Sequestered High Lumbar Intradural Disc Herniation Mimicking A Spinal Tumor: Case Report And Review Of The Literature

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Citation

Abstract
We report an unusual case of a sequestered high lumbar intradural disc herniation mimicking a spinal tumor radiologically in a 49-year-old man who presented with bilateral radicular pain and weakness of the lower extremities. Magnetic resonance (MR) imaging of the lumbo-sacral spine revealed the presence of an intradural extramedullary lesion at the L1-2 level abutting the conus medullaris. The patient underwent laminectomy and microsurgical resection of the mass. Pathological examination of the resected lesion revealed it to be of disc material. Postoperatively, the patient experienced complete remission of symptoms.

The differential diagnosis of a sequestered intradural enhancing lesion in the lumbar region causing nerve root compression or cauda equina syndrome must include disc herniation. In addition, early surgery provides the best opportunity for full neurological recovery.

INTRODUCTION
Intradural disc herniation, although comprising only 0.26-0.30% of all cases of disc herniation, is a serious complication of intervertebral disc disease[1, 5]. First described by Dandy[4] in 1942, more than 100 cases of intradural disc herniation have since been reported in the literature[7,8]. The majority of these cases (92%) involve the lumbar spine, most commonly the L3-L4 and L4-L5 disc spaces. High lumbar intradural disc herniations constitute just over 17% of the total cases of lumbar intradural disc herniation, occurring at the L1-L2 (8%) and L2-L3 (9%) disc spaces[5,8]. We report an unusual case of a sequestered intradural lumbar disc herniation occurring at the L1-L2 interspace, minimally attached to the ventral dura, which mimicked an intradural extramedullary tumor on radiological evaluation.

CASE REPORT
PRESENTATION
This 49-year-old man, with a 2-year history of back pain, presented after three months of worsening low back pain that radiated bilaterally down the back of his thighs and legs. The patient had right lower extremity numbness that was most profound distally, and he reported substantial subjective weakness on the right side to an extent that caused him to use a single prong cane for added stability while walking. He reported no bladder or bowel symptoms. One year before presentation, the patient had developed similar symptoms of radiating pain down the backs of both legs and bilateral lower extremity numbness. Magnetic resonance (MR) imaging at the time showed generalized degenerative disease with no specific abnormality, and his symptoms were minimized after a few months of physical therapy and weight loss.

EXAMINATION
General examination revealed an obese male weighing 260 lbs with multiple skin tags and pigmentation on his torso. Neurological examination revealed decreased sensitivity to pinprick sensation in the distal right lower extremity involving multiple dermatomes. Joint proprioception was impaired in the right toes but was intact in the right ankle and knee as well as in the entire left lower extremity. Strength in his lower extremities was 5/5 in all muscle groups. Deep tendon reflexes were present and symmetric in the upper extremities but were essentially absent in the lower extremities. His toes were down-going bilaterally, and the Hoffmann sign was absent in both upper extremities. The
patient had a slight limp while walking and favored the right side.

**PREOPERATIVE IMAGING STUDIES**

MR imaging of the lumbosacral spine with and without gadolinium showed mild to moderate multilevel degenerative disc and facet changes. At the L1-L2 disc space level, an intradural lesion characterized by prolonged T1- and T2-weighted signal intensity and peripheral enhancement was detected (Fig. 1).

**Figure 1**

Figure 1: Preoperative sagittal T1-weighted MR images (A) without and (B) with gadolinium show an intradural enhancing lesion (arrows) just below the conus medullaris.

The lesion was occupying the right side of the spinal canal primarily and was displacing the adjacent cauda equina to the left (Fig. 2). It appeared to be sitting just below the tip of the conus medullaris. The central portion of the lesion did not enhance and appeared to be an area of cyst formation or necrosis. The differential diagnosis included a nerve sheath tumor, an ependymoma, and a meningioma.

**OPERATION**

A thoracolumbar laminectomy and durotomy revealed the presence of an intradural mass directly below the conus medullaris and displacing the cauda equina to the left (Fig. 3).
Figure 3
Figure 3: Intraoperative photograph. After durotomy was performed, a lobulated glistening mass (arrow) was identified that was compressing the cauda equina nerve roots (asterisk).

The lesion seemed to arise from the ventral dura. A biopsy of the mass was sent for pathological examination in frozen sections. It revealed that the lesion was of disc material. A total resection of the lesion was performed. Post resection inspection revealed an intact and mobile ventral dura. The dorsal dura was then closed primarily, and wound closure completed the procedure.

POSTOPERATIVE COURSE
The patient recovered uneventfully and exhibited complete remission of his neurological symptoms on follow up visit.

DISCUSSION
The highest incidence of intradural disc herniation occurs in the fifth decade; more than 50% of patients present in their 40s and 50s, and 75% are males. Dandy, in 1942, reported the first case among 300 patients who underwent surgery for lumbar disc herniation. Since then, more than 100 additional cases have been reported.

Although the pathogenesis of intradural disc herniation is not understood, a number of theories have emerged over the years seeking to explain this unexpected occurrence. Dandy hypothesized initially that the sudden pressure created by a herniated disc leads to erosion and penetration of the underlying dura. Blikra studied the relationship between the ventral dura and the posterior longitudinal ligament in 40 cadaveric specimens. He found frequent dense adhesions between the two structures throughout the lumbar spine. In addition, he found in some cases that the connection between the dura, the posterior longitudinal ligament, and the annulus fibrosis of the disc was so firm that it could not be divided by blunt dissection. Blikra concluded that in cases when these structures were firmly adhered, a herniated disc could penetrate through them as if they were one structure.

The idea that adhesions predispose to intradural disc herniation has been a common theme in the literature since Blikra's study. Adhesions are thought to develop from a number of different mechanisms including chronic irritation from a herniated disc, previous surgery, chronic local inflammation, or congenital/iatrogenic factors. Other possible explanations include chronic osteophytosis, which may predispose to medial herniation through the dura, and congenital narrowing of the spinal canal, which results in less epidural space. Our case is unique in that the disc fragment was completely sequestered intradurally with minimal attachment to the ventral dura which was intact at surgery.

As seen in our case, intradural disc herniations frequently develop in patients with a history of chronic back pain. Seventy-nine percent of the cases of intradural lumbar herniation reported in the literature occurred in patients who had suffered from low back pain or sciatica for more than one year. In addition to their chronic symptoms, patients typically develop an acute progressive neurological deficit such as cauda equina syndrome or bilateral lower extremity radiculopathy, often after a precipitating event. Generally, patients with intradural disc rupture tend to have more severe neurological symptoms than would be expected from an uncomplicated herniated disc. Cauda equina syndrome in particular has been found in more than 60% of patients with intradural lumbar herniations. In addition to lower extremity weakness and sensory deficits, these patients often develop other symptoms typical of cauda equina syndrome such as saddle anesthesia or bladder and bowel dysfunction. Urinary disturbance was present in 31 of 46 patients with intradural lumbar disc herniation, while anal sphincter disease has a reported incidence of 30%.

Therefore, intradural lumbar disc herniation should be considered in the differential diagnosis of intradural, extramedullary mass lesions that cause nerve root or cauda equina compression.

The most common intradural mass lesions found in the lumbar spine are benign tumors of the spine and its surrounding structures. These lesions include neurofibromas, meningiomas, ependymomas, arachnoid cysts, and epidermoid/dermoid cysts. Metastatic disease also must be considered in any patient with spinal lesions. Differentiating
these intradural lesions radiographically from an intradural herniated disc is difficult, and this difficulty is one of the main reasons that this condition is rarely diagnosed preoperatively. In the past, myelography was the imaging modality of choice, and it would characteristically demonstrate an irregularly marginated intradural filling defect at the level of the ruptured disc[4]. In comparison, benign intradural tumors were found to cause more smoothly marginated rounded filling defects[5]. The addition of computed tomography (CT) to myelography allowed for better visualization of the intradural component of a herniated disc, thus improving diagnostic accuracy[6]. Myelography essentially has been abandoned in favor of contrast enhanced MR imaging, which Holtas et Al[7], described as superior in visualizing all components of the herniation including surrounding inflammation and adhesions. On T1- and T2-weighted MR images[8] intradural discs typically appear as homogeneous isodense lesions. With the addition of gadolinium, however, these lesions develop a characteristic peripheral ring enhancement, which is believed to represent granulation tissue around the nonenhancing disc[9]. Neurofibromas and meningiomas, by contrast, both show homogeneous enhancement on contrast MR imaging, while ependymomas typically appear hyperdense on T2- weighted images[2]. Despite these clear-cut radiographic differences, however, complications in differentiating lesions on MR imaging commonly occur. For example, in cases when intradural disc herniation has occurred acutely and granulation tissue has not yet developed, the lesion will fail to show ring-enhancement on MR imaging[10]. When the disease process has been ongoing for a while and granulation tissue has infiltrated the disc fragment, the lesion tends to enhance homogeneously[11]. Therefore, although MR imaging is an effective diagnostic tool, intradural disc herniations continue to be diagnosed most frequently intraoperatively. Diagnosis of the mass in our patient was imprecise until the pathologist reviewed frozen sections of the lesion. Given the location, rare occurrence, and radiological finding, our differential diagnosis favored an intradural extramedullary tumor over a herniated disc.

Treatment of intradural lumbar disc herniations requires a laminectomy followed by dural opening and removal of all ruptured disc material. In cases of ventral herniation, the associated dural tear must be sutured or covered by an adhesive to prevent cerebrospinal fluid (CSF) leakage[12]. If adequate nerve root decompression is achieved, full neurological recovery can be expected in 67% of patients, whereas 33% will continue to show some residual neurologic deficit after surgery[13]. The most important prognostic factor for which patients will likely achieve full recovery, besides the adequacy of disc removal during surgery, is the preoperative duration of neurologic symptoms[14]. Shorter duration strongly correlates with better outcome. It is therefore essential, that these patients be both diagnosed early in the disease process and treated with prompt surgical intervention.

CONCLUSIONS

We present an unusual case of a sequestered high lumbar intradural disc herniation at the L1-2 interspace abutting the conus medullaris mimicking an intradural tumor. Despite its infrequent occurrence, disc herniation must be entertained in the preoperative diagnosis of patients who present with neurological deficits and in whom intradural mass lesions of the lumbar spine are revealed radiographically. Early surgery should be offered to provide the best opportunity for full neurological recovery.

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