Kyphoscoliosis correction with recurrent tethered cord syndrome due to myelomeningocele and neurofibromatosis type 1

Rachana Tyagi, Smit Shah

ABSTRACT

Introduction: Recurrent tethered cord syndrome (TCS) is a neurosurgical condition that is associated with spinal dermoid cysts, myelomeningoceles and lipomyelomeningoceles most commonly. However, recurrent TCS due to myelomeningocele and neurofibromatosis type 1 (NF1) has not been reported.

Case Report: We describe a case of a 13-year-old boy with recurrent TCS and progressive spinal deformity with a severe kyphoscoliosis due to myelomeningocele and NF1. We also review the anterior and posterior surgical approaches to vertebrectomies along with their indications and contraindications. Finally, we also discuss specific risks of scoliosis repair in NF1 patients due to high probability of pseudoarthrosis.

Conclusion: We conclude that in patients with recurrent TCS, posterior fusion with shortening and without detethering was a successful procedure which should be considered as a primary surgical option. Anterior fusion may be delayed in such patients and used only if necessary given the additional risks in such medically fragile patients.

International Journal of Case Reports and Images (IJCRI)

International Journal of Case Reports and Images (IJCRI) is an international, peer reviewed, monthly, open access, online journal, publishing high-quality, articles in all areas of basic medical sciences and clinical specialties.

Aim of IJCRI is to encourage the publication of new information by providing a platform for reporting of unique, unusual and rare cases which enhance understanding of disease process, its diagnosis, management and clinico-pathologic correlations.

IJCRI publishes Review Articles, Case Series, Case Reports, Case in Images, Clinical Images and Letters to Editor.

Website: www.ijcasereportsandimages.com

CASE IN IMAGES

PEER REVIEWED | OPEN ACCESS

Kyphoscoliosis correction with recurrent tethered cord syndrome due to myelomeningocele and neurofibromatosis type 1

Rachana Tyagi, Smit Shah

ABSTRACT

Introduction: Recurrent tethered cord syndrome (TCS) is a neurosurgical condition that is associated with spinal dermoid cysts, myelomeningoceles and lipomyelomeningoceles most commonly. However, recurrent TCS due to myelomeningocele and neurofibromatosis type 1 (NF1) has not been reported. **Case Report:** We describe a case of a 13-year-old boy with recurrent TCS and progressive spinal deformity with a severe kyphoscoliosis due to myelomeningocele and NF1. We also review the anterior and posterior surgical approaches to vertebrectomies along with their indications and contraindications. Finally, we also discuss specific risks of scoliosis repair in NF1 patients due to high probability of pseudoarthrosis. **Conclusion:** We conclude that in patients with recurrent TCS, posterior fusion with shortening and without detethering was a successful procedure which should be considered as a primary surgical option. Anterior fusion may be delayed in such patients and used only if necessary given the additional risks in such medically fragile patients.

Keywords: Neurofibromatosis type 1, Pseudoarthrosis syringohydromyelia, Recurrent tethered cord syndrome, Vertebrectomy

How to cite this article

Tyagi R, Shah S. Kyphoscoliosis correction with recurrent tethered cord syndrome due to myelomeningocele and neurofibromatosis type 1. Int J Case Rep Images 2017;8(12):826–833.

Article ID: Z01201712CI10022RT

doi: 10.5348/ijcri-201704-CI-10022

INTRODUCTION

Recurrent tethered cord syndrome (TCS) is a progressive neurological and orthopedic condition that occurs due to recurrent fixation or tethering of the distal spinal cord after an initial detethering procedure in pediatric and adult patients [1]. It develops due to scar tissue adhering to the spinal cord after an initial detethering procedure for spinal cord anomalies including: dorsal lipomas, congenital spinal dermoid cyst or sinus tract and myelomeningoceles and lipomyelomeningoceles [2]. As the child grows, there is a progressive stretching of the spinal cord which can lead to worsening neurological and orthopedic conditions along with deterioration of blood supply to the spinal cord.

Based on animal studies, Yamada et al. have demonstrated TCS with a tight filum to be a stretch induced functional disorder at the lumbosacral spinal cord that occurs due to failure of oxidative metabolism at the level of the mitochondria due to marked reduction in cytochrome *a* and *a3* which possibly results in abnormally inelastic filum [3]. As a result, ‘...abnormal inelastic filum

Rachana Tyagi¹, Smit Shah²

Affiliations: ¹Rutgers Robert Wood Johnson University Hospital, Pediatric Neurosurgeon, Department of Neurosurgery, New Brunswick, New Jersey, USA; ²Rutgers Robert Wood Johnson Medical School, Medical Student Year 3, New Brunswick, New Jersey, USA.

Corresponding Author: Smit Shah, 112 Montgomery Street, Apt 3G, Highland Park, New Jersey, USA - 08904; Email: spshah1991@gmail.com

Received: 14 September 2017

Accepted: 03 October 2017

Published: 01 December 2017

interferes with normal cord ascension and results in low lying conus medullaris...’ which lies below the level of L1 and L2 [4]. Thus symptoms are increased with activity, particularly flexion and extension which increases tension on the distal cord. It is possible that similar mechanisms of spinal cord dysfunction are likely in play with recurrent tethered cord syndrome due to more extensive tethering lesions.

Clinical manifestations of recurrent TCS vary between pediatric and adult patients. Unlike adult patients, pediatric patients usually complain of poorly localized diffuse pain in the lower extremities and perineum, or back mostly confined to the lower back with infrequent radiation to legs [5]. Neurological symptoms include bowel/bladder dysfunction, weakness and lethargy with activity, exacerbating gait and balance problems, numbness or paresthesia along with frequent bruising. Spinal signs/symptoms include muscle spasms, tenderness on palpation, scoliosis and kyphosis with possible truncal decompensation. The lower extremities may show deformities or atrophy, commonly worse on one side, with weakness and sensory loss on examination. Urodynamics may be the most sensitive test showing minor changes consistent with neurogenic bladder even without obvious physical findings [6]. Magnetic resonance imaging scan will show recurrent tethering at the site of the previous surgery. A proximal syrinx may also be noted, or arachnoiditis of the cauda equina.

Neurofibromatosis 1 (NF1) is an autosomal dominant syndrome including café-au-lait lesions, Lisch nodules, intracranial tumors, peripheral neurofibromas, capillary hemangiomas, dural ectasia, scoliosis and other bony abnormalities. Despite its dominant genetic inheritance, spontaneous mutations of NF1 gene commonly occur. Due to variable genetic expressivity, severity of symptoms varies. Anterior sacral neural tube defects and neurofibromatosis type 1 are closely associated [7]. Even though, neurofibromatosis type 1 (NF1) is rarely associated with tethered cord, it is highly associated with dural meningocele [8, 9]. Scoliosis is relatively common in these patients, and surgery is often indicated. The poor bone quality associated with this disease increases the risks of pseudoarthrosis and often requires more aggressive fusion techniques or repeated procedures [10].

Surgery for repeated untethering has lower success rates with increased risk of complications, particularly new neurologic deficits. For pediatric patients with meningomyelocele and a Gibbus, possible treatment options include kyphectomy of the vertebrae responsible for kyphosis. This results in improved seating balance as well as restoration of an intact skin envelope by decreasing the incidence of pressure sores at the apex. For patients with severe kyphoscoliosis, a combined anterior and posterior spinal reconstruction can be used. This approach allows for maximal exposure, enhanced mobilization of the spinal elements, safety, and efficiency when operating around the spinal cord.

Such an approach facilitates posterior compression and anterior distraction, thereby allowing for greater correction and control [11].

Shortening the spine also allows for significant deformity correction without performing an initial detethering procedure, but still reducing the risk of a new neurologic deficit due to manipulation of the neural elements [11].

CASE REPORT

The patient was a 13-year-old boy with a history of de novo NF1 and thoracolumbar myelomeningocele. He had a progressive kyphoscoliosis with a large Gibbus complicated by pressure ulcers over the Gibbus as well as the sacrum. The patient also had a past medical history of Chiari Type II malformation with tethered cord, syringohydromyelia and hydrocephalus shunted at birth. He had significant difficulty with positioning in his chair and chronic back pain. He did have 4/5 iliopsoas function, but no significant function distally, with contractures of the knees and ankles. He also had worsening neurogenic bladder symptoms associated with left sided hydronephrosis.

We can see severe progression of scoliosis deformity as the age progresses (Figures 1–3). In Figure 1, when patient was nine years and eleven months old, there was severe right convex 20 degree curve from L1 to L3 which

Figure 1: Right convex 20 degree curve from L1 to L3 with level hips in sitting position; age nine years and eleven months.

progressed to severe 85 degree curve from T9 to L3 with pelvic tilt at age 12 years and 5 months (Figure 3). In addition, preoperative imaging at age 12 demonstrated significant kyphosis in the thoracolumbar area with holocord syrinx and thinned neural tissue with minimal soft tissue covering at closure site (Figure 2).

Patient's mother expressed significant reservations regarding detethering prior to deformity correction due to the risk of loss of hip flexion. He was currently able to assist with diaper changes by flexing his hips. Therefore, a detethering procedure was not planned, but instead a vertebrectomy to shorten the spine and prevent stretch on the tethered cord after deformity correction.

The initial procedure was placement of a halo head frame, with upright traction for one week. Minor improvement in the scoliosis at age 12 years 11 months was noted from 85–78 degrees, with no change in the neurologic examination (Figure 4). Therefore, posterior releases were performed with Ponte osteotomies from T6 to L2 and pedicle screw placement from T5 to L4, and the patient again placed in traction for two weeks,

with further decrease in the deformity to 45 degrees (Figure 5). As adequate correction was not obtained with these measures, a hemivertebrectomy from the posterior approach was then performed at the vertex of the curve at T11, and the posterior fusion was performed with successful correction of the deformity and no loss of hip function, extending the fusion to the pelvis with iliac bolts with a final coronal curve of 16 degrees along with good sagittal balance (Figure 6 and Figure 7). He was maintained in a rigid TLSO clamshell brace for three months postoperatively to decrease the risk of pseudoarthrosis, with a plan for possible further anterior surgery if needed.

Follow-up studies at 41 months show excellent fusion with no loss of correction, and the patient had developed no further pressure ulcers, back pain, or change in lower extremity function. As we can see in Figures 8–11, there was no loss of correction two years postoperative with 15 degree curve (Figure 8), 41 months postoperative at age 16 years 5 months showing successful fusion of facets bilaterally (Figure 9) and no loss of correction along with mild kyphosis at age 18 years and 9 months (approximately six years status post initial surgery) (Figure 10).

Figure 2: Preoperative magnetic resonance imaging scan at age 12 depicts significant kyphosis in the thoracolumbar area; holocord syrinx and thinned neural tissues with minimal soft tissue covering at closure site seen.

Figure 3: Anteroposterior and lateral images showing (A) Right convex 85 degree curve from T9 to L3, pelvic tilt age 12 years and 5 months, (B) Forward sagittal balance along with a forward tilt before traction.

Figure 4: (A) Initial upright traction with minimal improvement in curve to 78 degrees age 12 years and 11 months, (B) After 48 hours, curve decreased to 60 degrees.

Figure 5: Further improvement in curve after posterior release with Ponte osteotomies to 45 degrees with continued traction.

DISCUSSION

The thoracolumbar spine can be accessed anteriorly, posteriorly or in combination [12, 13].

Anterior access [7] is indicated in spinal conditions like kyphosis, scoliosis, lordosis, compression fracture or dislocation of the vertebral body, degenerative disc disease, thoracolumbar disc herniation, pyogenic or parasitic infection of spine and malignancy involving primary tumor of the vertebral body. The retroperitoneal

Figure 6: (A) Corrected pelvic tilt and Gibbus reduction after final hemivertebrae resection and fusion with posterior instrumentation eight days after final surgery, and (B) Final coronal curve 16 degrees, good sagittal balance on lateral film.

Figure 7: (A) Postoperative lateral view showing correction in sagittal imbalance; no residual Gibbus and (B) Postoperative image showing pedicle screws placement from T5 to L4; Significant reduction in scoliosis; correction of coronal balance; leveled pelvis seen in seated position post hemivertebrectomy.

area is accessed after the abdominal organs are retracted anteriorly. The discectomy above and below and the vertebrectomy/hemivertebrectomy is performed with placement of graft and possible intervertebral instrumentation with lateral plating to provide stability to fuse the vertebral column rostrally and caudally.

Thus, an anterior approach vertebrectomy reduces the risk of spinal cord or dural injury during the vertebrectomy, by eliminating the need for retraction and allows for greater sagittal deformity correction by allowing a larger lordotic cage to be placed without risk of neurologic injury. Anterior stability of the construct with a true 360 degree fusion construct also reduces the risk of pseudoarthrosis and crankshafting in the young patient. However, as compared to posterior approach, morbidity due to respiratory failure is more common with anterior approach [14]. A shortening procedure also cannot be performed without resection of the posterior elements, and thus would actually require

Figure 8: No loss of correction at second year postoperative with 15 degree curve.

Figure 9: Coronal computed tomography scan showing successful fusion of facets bilaterally at 41st months postoperative; age 16 years and 5 months.

a posterior laminectomies/facetectomies prior to the anterior fusion.

The posterior approach [15, 16] is more commonly used for fusion and stabilization of the vertebral column as compared to the anterior approach that is reserved for anterior column reconstruction and vertebrectomy. Since, the hemivertebrae in the thoracolumbar region deviates dorsolaterally with a kyphotic deformity, it can be identified posteriorly without significantly increased risk of neurologic injury. After the vertebral disc and endplate in both the cranial and caudal adjacent segments are curetted, bone chips obtained during the vertebrectomy can be inserted into the defect rather than a cage to shorten the spinal column, but with less correction of the kyphosis. A long construct with extensive deformity correction is possible with the posterior approach using osteotomies at the facets to further mobilize the segments [17]. Posterior only approach does have higher recurrence

Figure 10: (A) No loss of correction noted at 5.5 years postoperative at age 18 years and 7 months, and (B) Mild Kyphosis noted without any loss of correction at 5.5 years post op at age 18 years and 7 months.

rates of vertebral fractures along with contraindications like fragmented fractures of thoracolumbar spine [6, 7] for example: burst fractures which occurs due to vertical compression of the spine.

Scoliosis correction in NF1 patients carries increased risks relative to other causes of deformity. Crawford et al. demonstrated increased risk of pseudoarthrosis, paraplegia and intraoperative hemorrhage with complex spinal surgeries specifically in patients with NF1. Furthermore, dystrophic spinal deformities like scoliosis, kyphosis or kyphoscoliosis are challenging to manage postoperatively because they are rapidly progressive in nature and result in pseudoarthrosis very quickly. Decortication, abundant autogenous bone grafting, and segmental instrumentation are necessary to minimize the occurrence of pseudoarthrosis [11]. In addition, during the anterior approach, it is extremely important to resect all the pathologic disc soft tissue in order to prevent the resorption in the mid portion of the graft material due to incomplete resection. Finally, even if rigid fixation has been achieved using modern instrumentation, orthotic immobilization is recommended until a fusion mass with trabecular pattern is seen in CT scan performed six

months after surgery [18]. In these patients, an anterior and posterior construct for increased construct rigidity is recommended.

For the current patient, the risk of new neurologic deficit with loss of hip function was a significant concern, and would have adversely affected his mother's ability to provide care for the patient, likely necessitating two care givers for diaper changes as he grew, and further limiting his ability to reposition himself with increased risk of pressure ulcers. Correction of the deformity without detethering placed him at risk for increased stretch on the placode with likely new neurologic deficit. However, due to the holocord syrinx and severely thinned placode, the repeat detethering procedure itself would likely have led to permanent neurologic deficits from manipulation of the neural elements. Thus, a shortening procedure was planned if traction alone did not provide adequate deformity correction.

This case was discussed with a group of experienced spine surgeons with a consensus of proceeding with staged surgeries with initial posterior fusion only, and a delayed anterior fusion at sixth month if necessary. A step-wise escalation of deformity correction was followed with initial awake vertical traction in his wheelchair to allow for continuous monitoring of his neurologic function and to limit the risk of surgery. The initial surgical procedure of only osteotomies and placement of screws was then performed. Inadequate correction determined the hemivertebrectomy was required, with excellent coronal and sagittal balance and no loss of function. Subsequent imaging showed excellent fusion of the posterior columns with no loss of correction, and no need for anterior stabilization.

CONCLUSION

In this patient with recurrent tethered cord and a severe kyphoscoliosis due to myelomeningocele and NF1, posterior fusion with shortening and without detethering was a successful procedure. This technique may be considered as a primary surgery in similar patients. Although scoliosis correction in NF1 is known to have an increased risk of pseudoarthrosis, and therefore an anterior fusion was considered, our patient did have a successful posterior-only procedure. Anterior fusion may be delayed in such patients and used only if necessary given the additional risks in these medically fragile patients.

Author Contributions

Rachana Tyagi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Smit Shah – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None

Conflict of Interest

Authors declare no conflict of interest.

Copyright

© 2017 Rachana Tyagi et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES

1. Shih P, Halpin RJ, Ganju A, Liu JC, Koski TR. Management of recurrent adult tethered cord syndrome. *Neurosurg Focus* 2010 Jul;29(1):E5.
2. Caldarelli M, Boscarelli A, Massimi L. Recurrent tethered cord: Radiological investigation and management. *Childs Nerv Syst* 2013 Sep;29(9):1601–9.
3. Yamada S, Knerium DS, Mandybur GM, Schultz RL, Yamada BS. Pathophysiology of tethered cord syndrome and other complex factors. *Neurol Res* 2004 Oct;26(7):722–6.
4. Bui CJ, Tubbs RS, Oakes WJ. Tethered cord syndrome in children: A review. *Neurosurg Focus* 2007;23(2):E2.
5. Rajiah P, Shabani A. *Pediatric Radiology: Clinical Cases*. Cheshire: PasTest; 2008. p. 384–5.
6. Chou FH, Ho CH, Chir MB, Linsenmeyer TA. Normal ranges of variability for urodynamic studies of neurogenic bladders in spinal cord injury. *J Spinal Cord Med* 2006;29(1):26–31.
7. Hsien H, Ming T, Yang S, et al. Anterior approach for surgery of thoracolumbar spine: Surgical outcomes of series of one self-trained neurosurgeon. *Formosan Journal of Surgery* 2013;46(5):157–65.
8. Rengachary SS, Wilkins RH. *Neurosurgical Operative Atlas*. 2ed. New York: The American Association of Neurological Surgeons; 2007. p. 257–8.

9. Swetz KM, Spinner RJ. Large intrathoracic meningocele associated with neurofibromatosis type 1. *Mayo Clin Proc* 2009 Sep;84(9):769.
10. Tsirikos AI. Development and treatment of spinal deformity in patients with cerebral palsy. *Indian J Orthop* 2010 Apr;44(2):148–58.
11. Dalgic A, Uckun O, Acar HI, et al. Single-stage posterolateral Corpectomy and circumferential stabilization without laminectomy in the upper thoracic spine: Cadaveric study and report of three cases. *Turk Neurosurg* 2010 Apr;20(2):231–40.
12. Mayer S, Weisser M, Till H, Gräfe G, Geyer C. Congenital myelomeningocele - do we have to change our management? *Cerebrospinal Fluid Res* 2010 Oct 14;7:17.
13. Singh K, Samartzis D, An HS. Neurofibromatosis type I with severe dystrophic kyphoscoliosis and its operative management via a simultaneous anterior-posterior approach: A case report and review of the literature. *Spine J* 2005 Jul–Aug;15(4):461–6.
14. Nadir A, Sahin E, Ozum U, Karadag O, Tezeren G, Kaptanoglu M. Thoracotomy in spine surgery. *Thorac Cardiovasc Surg* 2008 Dec;56(8):482–4.
15. Nakamura H, Matsuda H, Konishi S, Yamano Y. Single-stage excision of hemivertebrae via the posterior approach alone for congenital spine deformity: Follow-up period longer than ten years. *Spine (Phila Pa 1976)* 2002 Jan 1;27(1):110–5.
16. Crocker M, James G, Ibrahim A, Thomas N, Chitnavis B. Posterior approach vertebrectomy in the thoracolumbar spine with expandable cage reconstruction: Indications and techniques based on eight cases. *Br J Neurosurg* 2008 Apr;22(2):235–40.
17. Crawford A, Lykissas M, Schorry E, et al. Neurofibromatosis: Etiology, commonly encountered spinal deformities, common complications and pitfalls of surgical treatment. *Spine Deformity* 2012;85–94.
18. Solmaz I, Izci Y, Albayrak B, et al. Tethered cord syndrome in childhood: Special emphasis on the surgical technique and review of the literature with our experience. *Turk Neurosurg* 2011;21(4):516–21.

SUGGESTED READING

- <https://rarediseases.org/rare-diseases/tethered-cord-syndrome/>
- Greenberg MS. *Handbook of Neurosurgery*. 7ed. New York: Thieme; 2010.
- Lew SM, Kothbauer KF. Tethered cord syndrome: An updated review. *Pediatr Neurosurg* 2007;43(3):236–48.

Access full text article on
other devices

Access PDF of article on
other devices

Edorium Journals: An introduction

About Edorium Journals

Edorium Journals is a publisher of international, high-quality, open access, scholarly journals covering subjects in basic sciences and clinical specialties and subspecialties.

Invitation for article submission

We sincerely invite you to submit your valuable research for publication to Edorium Journals.

Why should you publish with Edorium Journals?

In less than 10 words: "We give you what no one does".

Vision of being the best

We have the vision of making our journals the best and the most authoritative journals in their respective specialties. We are working towards this goal every day.

Exceptional services

We care for you, your work and your time. Our efficient, personalized and courteous services are a testimony to this.

Editorial review

All manuscripts submitted to Edorium Journals undergo pre-processing review followed by multiple rounds of stringent editorial reviews.

Peer review

All manuscripts submitted to Edorium Journals undergo anonymous, double-blind, external peer review.

Early view version

Early View version of your manuscript will be published in the journal within 72 hours of final acceptance.

Manuscript status

From submission to publication of your article you will get regular updates about status of your manuscripts.

Our Commitment

Six weeks

We give you our commitment that you will get first decision on your manuscript within six weeks (42 days) of submission. If we fail to honor this commitment by even one day, we will give you a 75% Discount Voucher for your next manuscript.

Four weeks

We give you our commitment that after we receive your page proofs, your manuscript will be published in the journal within 14 days (2 weeks). If we fail to honor this commitment by even one day, we will give you a 75% Discount Voucher for your next manuscript.

Favored author program

One email is all it takes to become our favored author. You will not only get 15% off on all manuscript but also get information and insights about scholarly publishing.

Institutional membership program

Join our Institutional Memberships program and help scholars from your institute make their research accessible to all and save thousands of dollars in publication fees.

Our presence

We have high quality, attractive and easy to read publication format. Our websites are very user friendly and enable you to use the services easily with no hassle.

Something more...

We request you to have a look at our website to know more about us and our services. Please visit:
www.edoriumjournals.com

We welcome you to interact with us, share with us, join us and of course publish with us.

Edorium Journals: On Web

Browse Journals

CONNECT WITH US