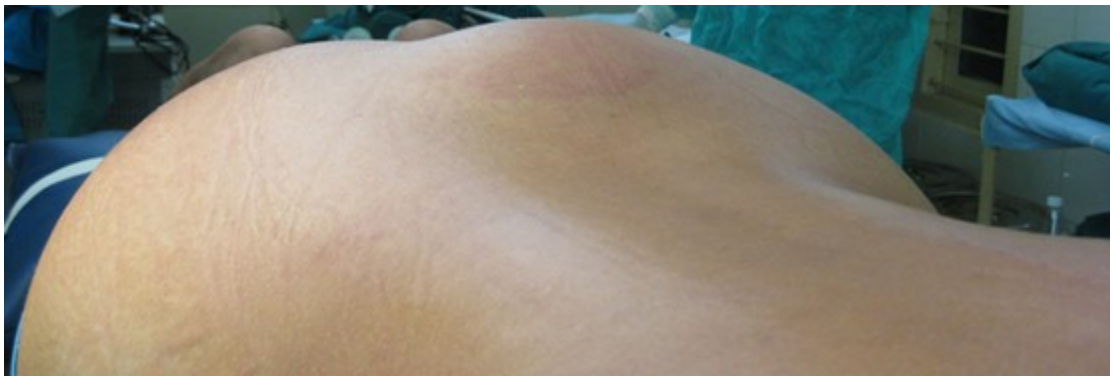

Figure 3: Lipoma of the filum terminale with a neavus at the LS area

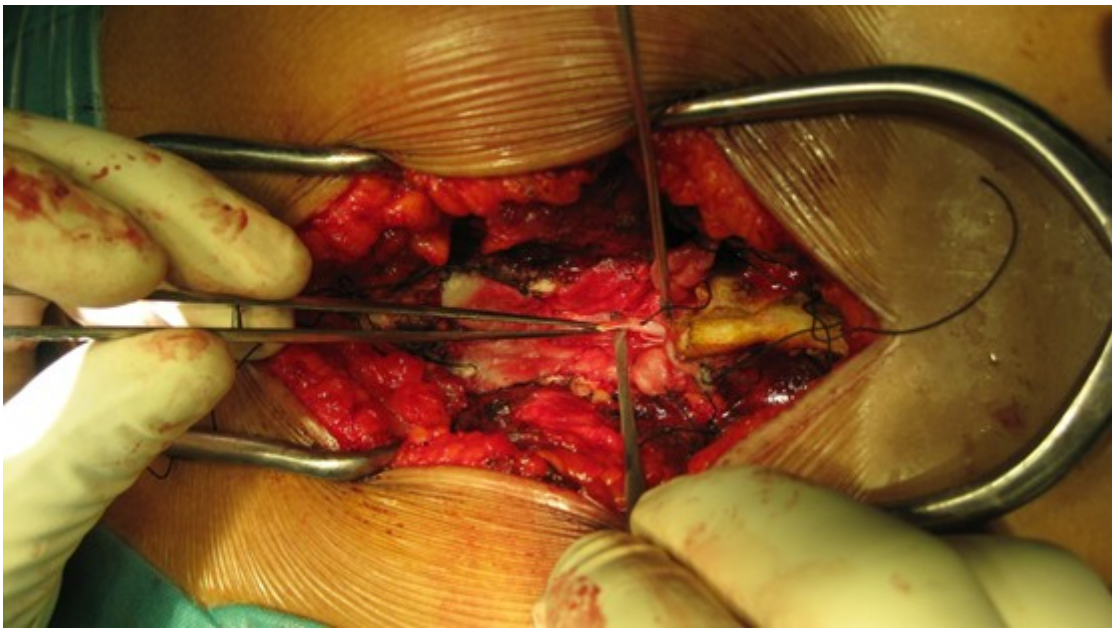

Figure 4: Isolated filum terminale - Intra operative picture

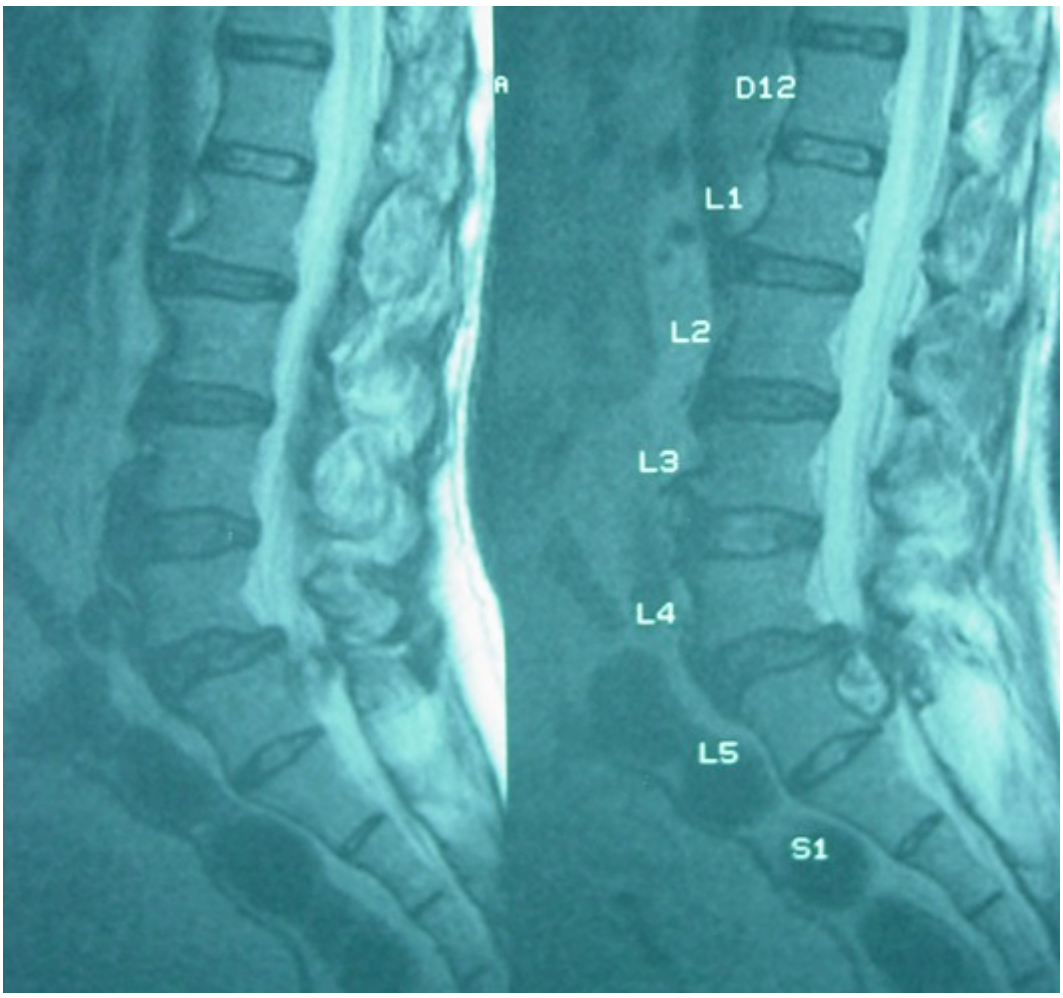

Figure 5: Neurenteric cyst with tethering of the cord

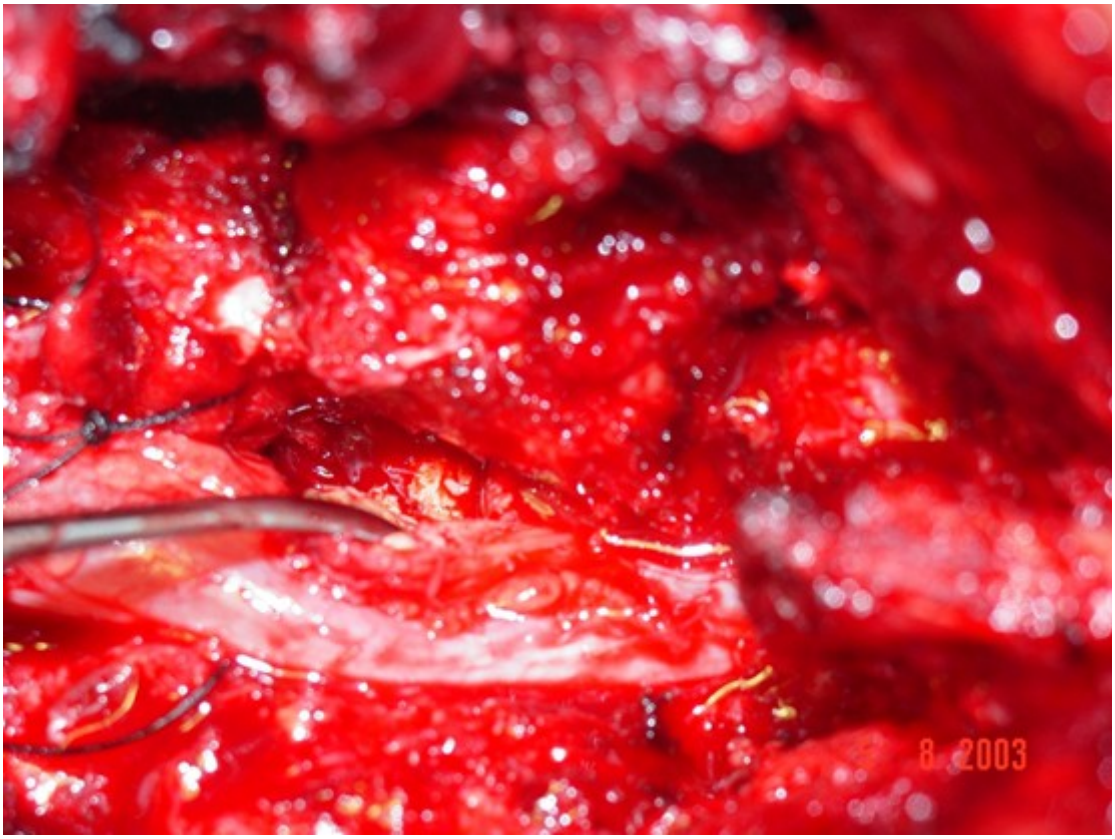

Figure 6: Intra operative picture - removal of the neurenteric cyst

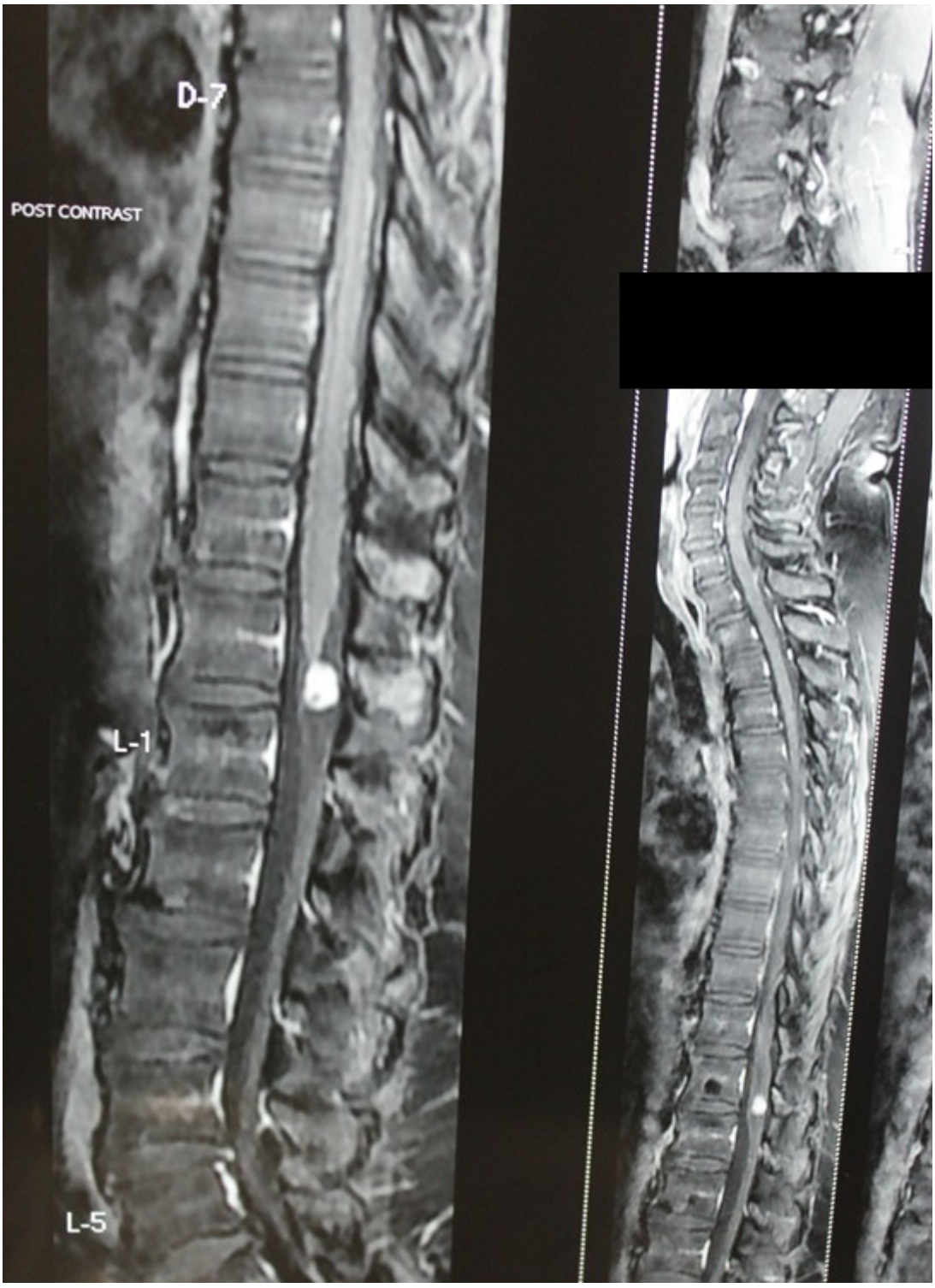

Figure7: Ependymoma of the conus causing tethering of the cord in an elderly person causing tethering - MRI

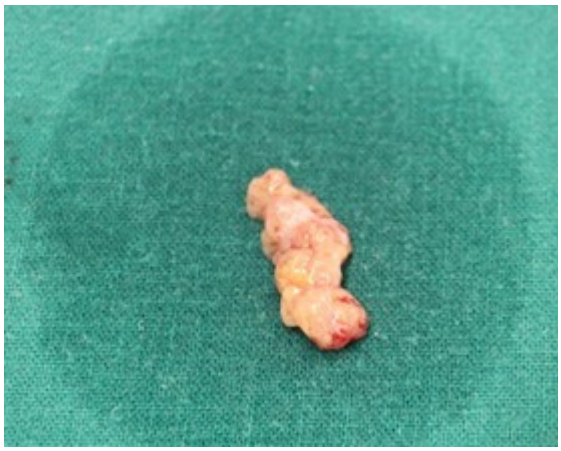

Figure 8: Removed ependymoma

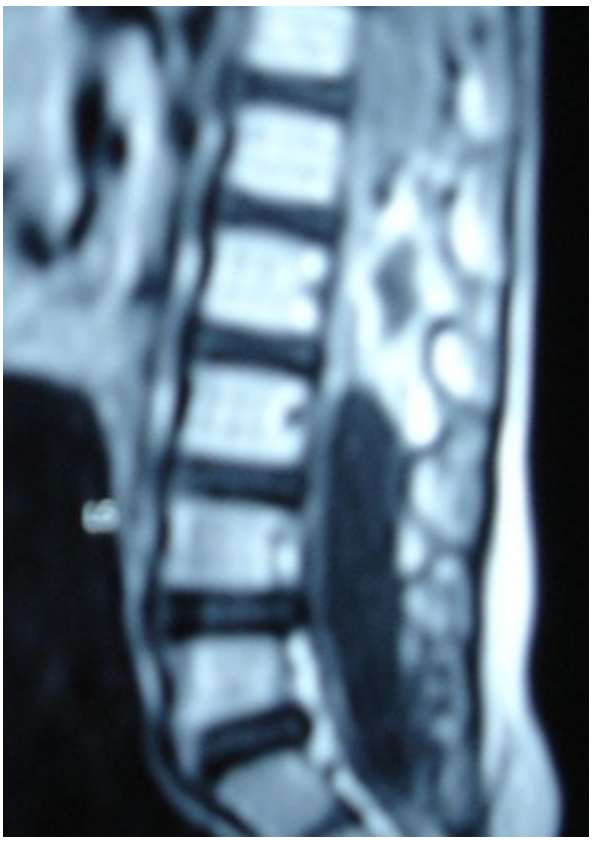

Figure 9: Intradural epidermoid in a toddler

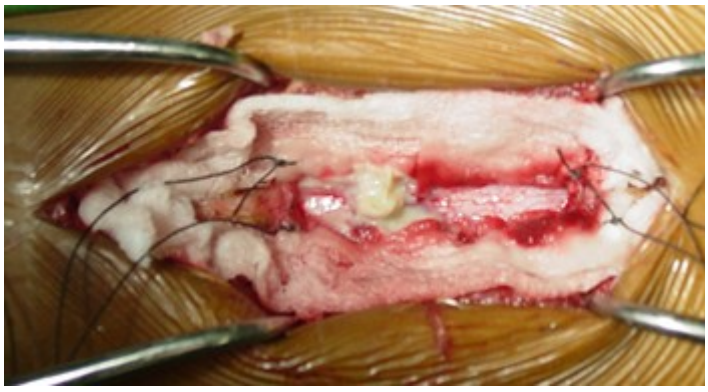

Figure 10: Infected epidermoid after durotomy

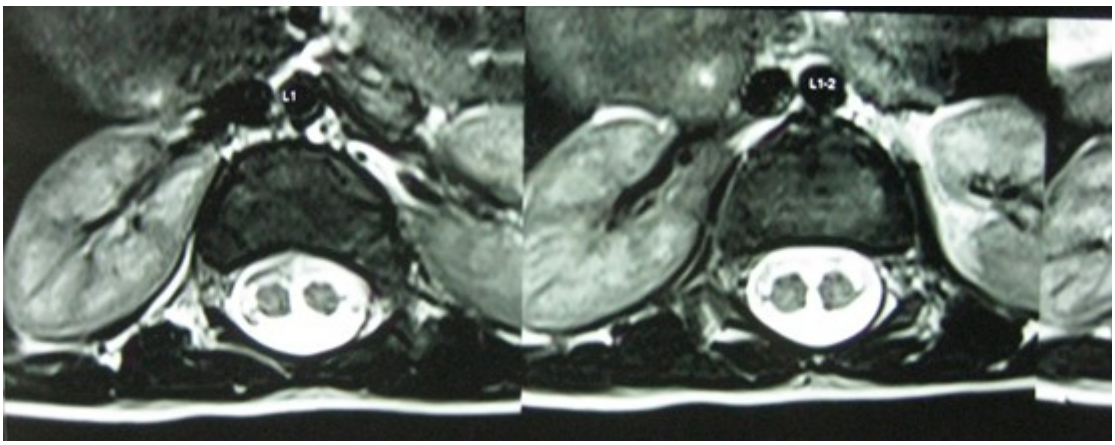

Figure 11: Type 2 diastamatomyelia in a child with tethering

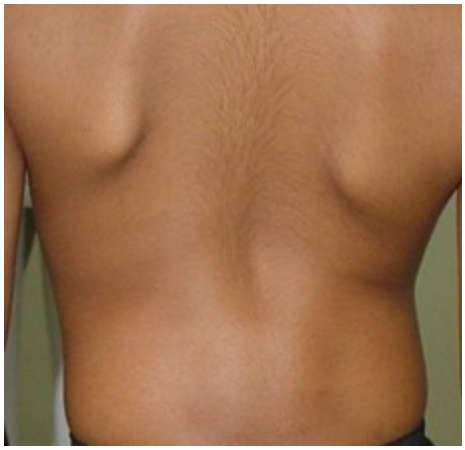

Figure 12: Increased lumbar lordosis in a young child with TCS

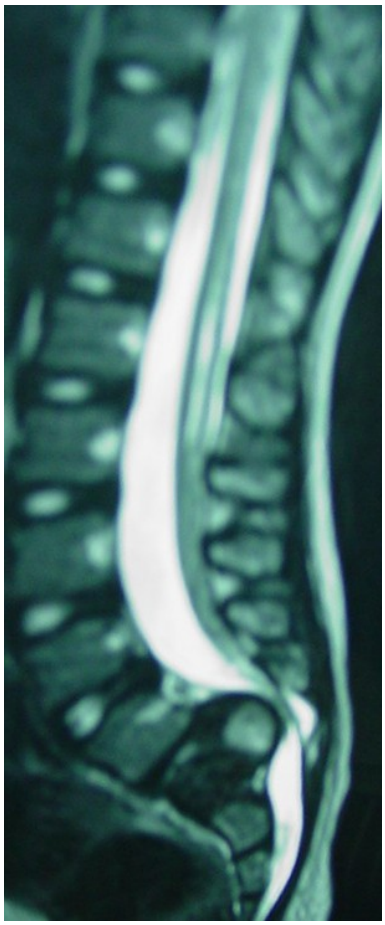

Figure 13: MRI showing hemivertebra, tight filum with syrinx above

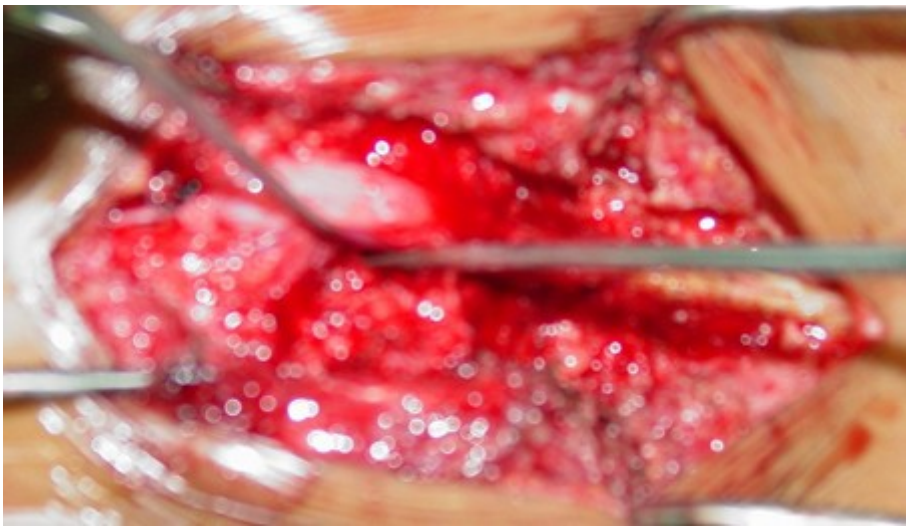

Figure 14: Resection of the hemi vertebra with de tethering of the cord

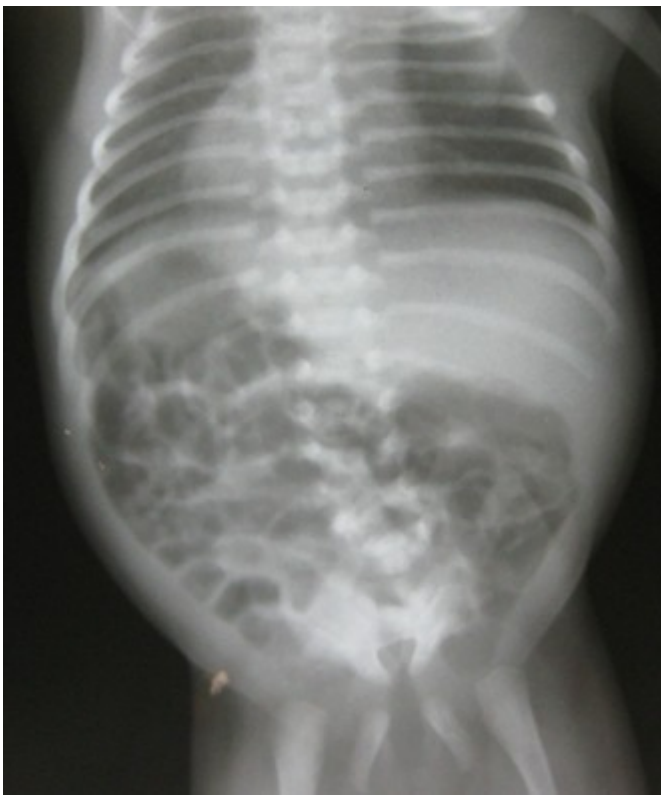

Figure 15: X-ray showing sacral agenesis

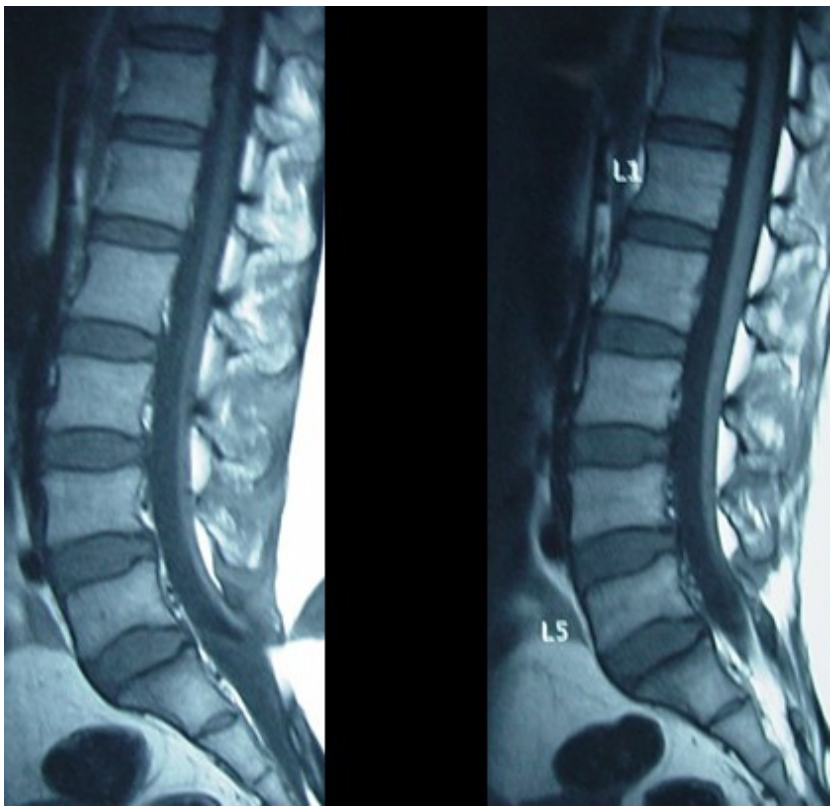

Figure 16: Myelomeningocele in a middle aged lady with TCS, urinary retention - a delayed presentation

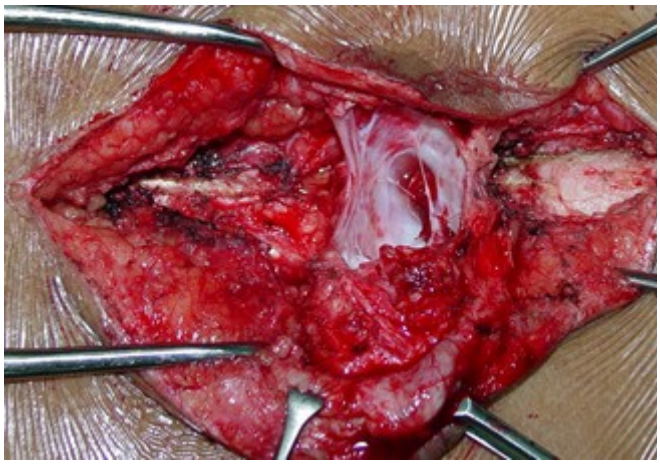

Figure 17: Intra operative picture showing isolation of the sac

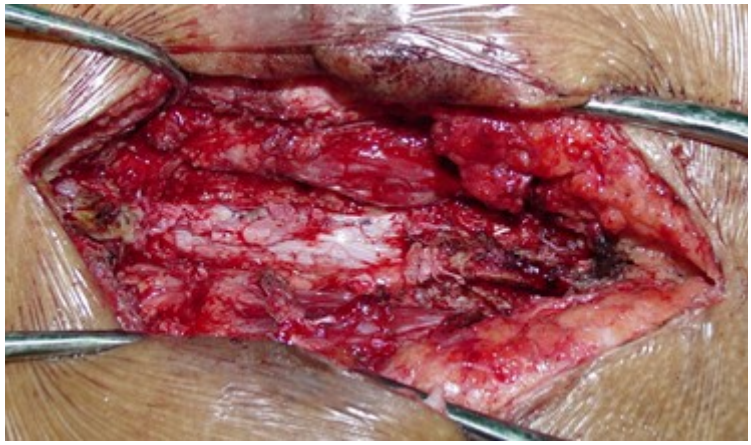

Figure 18: After dithering and repair of the sac and dura. Post operatively patient improved and the post void residual urine became 25 ml from 700 ml at 1 year

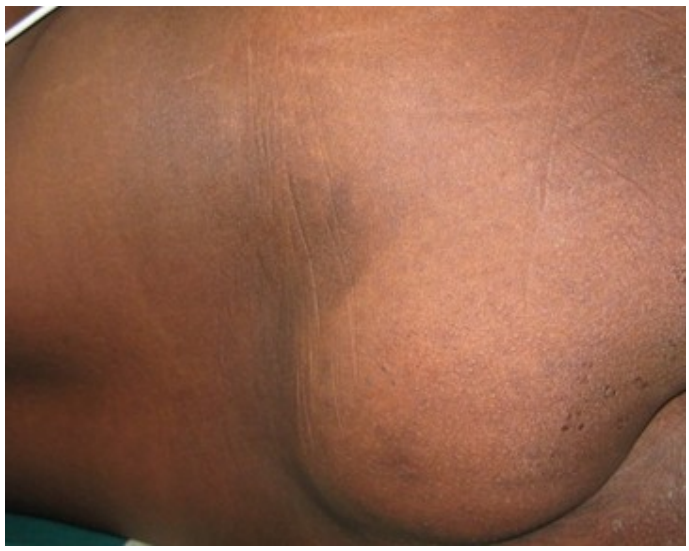

Figure 19: Lipomeningomyelocele in a young person with urinary retention

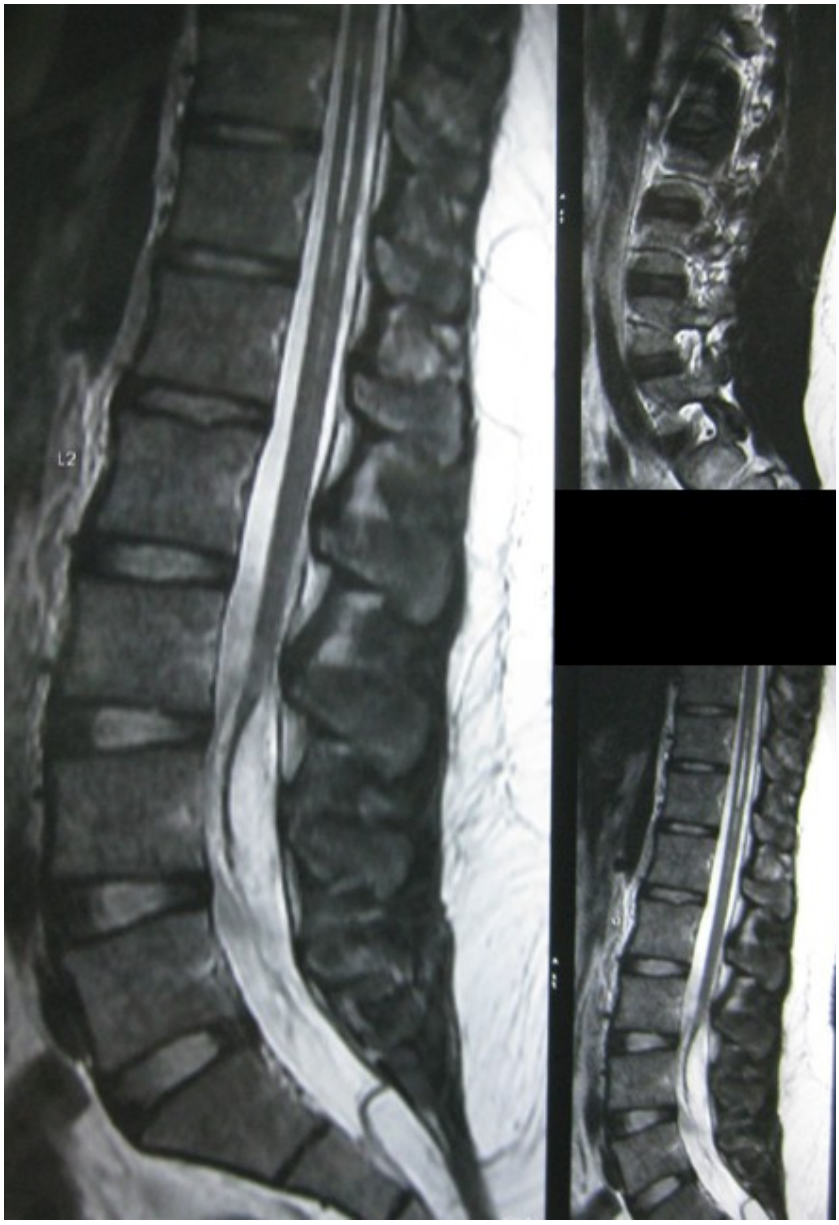

Figure 20: MRI showing lipomeningomyelocele, TCS and syrinx above

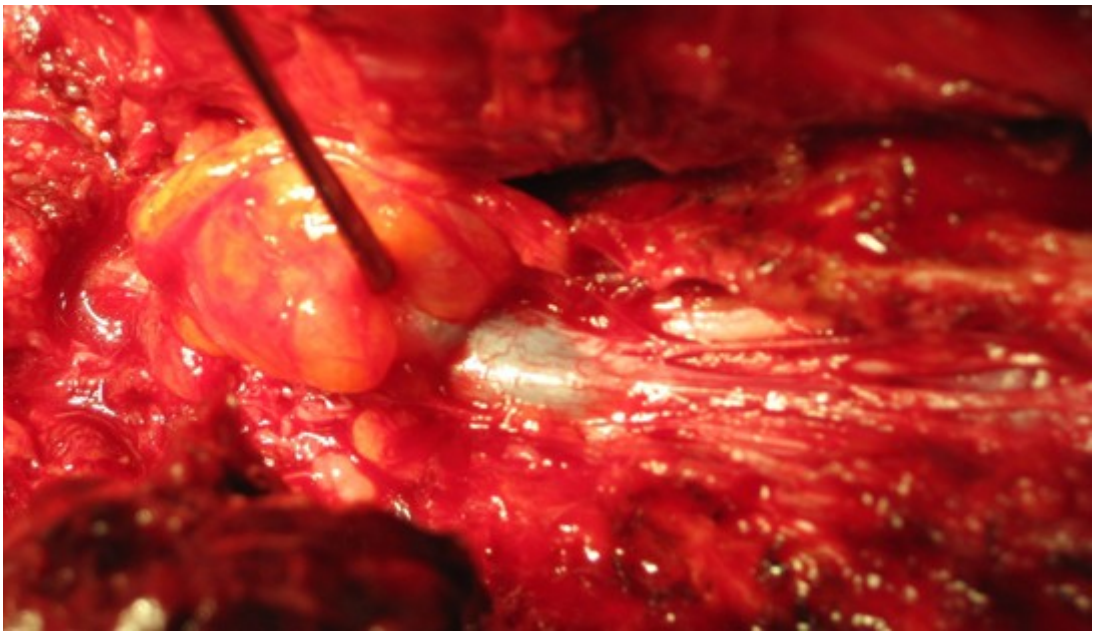

Figure 21: Intra operative picture showing removal of fat , dithering and repair

Conclusion

The filum diameter is 1.1 or 1.2 mm. The minimal filum diameter claimed to be diagnostic for tethered cord syndrome ranges from 1-2 mm. The main cause of TCS is traction effect of the inelastic filum. The diagnosis of tethered cord syndrome should be considered for any patient presenting with back or leg pain, scoliosis, neurogenic foot deformity, trophic foot ulceration or sphincter disturbance. This is especially so when the patient has spina bifida occulta or cutaneous skin markers like hairy patch, lipoma, hemangioma, skin dimple etc. Early evaluation and urgent intervention of these patients before neurological deterioration is recommended. Prophylactic untethering in asymptomatic patients are also advised, but the pros and cons of the surgery should be weighed against it. Significant neurological and symptomatic improvement is documented with timely intervention.

References

1. Hoffman HJ, Hendrick EB et al - The tethered spinal cord: its routine manifestations diagnosis, and surgical correction.Childs Brain 1976;2:145-155.

2. Tubbs RS, Salter G et al - The dendiculate ligament: anatomy and functional significance. J Neurosurg 2001;94(2,suppl):271-275.

3. Tunturi AR - Elasticity of the spinal cord, pia, and dendiculate ligament in a dog. J Neurosurg 1978;48:975-979.

4. Hawass LD, el-Badawi MG, Fatani JA et al - Myelographic study of the spinal cord ascend during fetal development. AJNR Am J Neuroradiol 1987;8:691-695.

5. Breig A - Over stretching and circumscribed pathological tension in the spinal cord: A basic cause of symptoms in cord disorders. J Biomech 1970;3:7-9.

6. Yamada S, Won DJ et al - Pathophysiology of tethered cord syndrome and similar complex disorders. Neurosurg Focus 2007;23:1-10.

7. Yamada S, Zinke DE et al - Pathophysiology of "Tethered Cord Syndrome" J Neurosurg

1981;54:494-503.

8. Kaufman BA - Neural tube defects. Pediatr Clin North Am 2004;51:389-419.

9. Barson AJ - The vertebral level of termination of spinal cord during normal and abnormal development. J Anat 1970;106(PT3):489-497.

10. Wilson DA, Prince JR et al - MR imaging determination of the location of the normal conus medullaris throughout childhood. AJR Am J Roentgenol 1989;152:1029-1032.

11. James CCM, Lassman LP - Spinal dysraphism: spina bifida occulta, New York: Appleton - Century – Crofts;1972.

12. Rayman AF, Anson BJ - Vertebral level of termination of spinal cord with report of a case of sacral cord. Anat Rec 1944;88:127-138.

13. McLendon RE, Oakes WJ et al - Adipose tissue in the filum terminal - a computed tomographic finding that may indicate tethering of spinal cord. Neuro surgery 1988;22:873-876

14. Emory JL, Lendon RG - Lipomas of the cauda equina and other fatty tumors related to neurospinal dysraphism. Dev Med Child Neurol Suppl 1969;20:62-70.

15. Naidich TP, McLone PG - Congenital pathology of the spine and spinal cord. In:Taveras JM, ed.Radiology. Philadelphia:JB Lippincott;1986:1-23.

16. Hochhauser L, Chuang S et al - The tethered cord syndrome revisited(abstract) AJ NR Am J Neuroradiol 1986;7:543.

17. Hendrick EB, Hauffman HJ et al. The tethered spinal cord. Clin Neurosurg 1983;30:457-463.

18. Anderson FM - Occult spinal dysraphism:A series of 73 cases. Pediatrics 1975;55:826-835.

19. Witkamb TD, Vandertop WP et al - Medullary cone movement in subjects with a normal spinal cord and in patients with a tethered spinal cord. Radiology 2001;220:208-212.

20. Yamada S, Won DJ et al - Adult tethered cord syndrome: New classification correlated with symptomatology, imaging pathophysiology. Neurosurg Q 2001;11:260-275.

21. Shokei Yamada et al - Tethered cord syndrome in children and adults. Second edition AANS January 12 2010.

22. Naidich TP, McLone DG et al - A new understanding of dorsal dysraphism with lipoma (lipomyeloschisis): Radiologic evaluation & surgical correction. AJRAm J Roentgenol 1983;140:1065-1078.

23. Pillai S, Cherian A T. Iatrogenic Intraspinal Epidermal Inclusion Cyst with Secondary Infection: A Cause for Acute Paraparesis. Journal of Orthopaedic Case Reports 2018 Nov-Dec; 8(6): 96-99.

24. Pang D, Wilberger JE Jr - Tethered cord syndrome in adults J Neurosurg 1982;57:32-47.

25. Austin GM - Dermatomes, sclerotomes and spinal cord relationship in IN:Austin GM, ed the spinal cord third ed New York: Igaku-Shoin:1983:727-750.