Conus Medullaris Enterogenous Cyst

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INTRODUCTION

This report discusses the unique presentation and outcome of a patient with a conus medullaris enterogenous cyst. The incidence of enterogenous cysts, differential diagnosis, imaging, presentation, and management are discussed. The unique aspects of this case presentation include a patient of advanced age with an atypical enterogenous cyst at clinical presentation and postoperative motor recovery despite prolonged cord compression.

CASE PRESENTATION

A 66-year-old white woman presented with a 2-year history of constant pain in her left anteromedial thigh and knee. She had experienced multiple falls due to left knee buckling, although she had experienced no specific left knee injury to cause the weakness. Due to this left lower extremity weakness, she required a walker to ambulate. There was no history of trauma, fever, back pain, recent weight loss, paresthesias, or changes in bowel or bladder. Her medical history was significant for obesity and hypothyroidism. Review of systems was otherwise unremarkable. Results of a physical examination revealed visible left thigh atrophy (thigh circumference was 6 cm smaller on the left side). Manual muscle testing demonstrated left iliopsoas and quadriceps strength of 2/5. Bilateral gluteus medius, hamstring, anterior tibialis, gastrocsoleus complex, and extensor hallucis longus strength, and right iliopsoas and quadriceps strength were all 5/5. Sensory examination was normal for light touch, pinprick, and vibration, with the exception of anesthesia in the left anterior and lateral thigh that extended to the proximal medial tibial region. Deep tendon reflexes were normal, with the exception of an absent left quadriceps reflex. Toes were downgoing bilaterally to plantar stimulation.

Electrodiagnostic testing of the bilateral lower extremities and related paraspinals revealed a left L2-L4 radiculopathy (anterior limb muscles and posterior paraspinal denervation evident). There was no evidence of right lumbosacral radiculopathy or plexopathy. Magnetic resonance imaging (MRI) of the lumbar spine without contrast performed to evaluate for a presumed diskal-structural lesion revealed a cystic structure in the distal cord–conus medullaris. A thoracic spine MRI with contrast demonstrated an expansile lesion of T1 and T2 prolongation signal in the distal cord–conus medullaris that was approximately 15 × 15 × 40 mm in the anterior and posterior, transverse, and cephalocaudad diameter, without pathologic contrast enhancement (Figure 1).

Initial conservative treatment included physical therapy, adaptive equipment, and observation because the patient was concerned about potential paraplegia with surgical intervention on the distal spinal cord. Needle electromyogram examination demonstrated the presence of active denervation of the left anterior thigh musculature. Physical therapy was initiated to strengthen the involved musculature, to instruct in compensatory gait mechanics and/or assistive device use, and to prevent falls. The strength of the left anterior thigh musculature did not improve, but compensatory strategies were used by the patient to prevent falls. A surgical solution was pursued approximately 18 months later when right quadriceps weakness became evident, upper motor long tract signs appeared, and the potential for long-term wheelchair use became obvious. The patient underwent a T11 to L1 laminectomy and subtotal removal of the conus cyst, with marsupialization and shunting to the subarachnoid space under microdissection technique. The conus medullaris cyst pathology revealed the cyst lumen to be lined by a cuboidal epithelium laying on a thin

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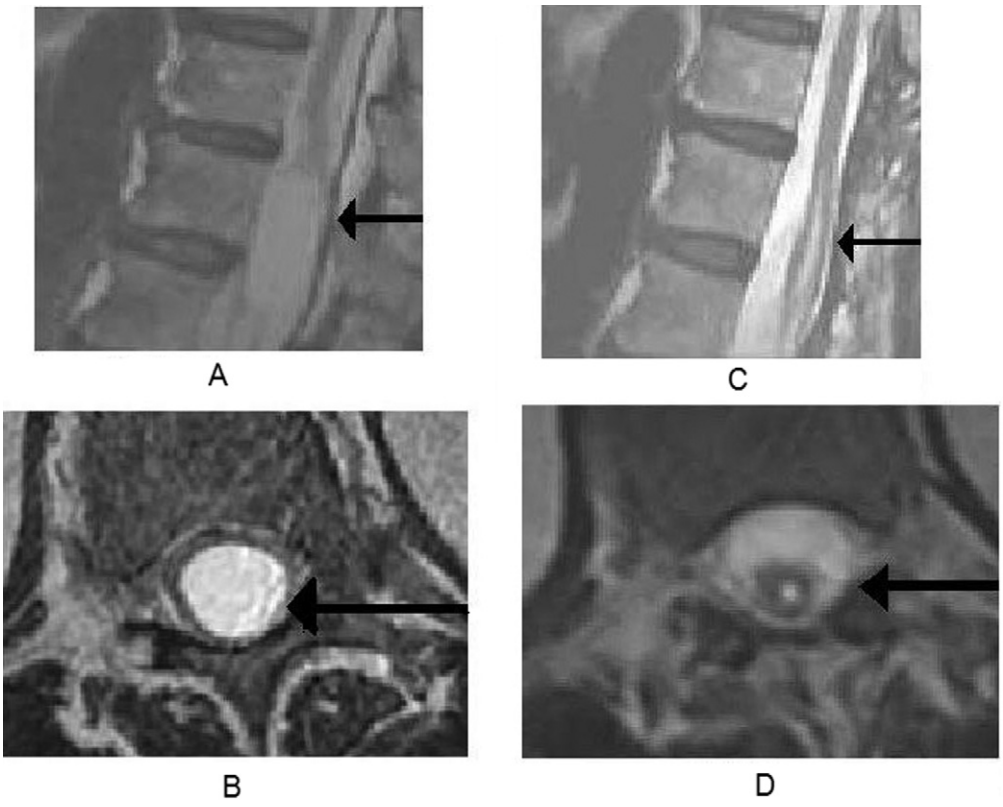

Figure 1. Preoperative sagittal (A) and axial (B) and postoperative sagittal (C) and axial (D) magnetic resonance image of a conus medullaris cyst.

continuous layer of fibrocollagenous tissue. Pathology was consistent with epithelial cyst of endodermal origin. After surgery, the patient's strength improved in the lower extremities. On her 2-month postoperative follow-up, she had complete recovery of right quadriceps strength. Her left iliopsoas and adductor muscle strength improved to 4/5. However, strength in her left quadriceps was unchanged, at 2/5. She had normal strength in the remainder of her bilateral lower extremity musculature.

DISCUSSION

Intradural spinal cysts are rare. They may be secondary to ependymoma, astrocytoma, and hemangioblastoma [1-3]. Benign intradural cysts usually are isointense with spinal fluid on MRI and show no enhancement with gadolinium–diethylenetriamine penta-acetic acid [3-6]. The lack of enhancement helps to differentiate such cysts from neoplastic lesions [4-6]. Benign intradural spinal cysts are histologically classified as arachnoid, enterogenousand ependymal cysts [7].

Our patient had a benign enterogenous intradural spinal cyst. Enterogenous cysts are believed to originate from embryonal dysgenesis. Normal development in the third week of embryonal life involves closure of the neuroenteric canal and notochordal separation from the primitive gut. Disruption of this process may lead to the inclusion of endodermal tissue and cystic formation [8]. Enterogenous cysts arise within the spinal cord and do not arise from the meninges. Enterogenous cysts tend to present earlier in life because of their origin from embryonal dysgenesis. Potential etiologies of enterogenous cysts include the following: aberrant vascular supply to the neuroenteric tube, adhesions between ectoderm and endoderm, and persistent primitive neuroenteric canal.

Isolated spinal cord enterogenous cysts are rare lesions. Fewer than 100 have been reported in which there were no associated bone or soft-tissue malformations [3]. These enterogenous cysts comprise 0.7%-1.3% of all spinal cord masses or tumors [4]. Enterogenous cysts affect the spinal cord far more commonly than the cranium, skull base, and brain. Of those cysts within the cord, Lipman et al [4] reported that approximately 54% occurred in the cervical, 16% in the thoracic, and 18% in the thoracolumbar regions, whereas the remainder were located more caudally (conus medullaris) [4].

Enterogenous spinal cord cysts are benign formations that infrequently result in neurophysiologic deficits. Most en-

terogenous cysts are asymptomatic incidental imaging findings that are observed and remain stable over time. Symptomatic enterogenous spinal cord cysts present with upper motor neuron signs, and functional deficits are typically diagnosed in early adulthood. In some patients, the clinical course is variable, with symptomatic exacerbations and remissions [4]. Patients present with spinal pain, paresthesias, and paraparesis [9]. Bowel and bladder dysfunction may be observed [1,5,10]. Patients typically present for medical care after the development of motor weakness [4]. During the first decade of life, the presentation usually is an acute onset with progressive neurophysiologic decline [3]. Teens and young adults in the second decade have an insidious presentation, with either intermittent or rapid progression [3,11]. Enterogenous cysts are almost 3 times more likely to occur in men than in women [4]. Anterior spina bifida and vertebral anomalies are present in as many as 50% of documented cases [3]. Our patient did not have any form of spina bifida or any vertebral abnormalities.

Early diagnosis is imperative to avoid permanent neurophysiologic loss that results from prolonged neural compression within the spinal cord [1,5]. Management strategies differ substantially, depending on the kind of lesion. Surgical treatment approaches include aspiration, total resection, partial resection, marsupialization of the cyst, and cystosubarachnoid shunt [4]. Recurrence of the cyst has been reported with simple aspiration and, as a result, is not a recommended treatment [4]. Positive outcomes are reported with subtotal resection and marsupialization [4].

Our patient presented late in life with a rare congenital enterogenous cyst of the epithelium with immunohistochemical features of endodermal origin in the conus medullaris. This presentation was atypical in a variety of aspects. The large size of the cyst, the lengthy duration of cord compression, the patient's gender, and her advanced age at symptom presentation were all atypical. The favorable outcome was remarkable, given the severity of neurophysiological loss. Upper motor neuron signs were not evident on clinical examination but were expected when considering that the cyst occupied a large area of the distal cord and distorted the shape of the cord from T11 to L1 (Figure 1A, B). Slowly accumulating fluid within the soft tissues of the cord presumably allowed the neural tissue time to adapt to the enlarging space–occupying lesion within the spinal cord over many years before neurophysiological change was evident. The significant change in the appearance of the cord with cord nervous tissue pushed out into a ring within the spinal canal, as demonstrated in Figure 1A and B. Preoperative cord appearance and the evident rebound of the tissues back into a more normal configuration after surgery are demonstrated in Figure 1C and D.

In our patient, sensory and motor deficits occurred for at least 24 months before definitive surgical treatment, and the degree of postoperative motor recovery was remarkable in the right lower extremity, even though left quadriceps weakness persisted. Marsupialization of enterogenous cysts is a technically simple procedure for appropriately skilled neurosurgeons that is associated with a low risk of significant complication, paraplegia in this instance. The earlier that enterogenous cyst marsupialization can be performed, the better the functional outcome can be expected. Our patient preferred conservative treatment options rather than surgical intervention. The physiatrist communicated with the treatment team (neurosurgeon, therapists), performed serial clinical examinations, and counseled the patient over a continuum of time in the treatment process. In the final analysis, the patient was well informed, actively made her own decisions about treatment, maintained the ability to walk since she regained right lower extremity motor function, and was able to perform both basic and advances activities of daily living with the residual left anterior thigh weakness. The physical examination, anatomical imaging, electrodiagnostic testing, and a team approach across medical specialties worked in concert to properly discern symptom, condition, and causation, which led to an effective treatment for our patient.

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