Tethered Cord Syndrome in Adulthood

Symptoms related to a congenital tethered cord occur most commonly in childhood, so it was initially regarded as a pediatric problem; but in many patients, the diagnosis is not established until symptoms manifest in adulthood.

The number of adults in whom congenital TCS is diagnosed continues to grow as a result of better imaging and recognition of this syndrome. Pediatric TCS has been well studied in the literature, but much of the information regarding the adult population is still being defined. Patients who never undergo treatment for TCS likely have an elevated risk of developing symptoms with advancing age [1)].

Epidemiology

Adult tethered cord syndrome is a rare neurological disorder that classically presents with back or leg pain, weakness, and urinary dysfunction. Spinal cord tethering has been associated with acquired Chiari malformations.

Evaluation

Radiographically: low conus medullaris (below L2) and thickened filum terminale. NB:apparent filum terminale diameter on CT myelogram may vary with concentration of contrast material.

Preoperative cystometrogram is strongly recommended, especially if the patient seems continent (postoperative changes in bladder function are not uncommon, possibly due to stretching of the lower fibers of the cauda equina).

Differential diagnosis

It is difficult to differentiate a tethered cord from a congenitally low lying conus (filum diameter is generally normal in the latter).

Treatment

Standard treatment for TCS diagnosed in adulthood remains controversial. Surgical intervention is usually indicated based on an expected natural history of disease progression in the absence of treatment.

Some adults with TCS decline surgery despite severe neurologic deficit [2)].

Surgical treatment

If the only abnormality is a thickened, shortened filum terminale, then a limited lumbosacral laminectomy may suffice, with division of the filum once identified.

If a lipoma is found, it may be removed with the filum if it separates easily from neural tissues.

The filum is differentiated from nerve roots by presence of characteritics squiggly vessel on surface of filum. Also, under the microscope, the filum has a distinctively whiter appearance than the nerve roots, and ligamentous-like strands can be seen running through it. NB: intra-op electrical stimulation and recording of anal sphincter EMG are more definitive.

Technique

In the series of Gao et al. all patients received general anesthesia and took their prone position, neural electrophysiological monitoring electrode were then placed, followed by the acquisition and collection of muscle electromyography signals from the anal sphincter, bilateral musculus vastus lateralis, gastrocnemius and mesothenar. A total of 72 cases applied positive straight incision, 10 cases of lumbosacral lipoma with longitudinal incision. After exposing the dura mater spinalis, it was cut from the normal anatomical structure to the lesion. Cauda equina was managed by sharp releasing adhesion under the nerve electrophysiological monitoring, tumors were removed with the use of medical ultrasonic dissector. After the tumor was removed, the dura mater spinalis with low tonus was closed by water, and the dura mater spinalis with high tonus was formed by the autogenous fascia. For patients combined with subcutaneous giant lipoma in the lumbosacral region, the subcutaneous tumor was removed, and the drainage tube was placed into the left empty cavity, followed by pressurized dressing and vacuum aspiration [3].

Outcome

Surgical release is usually good for pain relief. However, it is poor for return of bladder function.

Results of clinical studies of surgical intervention in adulthood are encouraging [4]. [5] [6].

It is safe and effective for improving pain and neurological status in the majority of patients; however, patients who have undergone previous intradural detethering procedures in general fare less well, and considerable judgment is required in their management [7].

In a multivariate regression model, laminectomy, bladder dysfunction when associated to muscular weakness, and long-term (>6 months) symptoms were selected as the independent risk factors associated with poor or minimally improved (almost unchanged) surgical outcomes. When the urodynamic test showed overactive detrusor muscle, no improvement was recorded in postoperative urodynamic test. Laminoplasty (or hemilaminectomy), short-term (<6 months) symptoms, patients without lipomas, and presentation with moderate or mild symptoms seem to be proper predictors for good surgical outcomes. Further prospective studies are necessary to investigate these findings systematically. Urodynamic study can be used as a predictive tool for close follow-up of asymptomatic adult patients involved with TCS [8].

Case reports

A 68-year-old man with a history of distant T12-level spinal cord injury who presented with two weeks of progressive bilateral lower extremity weakness. The patient underwent a T12-L1 laminectomy in 1977, complicated by arachnoiditis and syringomyelia, with eventual placement of a syringo-pleural shunt. He remained neurologically stable until 2012, when he underwent a suboccipital craniectomy for Chiari decompression for new-onset headache and dysphagia. Ten days later, the patient noted progressive leg weakness and radiographic evidence of spinal cord tethering at the T11-T12 level. A T10-L1 laminectomy and medical facetectomy was undertaken for detethering with postoperative recovery of ambulatory function with assistance.

The patient presented with an unusual acquisition of tethered cord syndrome. The tethering of the spinal cord may have been triggered by arachnoid adhesions from initial lumbar surgery 35 years prior to presentation and subsequently exacerbated by alterations of CSF dynamics following Chiari decompression. Given the potentially devastating sequelae of tethered cord syndrome, investigation of CSF flow dynamics may be beneficial prior to operative intervention in patients with risk factors for a tethered cord who present with adult-onset Chiari malformation [9].

[1], [4]
Rajpal S, Tubbs RS, George T, Oakes WJ, Fuchs HE, Hadley MN, Iskandar BJ. Tethered cord due to spina bifida occulta presenting in adulthood: a tricenter review of 61 patients. J Neurosurg Spine. 2007 Mar;6(3):210-5. PubMed PMID: 17355019.

[2]
Düz B, Gocmen S, Secer HI, Basal S, Gönül E. Tethered cord syndrome in adulthood. J Spinal Cord Med. 2008;31(3):272-8. PubMed PMID: 18795476; PubMed Central PMCID: PMC2565560.

[3]
Gao J, Kong X, Li Z, Wang T, Li Y. Surgical treatments on adult tethered cord syndrome: A retrospective study. Medicine (Baltimore). 2016 Nov;95(46):e5454. PubMed PMID: 27861396; PubMed Central PMCID: PMC5120953.

[5], [7]
Lee GY, Paradiso G, Tator CH, Gentili F, Massicotte EM, Fehlings MG. Surgical management of tethered cord syndrome in adults: indications, techniques, and long-term outcomes in 60 patients. J Neurosurg Spine. 2006 Feb;4(2):123-31. PubMed PMID: 16506479.

[6]
van Leeuwen R, Notermans NC, Vandertop WP. Surgery in adults with tethered cord syndrome: outcome study with independent clinical review. J Neurosurg. 2001 Apr;94(2 Suppl):205-9. PubMed PMID: 11302621.

[8]
Abdallah A, Emel E, Abdallah BG, Asiltürk M, Sofuoğlu ÖE. Factors affecting the surgical outcomes of tethered cord syndrome in adults: a retrospective study. Neurosurg Rev. 2018 Jan;41(1):229-239. doi: 10.1007/s10143-017-0842-z. Epub 2017 Mar 14. PubMed PMID: 28293750.

[9]
Jackson C, Yang BW, Bi WL, Chiocca EA, Groff MW. Adult tethered cord syndrome following Chiari decompression. World Neurosurg. 2018 Jan 31. pii: S1878-8750(18)30208-0. doi: 10.1016/j.wneu.2018.01.165. [Epub ahead of print] PubMed PMID: 29409774.