Case Report

Recovery of Function Following Surgery and Rehabilitation for Tethered Cord Syndrome

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Adult patients who have had lower motor neuron weakness since childhood and who were born in countries without a well-structured childhood vaccination program are often assumed to have had poliomyelitis. Obtaining a detailed history and performing a thorough physical examination, with emphasis on limb-length differences, sensory changes, and bladder or bowel dysfunction, can lead to a different and potentially more remediable diagnosis. The use of neuroimaging techniques, such as magnetic resonance imaging (MRI) of the spinal cord or myelography with computed tomography (CT), is mandatory in cases in which a surgically remediable lesion is responsible for the weakness. This article presents the case of a 32-year-old woman who had received a diagnosis of poliomyelitis during childhood and later sought medical care because of leg atrophy and progressive radicular and back pain.

CASE PRESENTATION

Patient Presentation and History

A 32-year-old Puerto Rican woman came to the hospital’s neurology clinic because of dull pain in her lower back and sharp burning pain accompanied by paresthesias in a radicular pattern in her left leg for the past year. She reported no back trauma but mentioned that she collapsed in the street at age 11 years because of weakness in her left leg. Her doctor in Puerto Rico had diagnosed her condition as poliomyelitis. Two years later, she had undergone corrective surgery of the metatarsal bones of the left foot; surgical records were not available, but she remembered that the left leg had been smaller and shorter than the right. The patient continued to walk with a limp because of the leg-length discrepancy. When she came to the neurology clinic at age 31, she reported pain and numbness in her left foot and constipation but no bladder symptoms.

Physical Examination

Pertinent findings on physical examination included a soft, tender mass in the left paraspinal region at L3 to L5 without hypertrichosis. The left foot was fused at the ankle, and the left leg was 2 1/3 inches shorter than the right, with profound atrophy of the calf and foot. No scoliosis was present. A straight leg-raising test elicited no pain bilaterally. The patient’s gait was unsteady; muscle power was grade 4/5 in the left gastrocnemius, 2/5 in the anterior tibialis and extensor hallucis, 3/5 in the peroneus, and normal elsewhere. Pinprick sensation was decreased in the left L5 and S1 distributions. Deep tendon reflexes were brisk in both knees and in the right ankle but absent in the left ankle. Babinski’s sign was absent on the right foot and mute on the left foot.

Diagnostic Studies

A CT scan of the lumbosacral spine showed a lipoma of the distal filum with a tethered spinal cord and herniated disk at L5-S1. MRI of the cervical and thoracic spine revealed normal findings in the cervical region, a disk herniation to the right at T6-7 without cord compression, and a hemangioma of the T10 vertebral body. MRI of the lumbosacral spine revealed a lipoma (1.0 × 1.0 × 3.5 cm) at the L4-5 level that involved the filum terminale and was accompanied by tethering of the spinal cord, which was displaced posteriorly.

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Osteophytes were seen projecting from the end plates of L5 and S1, with disk desiccation. Urodynamic studies revealed normal bladder sensation and capacity, but voluntary bladder pressure was decreased.

**Treatment and Outcome**

The patient underwent a lumbosacral laminectomy from L4 through S2 with disattachment of the tethering from the cord. However, the lipoma itself was left in place, because several nerve rootlets were visualized inside it. Following surgery, the patient was placed on a tapering course of corticosteroids for 5 days. She continued to report severe back pain radiating to the entire left leg and described “a heavy feeling” of the left leg; sensation of the entire left leg was decreased to pinprick and vibration. The patient was unable to get out of bed or walk without assistance, and she was transferred to the inpatient service for intensive rehabilitation.

On admission to the rehabilitation medicine service, the patient’s physical examination revealed atrophy of the left anterior tibialis and gastrocnemius muscles and bony deformity and fusion in the left foot. Straight leg raising of the left leg was limited to 30 degrees because of pain. The patient experienced decreased sensation in the entire left leg and tenderness over the surgical scar, without warmth or erythema. Rectal sphincter tone was present, but the patient could not voluntarily contract the sphincter muscle. Functional evaluation showed that she was unable to ambulate without assistance and also required assistance for bed mobility, transfers, and activities of daily living (ADLs) because of pain. The patient underwent intensive rehabilitation for 17 days, including stretching and strengthening exercises of the abdominal and back muscles. In addition, she received training in ADLs, transfers, ambulation, and stair climbing. At the end of the rehabilitation program, she progressed to independence in bed mobility, transfers, and ADLs, could walk 300 feet using a straight cane, and was able to climb up and down 2 flights of stairs without assistance. Rectal sphincter function returned to normal. One month after discharge from the hospital, the patient continued to walk independently for functional distances and had tolerable pain for approximately 2 years.

Two years after her initial surgery, the patient returned to the hospital because of pain and dysesthesia in the left S1-2 distribution and progressive atrophy of the gluteal muscles on the left side. MRI revealed that the lipoma extended from L3 through L5 with a tethered cord, possibly caused by adhesions. After referral to a pediatric neurosurgeon, the patient underwent a second surgery through the original scar by a team specializing in congenital spinal diseases. Ultrasonography was used to localize the margins of the mass. The lipoma was debulked by 40%, and adhesions connecting the dura to the mass were removed under evoked potential surveillance. Postoperative MRI was remarkable for hemorrhage in the area of the lipoma. The extent of the untethering and lipoma removal was difficult to determine because the study was done only 2 days after the surgery. The patient was discharged from the hospital after a 2-week rehabilitation stay, and she continued to ambulate independently with no apparent pain or physical limitations.
operation. The patient has since resumed her previous level of activity with tolerable amounts of radicular pain and without losing bladder function.

DIFFERENTIAL DIAGNOSIS

Congenital malformations of the neural tube (ie, spinal dysraphism) can take many forms. In the basic pathology of tethered cord syndrome, the filum (ie, tail of the spinal cord) remains attached to the bony structures of the sacrum, causing stretching of the nerve roots or cord and resultant symptoms as a child grows. The tethering may be caused by adhesions from tumors, myelomeningocele, or infection or may result from spontaneous formation during development. The symptoms caused by tethered cord are quite variable. Signs of tethered cord syndrome in this part of the spinal cord always involve the lower motor neurons or nerve roots. Symptoms may include muscle weakness and atrophy, dysesthesias, neurogenic bladder, constipation, and sensory loss in a root distribution or pain in a radicular distribution, which may be elicited by straight leg-raising, causing further traction.

If a lipoma or benign fat-containing tumor is also present in the region of the cauda equina (as in the case patient), symptoms of root traction, compression, or pain in the lower back caused by hemorrhage also may be present. Lipomas can appear higher in the spinal canal, causing upper motor neuron symptoms (eg, spasticity, Babinski’s sign), but they are less common.

Differential Diagnosis

The patient was initially suspected to have had postpoliomyelitis syndrome, which is characterized by late, slowly progressive deterioration of strength in the same muscles initially affected by the poliomyelitis. Sensory loss is not a characteristic of this syndrome. Although some patients report pain with postpoliomyelitis syndrome, the pain is not described in the same manner as that seen with nerve root irritation (eg, herniated disk, pressure on the cauda equina from the lipomatous mass, stretching of roots from the tethered cord). The pain that accompanies postpoliomyelitis syndrome is described to be more cramp-like than tingling, and it may be similar to the pain and spasticity reported in amyotrophic lateral sclerosis, rather than to the numbness or paresthesia of neuropathy or sensory radiculopathy. Postpoliomyelitis pain is occasionally responsive to the antiepileptic drug gabapentin, which our patient took without benefit. Because of the sensory loss, dyesthesias, constipation, and limited progression of motor deficits, as well as the short duration of illness since the patient’s presumed poliomyelitis, it was doubtful on neuroanatomic grounds that she had postpoliomyelitis syndrome. In addition, the patient’s age placed her in an era of presumed vaccination for poliomyelitis. Although electrophysiologic studies would have contributed evidence for distinguishing the anterior horn cell dysfunction of poliomyelitis and postpoliomyelitis syndrome from the radicular involvement of a mass, we elected to pursue further work-up with radiographic investigation.

Diagnosis and Management

Following MRI of the lumbosacral spine in our patient, the diagnosis of lipoma with tethered cord syndrome was established, and the decision was made to perform surgical correction. Although information pertaining to the level of attachment of a lipoma, either at the filum or the conus, is valuable in determining the need for surgery, a final decision about the extent of the operation must be made after direct visualization of the contents of the lipoma during surgery. The resection is primarily performed to prevent further traction on the cord or compression of the cord or the nerve roots, but it is essential to avoid sacrificing or compromising any neural tissue during surgery. Similarly, the surgeon should avoid trauma to the posterior spinal cord that may result in sensory loss or interfere with the blood supply.

Unfortunately, both late deterioration of neurologic function and surgical complications (eg, nerve root injury, transection) are seen more often in patients with conus lesions. Nevertheless, with the accurate localization of pathology using MRI, the careful recognition and sparing of neural structures, and the use of intraoperative monitoring of spinal cord function by evoked potentials, a second surgery can be performed safely without undue damage to the caudal roots. Of course, it is preferable to avoid the difficulties of a repeated operation; therefore, all patients should be referred to an experienced neurosurgeon who uses the guidance of intraoperative evoked potential monitoring.

CONCLUSION

Although our patient underwent 2 surgical procedures and lost some use of the left gluteus muscle, overall her case had a good outcome. Successful arrest of progressive nerve root compression, pain management, and improved ability to perform ADLs were eventually achieved. Contributing to these results were the pursuit of the correct diagnosis of lipoma and tethered cord syndrome, performance of the appropriate surgical correction, and intensive rehabilitation, rather than acceptance of an incorrect diagnosis of postpoliomyelitis syndrome.
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REFERENCES