Fecal incontinence as a predominant symptom in a case of multiply recurrent tethered cord: diagnosis and operative strategies

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Recurrent manifestations of tethered spinal cord after an initial operative intervention for a simple fatty filum terminale is fairly uncommon. The authors present the case of an unusual clinical course in which there were 3 distinct episodes of recurrence, each time presenting predominantly as fecal incontinence and resolving with operative intervention. Typical signs of tethering were absent on radiological evaluation, and operative intervention was based on clinical grounds. Intraoperatively, sacral nerve roots to the anal sphincter were found tethered to the filar stump with electrophysiological evidence of regained activity on disentanglement. To the best of the authors' knowledge, a similar clinical course or operative findings have not been reported.

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Tethered cord syndrome (TCS) may be associated with myelomeningocele, lipomyelomeningocele, and lumbosacral lipoma or fibrolipomatous filum terminale or may be secondary to arachnoiditis or post-surgical scarring following an initial operative approach. Rates of retethering after sectioning a fatty filum terminale are low.[15,16] Retethering may produce neurological symptoms through nerve root traction and ischemia or tension distributed through the spinal cord.[3,4,12] We present a case of simple tethered cord due to a fatty filum terminale with 3 distinct episodes of recurrence, each presenting with fecal incontinence and resolving with operative intervention. At the last procedure, a discrete sacral nerve root was recognized and individual nerve roots were separated from the filum before its eventual complete resection. Symptoms had not returned at the 9-month follow-up. This case represents a unique clinical course and interesting pathophysiology that, to the best of our knowledge, has not been reported.

Case Report

History and Examination

A healthy 4-year-old boy initially presented with severe constipation and abdominal pain concomitant with low-back and leg pain for which he was being managed with bowel regimens without improvement. Magnetic resonance imaging of the spine demonstrated a fatty filum terminale and tethered cord. The patient underwent tethered cord release at another institution, and his symptoms partially improved for 6 months before a recurrence of his initial symptoms.

First Recurrence

The patient's physical therapist noted new mild weakness of the left leg. Magnetic resonance imaging confirmed release of the filum terminale with the lipoma clearly disconnected, and the surgeon declined to reexplore. The child presented to our office at this time for a second opinion. We offered to perform an exploration at the original surgical

level because his symptoms were identical to those before the release and were becoming disabling. At surgery—a reexploration of the exact primary surgical site—multiple nerve roots were seen tightly adherent to the dorsal dura mater. Intradural neurolysis was performed, and the filum terminale was clearly free without any obvious traction.

Second Recurrence

The symptoms immediately resolved after the second operation, but 20 months thereafter, the patient returned with recalcitrant leg pain, foot pain, and fecal incontinence. Lumbosacral MRI at this time was suggestive of recurrent scarring due to an unnatural position of the distal end of the fatty filum, which appeared to be in opposition to the dorsal dura at a right angle (Fig. 1). We proceeded to surgically reexplore and found that the filum again had several nerve roots wrapped over it and appeared in direct opposition to the dura. The adhesions were arachnoidal, and the nerve roots were easily and completely dissected off the filum. The visible segment of the fatty filum was resected in an attempt to avoid further recurrence. Again, symptom resolution was immediate but not long-lived.

Third Recurrence

After a period of improvement, fecal incontinence returned 8 months later. At that time, the patient was in the second grade and the social ramifications of being unable to control his bowels were becoming a significant burden and psychosocial strain on the family. Magnetic resonance imaging at the time was not suggestive of any clear pathology. We deferred reexploration for some time given concerns over the short-lived effects of the previous interventions. However, after attempts at behavioral modification and gastroenterology and general surgery consultations, reexploration was approached for a third time. At surgery, the laminectomy was extended rostrally to involve T-12 and L-1 with the intention of achieving radical resection of the filum lipoma, particularly where it emerged from the conus at L-1. Dural opening revealed several nerve roots tethered to the filar stump and wrapped around the lipoma at awkward angles traveling rostral and then caudal, making an S-shaped turn (Fig. 2). We directly stimulated these nerve roots using the microstimulator at 0.2 and 0.3 mA, obtaining robust signals from electrodes placed in the left and right anal sphincters, as well as the right extensor hallucis longus muscle. The nerve roots were completely microsurgically disentangled from the filum, and the remainder of the lipoma was resected up to the conus (Fig. 3). Noteworthy were the improved response and the strength of the signals obtained from stimulation of the same nerve roots after untethering, an observation unique to the final procedure. In earlier surgeries, monitoring was helpful in mapping the sacral roots but did not demonstrate this finding (Fig. 4). We placed the child on a long course of antiinflammatory medication in an attempt to minimize any intradural immune response. He regained complete sphincter control within days of the surgery, and at 9 months postoperatively, the symptoms had not returned.

Discussion
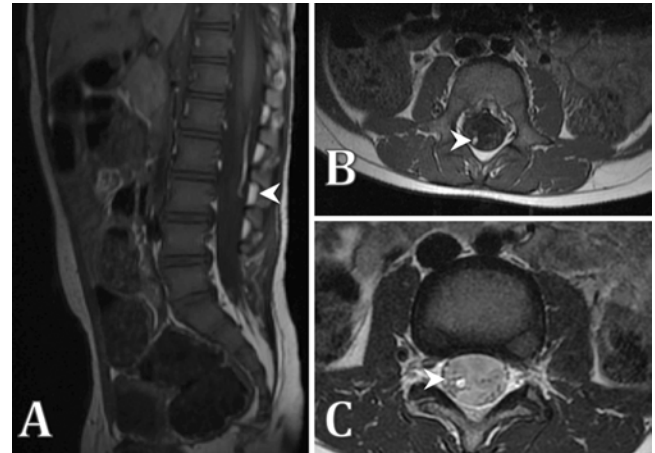
The surgical outcome of untethering in cases of fatty

FIG. 1. Magnetic resonance images of the lumbosacral spine obtained prior to the third procedure. Sagittal T1-weighted MR image **(A)** showing a right-angle bend of the stump of the fatty filum terminale (*white arrowheads*) to insert in the dorsal thecal sac. Axial T1-weighted **(B)** and T2-weighted **(C)** MR images depicting the proximity of the filum to the thecal sac and some nerve roots.

filum terminale is very good; some case series have reported success rates of 80% for the management of urological problems, 60%–80% improvement in neurological symptoms, and high levels of pain control.[1,10,15] Despite generally successful initial procedures, retethering is a very difficult to manage postoperative complication. Surgical series have shown retethering rates approaching 45% in more complex cases of myelomeningocele and lipomyelomeningocele; in cases of thick/fatty filum terminale sectioning, considerably lower rates have been reported, and scattered reports in the literature cite a retethering rate of about 8%–9% on long-term follow-up.[6,7,16] Most retethering is attributed to scar and arachnoid adherence around the neural elements attached to the spinal meninges, developing traction and
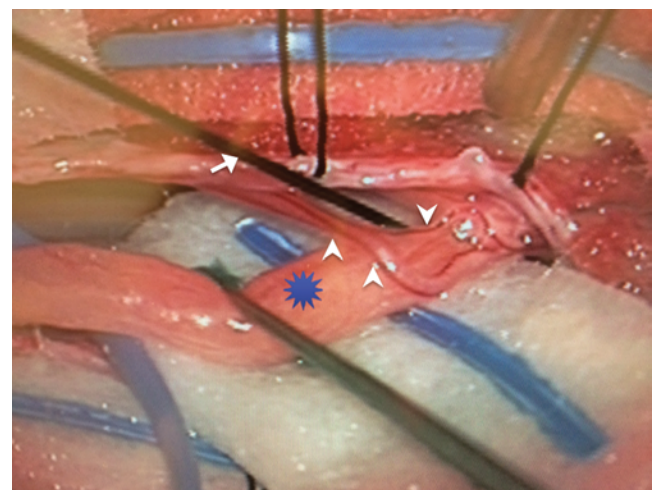

FIG. 2. Photograph from an intraoperative microscope showing the fatty filum (*asterisk*) and a stimulator probe (*white arrow*) used for mapping the entangled sacral nerve roots (*white arrowheads*). These nerve roots demonstrated stimulation of the electrodes placed within the right side of the anal sphincter and extensor hallucis longus muscle, confirming likely sacral origins. Figure is available in color online only.
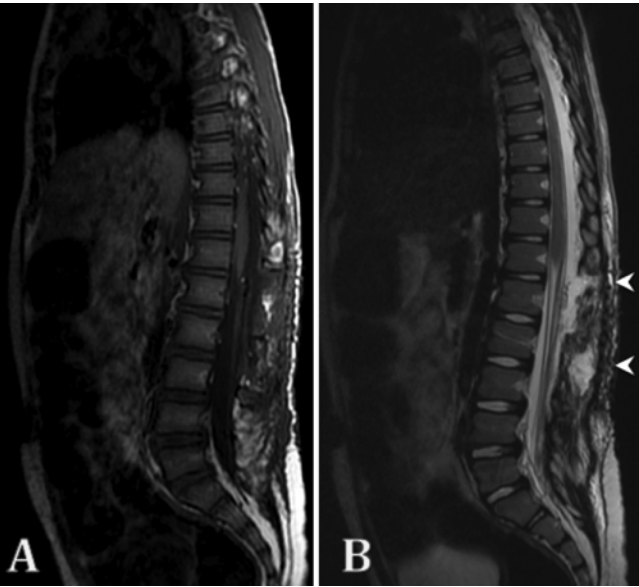
FIG. 3. Postoperative MR images obtained after the final untethering procedure, which was extended to T12–L1. Sagittal T1-weighted image **(A)** showing complete absence of the hyperintense signal of the fatty filum. Sagittal T2-weighted image **(B)** showing the different levels of the successive untethering procedures *(arrowheads)*; the upper level was the site of the most recent procedure.

nerve root dysfunction over time.[15] Maintenance of a pristine subarachnoid space during surgery and robust irrigation prior to dural closure have been espoused as technical nuances thought to minimize the risk of intradural adhesion. Expansile duraplasty has also been advocated to create more subarachnoid space, although it does introduce 2 new sites of potential adhesion at the suture line.[14]

Samuels et al.[9] treated 110 cases of symptomatic TCS, whose cause in 32% of the series was noncomplex, and 74% of the latter had fatty filum. The median length of follow-up was 42.5 months. Twenty-nine patients (26%) presented with symptomatic retethering; however, only 14% of these cases had a noncomplex etiology of TCS. In a large cohort, Ogiwara et al.[5] described 225 children with TCS treated by the release of fatty filum, and they established a 2.7% rate of recurrence. Yong et al.[16] described a cohort of 152 patients, 13 of whom (8.6%) had symptomatic retethering; the average time for symptom onset was 23.4 months. In all of the above studies, adhesions were described around the residual filar stump or other neural and meningeal elements being scarred, and this was generally regarded as the main etiology of the recurrent tethering. Other studies have also described similar findings.[2,11,13] However, in none of the studies were detailed specifications of intraoperative findings reported, nor were descriptions of discrete nerve root mapping outlined as the possible mechanism to specifically address symptom recurrence.
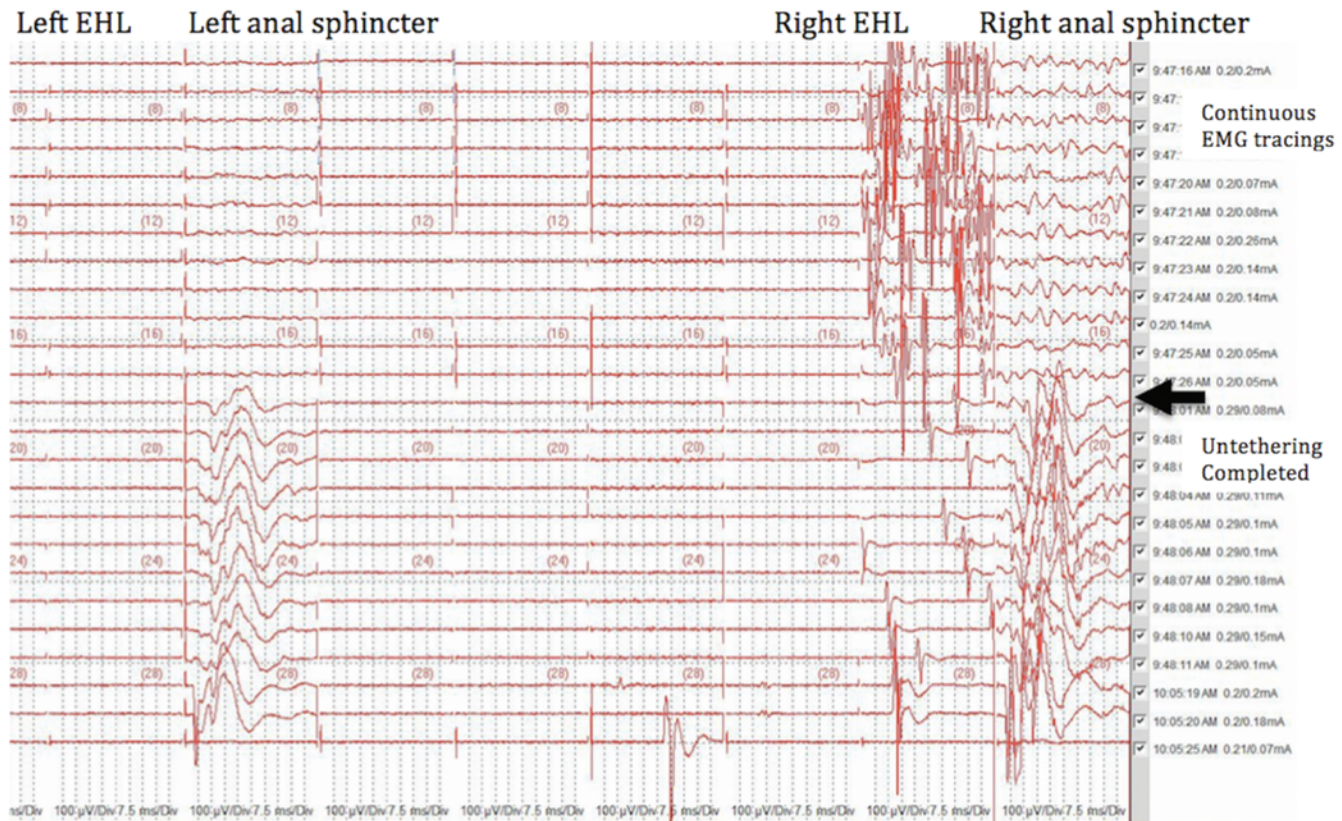

FIG. 4. Electromyographic (EMG) tracings of the left and right anal sphincter muscles demonstrating marked improvement subsequent to completion of the sacral nerve root neurolysis *(black arrow)*. Stimulation was applied with a standard 0.2-mA current. EHL = extensor hallucis longus. Figure is available in color online only.

Our case is unique in that we were able to demonstrate, fairly unequivocally, that the nerve roots of the sacral compartments became entangled and wrapped around the filar stump and were tethered to it, in this way producing objective symptoms. The use of monitoring is one interesting issue that this complicated case raises. Utilizing neurophysiological monitoring in this case, although often thought to be unnecessary in simple untethering procedures, was helpful in identifying nerves with anal sphincter innervation and was also useful to confirm maintained sphincter function after the last untethering procedure (Fig. 4). Others, however, use it routinely. Pang and colleagues, in 2009,[8] described extensive intraoperative monitoring as an important adjunct for untethering procedures. We similarly recommend its use in recurrent TCS procedures (regardless of the primary etiology), as they are often more complex interventions.

Clearly this patient had an unusual clinical course with multiple recurrences over a period of 34 months, from what we typically espouse to be a simple procedure. We felt this case would be a useful addition to the pediatric neurosurgical literature. The decision to pursue retethering surgically is often unappealing, but we suggest that the physiological pathways and specific symptoms producing the syndrome should be the primary guide, over radiographic findings, in the decision-making process. In this child's case, suggestive radiological findings that would otherwise unequivocally direct the surgical decision were absent, save once. Correlating symptoms with intraoperative electrophysiological studies should be emphasized. We also suggest that in cases of complex retethering, aggressive resection of the lipomatous elements may be considered to be part of the process to minimize retethering. Typically, we do not resect any of the lipoma at the time of fatty filum sectioning. This report along with the other documented cases in the aforementioned series serves as a reminder that thoughtful reexploration may be warranted when clinical symptom recurrence approximates the preoperative symptom profile even when imaging suggests a "successful" detethering.

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Disclosure

Author Contributions

Conception and design: Greenfield, Behaine. Acquisition of data: Behaine. Analysis and interpretation of data: Abdel Latif. Drafting the article: Behaine, Abdel Latif. Critically revising the article: Greenfield, Abdel Latif. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Greenfield.

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