Tethered Cord Syndrome with Lumbo-sacral Subcutaneous and Intradural Lipoma: An Adult Presentation

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Summary:

Tethering of spinal cord can occur from a number of causes such as myelomeningocele, lipomeningocele, lipoma, dermoid, epidermoid, spina bifida occulta, split cord syndrome etc. Usually occur at childhood but rarely can present in adult age. We report this rare adult case as it presented with combination of all clinical syndromes associated with tethered cord syndrome where tethered spinal cord extended downward up to second sacral level.

Key Words : Adult Tethered Cord . Lipoma . Lumbo-sacral.

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Introduction:

Adult tethered cord syndrome is less frequent and rare. Clinical presentation may be in the form of neurological, urological, orthopedic, cutaneous syndromes or in any combination of these forms. The common causes are thick short filum terminale, meningomyelocele, lipomeningocele, epidermoid, dermoid etc. The main goal of surgical treatment of these patients is to halt the neurological deterioration with some expectation for improvement.[2,4,5,8,9,11] Adult tethered cord presenting with combination of all clinical syndromes is very rare. For this reason we report the case.

Case Report:

A 25 years old young lady presented with low back pain for 8 years, more marked for last three years with bilateral intractable sciatica which made her almost home bound. There was progressive weakness and muscle atrophy in both legs for last three years. She also had decreased sensation in both legs, foots and perianal region; but there was no trophic changes (neuropathic ulcer) in lower extremities. She had extreme difficulty in initiation of micturition with increase frequency and occasional overflow incontinence. She also had some difficulty with defecation in opening bowel. Her back examination showed a large lumbo-sacral subcutaneous soft tissue mass measuring12x15cm, extending on both side of midline mostly on the left side. There was no spinal deformity like kyphosis or scoliosis. There was right sided foot drop. Both leg and foot muscles were atrophied, and muscle power was 3+/5 in both leg muscles. Muscle power above both knee was normal with normal bulk. Both ankle jerks were absent and planter reflexes responded less. Anal tone was reduced but reflex was present. All modalities of sensation were grossly diminished in L4,L5&S1 and mild to moderately diminished in S2,S3&S4 dermatome in both sides. There was bilateral pes cavus. Patient could walk with extra effort .On abdominal examination bladder was full but kidney ballottement was negative. MRI of lumbo-sacral spine showed, dilatation of spinal canal at lumbo-sacral region, spinal cord extension up to S2 with an intradural lipoma from distal cord to S2 extending to subcutaneous tissue through the spina bifida (Figure1A,B,C). X-ray lumbo-sacral spine showed spina bifida at L5,S1&S2 with widening of spinal canal (Figure1D) Ultrasonogram showed significant postvoidal residual urine with moderate hydronephrosis (bilateral).

Untethering of the cord was done by removing subcutaneous, intraspinal extradural portion of lipoma along with most of the portion of intradural lipoma (a portion of lipoma attached with cord was left in situ) and by releasing short thick and tight filum terminale.

After two months of the operation, back pain and sciatica were relieved completely. Sensory function improved significantly but did not become normal (in both lower limb).Bowel problem also subsided. Her bladder control improved significantly but still she had some difficulty

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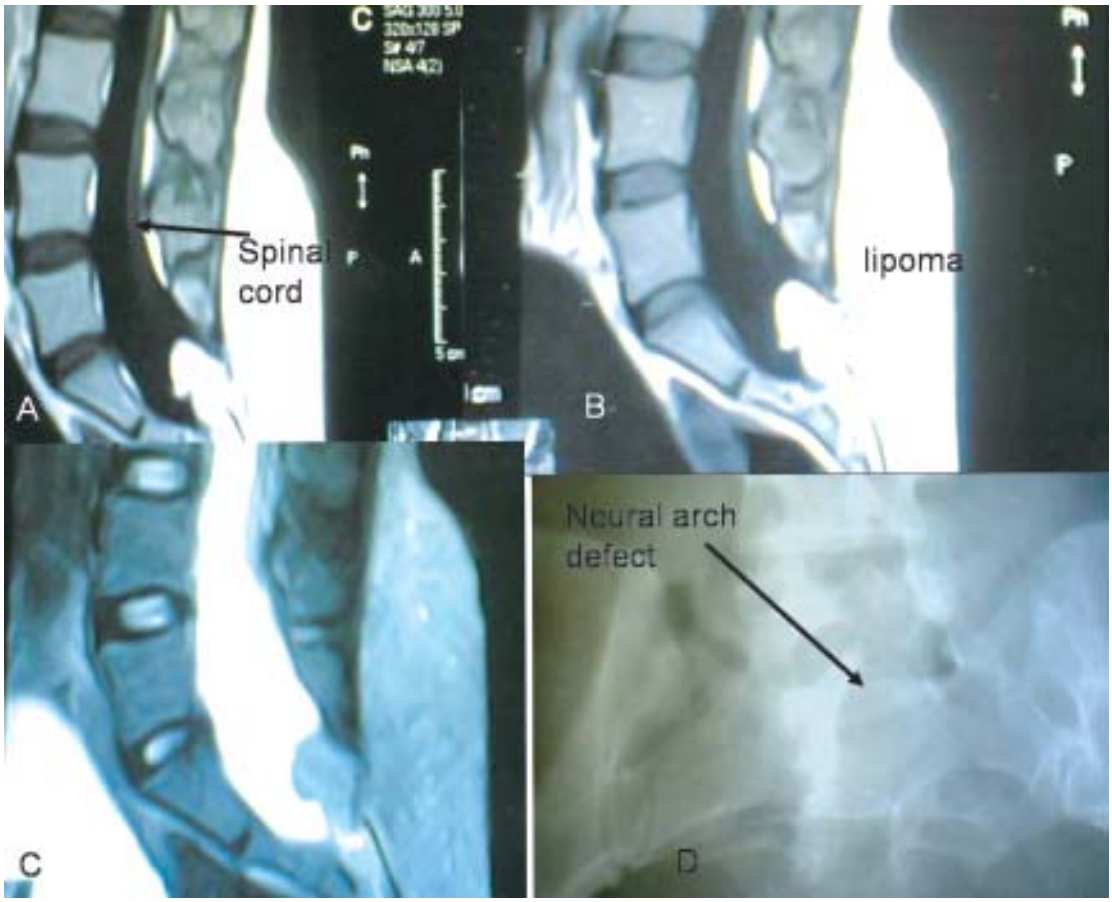

Fig.-1: *MRI of lumbo-sacral spine,A&B-T1 weighted saggital image, C-T2 weighted image showing low attached spinal cord with intra and extradural lipoma, D-Plain x-ray lumbo-sacral spine showing neural arch defect*

in emptying the bladder. Kidneys were normal in size and post voidal residual urine became insignificant on ultrasonography. Her motor function remained unchanged in both lower limbs.

Discussion:

The tethered spinal cord syndrome is most often encountered in children, but does also occur in adults .Its clinical spectrum comprises low back pain, sciatica, neurodeficit such as distal motor weakness and trophic & sensory changes in the legs (neurological syndrome), urological symptoms (urological syndrome) and musculoskeletal signs as scoliosis or foot deformities(orthopedic syndrome).[2,4,5,7,9] In addition, cutaneous lesions(cutaneous syndrome) or subcutaneous lipomas in the lumbo-sacral may be indirect signs of intraspinal pathology.[5] Intraspinal pathology includes a tight, thickened and some times shortened filum terminale ,an intraspinal lipoma, intradural scar formation or other lesion that lead to conus fixation.[4,7,9] The common mechanism of injury of these type of pathologies is an impairment of longitudinal movement of the spinal cord, especially the conus medullaris, which subsequently leads to chronic local ischaemia.[8] Diagnosis is most readily achieved by MRI.[6] Treatment is aimed at the restoration of cord mobility by means of microsurgical release of the conus, the cauda equina and the filum terminale. Further progression of neurological deficit can be effectively halted by microsurgical release; in fact almost half of the patient actually improve including urological symptoms.[4,6,9,10] Early diagnosis and adequate surgical release are the keys to success in tethered cord syndrome. Outcome of surgical release of tethered cord in children is very good.[1,11] In adult , result is not like that of children. During operation intraoperative monitoring like lower

extremity somato-sensory evoked potential, pudendal sensory evoked potential, external anal sphinter manometry and EMG can be used for prevention of iatrogenic further neurological deterioration.[3] Intradural arachnoid bands and fibrous stalks may attach to the filum terminale, lipoma, and conus medullaris, that may confuse the surgeon and all these should be cut for adequate release with preservation of rootlets and conus medullaris.[11]

In our case, though the patient had cutaneous syndrome from birth, she did not present in her childhood; rather she presented in her adult life typically with back pain and sciatica which is more common in adult type of tethered cord syndrome.2[1] She typically presented with all symptoms associated with tethered cord syndrome i.e. cutaneous syndrome, neurological syndrome, orthopedic syndrome and urological syndrome. MRI confirmed the diagnosis and lower end of cord seen at the sacral level; with such type of MRI finding, it is strange why the patient did not become symptomatic in her childhood .When the patient was referred to us late as the patient developed neurological deficit. We operated on her for prevention of further neurological deterioration. The tethering elements were removed by microsurgical operation. Post operatively with in short period of time patient improved significantly, though her motor function remained static.

Conclusion:

Every patients presenting with the clinical diagnosis of tethered cord syndrome, should be investigated properly and offered specialized microsurgical treatment to prevent neurological deterioration and in many cases, for practical hope of neurological improvement.

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