Spinal Intramedullary Lipoma without Dysraphism

Spinal cord lipomas are commonly associated with spinal dysraphism. The lipomas without spinal dysraphism have been reported to be only 1% of all spinal cord lipomas. We report two cases of patients with spinal intramedullary lipomas without dysraphism. One patient was a 67-year-old man who had lower back pain and right-sided sciatica for 20 years. Magnetic resonance imaging (MRI) of the spine demonstrated an intradural mass occupying L1 to L2. The other patient was a 27-year-old woman who had back pain and gait disturbance for 5 months. MRI of the spine showed an elongated intradural mass occupying T7 to T8. Total laminectomy with partial resection of the lesions was performed on both patients. Pathological studies confirmed the diagnosis of spinal cord lipoma. They exhibited no dysraphism. Postoperatively, neurological symptoms improved in both patients.

KEY WORDS: Intramedullary lipoma, Dysraphism.

INTRODUCTION

Intradural spinal cord lipomas are rare benign tumors of the spinal cord, accounting for less than 1% of all intradural spinal cord tumors. Spinal cord lipomas are commonly associated with spinal dysraphism, such as spina bifida, lipomyelomeningocele, myelomeningocele, diastematomyelia, cutaneous lipoma, fistula, pilosity and Klippel-Feil malformation. However, the lipoma without spinal dysraphism have been reported to be only 1% of all spinal cord lipomas. We report two cases of lipomas without dysraphism.

CASE REPORT

Case 1

A 57-year-old man presented with severe lower back pain, severe right-sided sciatica, and voiding difficulty of 20-year duration. Neurologic examination showed: straight leg raising test, (45 degrees/60 degrees); Naffziger's sign, (+) and ankle clonus bilaterally, a positive Babinski's sign. On plain radiographic examination of the lumbar spine, no evidence of dysraphism was demonstrated. Magnetic resonance imaging (MRI) of the spine demonstrated an intradural ovoid mass occupying L1 to L2, which was a hyper-intense lesion on T1-weighted image (Fig. 1).

We performed total laminectomy from L1 to L2. It revealed a pale, non-pulsatile and bulging dural sac. After opening the dura, a microscope was used to continue the procedure. The lipoma was exposed at posterior side of spinal cord and the cord was displaced anteriorly. A debulking of the lipoma was carried out by micro-scissor and cavitron ultrasonic surgical aspirator (CUSA). However, further removal was not attempted, because of no interface between lipoma and spinal cord. A watertight dural closure was per-
formed followed by a muscle and skin closure.

Histopathological examination revealed infiltration of mature adipocytes, which varied only slightly in size and shape and had small eccentric nuclei (Fig. 2). Postoperatively, the patient reported pain relief. But, he had residual urine and required regular intermittent catheterization but his problem resolved at discharge. On follow-up MRI taken 5 months after surgery, there was a residual intradural mass occupying L1 to L2 at decompressive laminectomy state (Fig. 3).

Case 2
A 27-year-old woman presented with severe back pain and marked gait disturbance of 5-month duration. Neurologic examination showed: hypesthesia below the T10 dermatome, weakness of both lower extremities with motor grade IV and ankle clonus bilaterally. On plain radiographs of thoracic spine, no evidence of dysraphism was demonstrated. MRI of the spine demonstrated an elongated intradural mass occupying T7 to T9, which was a high signal intensity lesion on T1-weighted image (Fig. 4).

We performed total laminectomy from T7 to T9. It revealed a pale, non-pulsatile and bulging dural sac. After removal of ligamentum flavum and opening the dura, a microscope was used to continue the procedure. The lipoma was exposed at posterior side of spinal cord and the cord was displaced anteriorly. A debulking of the lipoma was carried out by micro-scissor and cavitron ultrasonic surgical aspirator (CUSA). The lipoma was intermingled with spinal cord and thus no cleavage plan could be found between the normal neural tissue and the tumor. So, we performed a partial tumor mass removal. Partial resection of the lipoma was enough to decompress the spinal cord. Postoperatively, she had mild lower back pain and slight gait disturbance. Motor was weakness improved to leg elevation equivalent to grade V/V. Histopathological diagnosis was reported as a lipoma composed with mature adipocytes, which varied only slightly in size and shape and had small eccentric nuclei (Fig. 5). The postoperative MRI after 14 months demonstrated good decompression of the spinal cord with remnant lipoma (Fig. 6).
DISCUSSION

Spinal cord lipomas are very rare and account for approximately 1% of all spinal cord tumors in adults and 5-7% in children. Sixty percent are localized intradurally and 40% extradurally. Intradural lipoma most often occurs in young adults in their second and third decade of life without sex predilection. Spinal cord lipomas are classified into those with spinal dysraphism and those without spinal dysraphism. The first group of these occurs mostly in pediatric ages. These consist of lipomas located at the lumbosacral junction. They are generally associated with spinal dysraphism. The second group is less frequently observed, and tends to occur principally in the adult. Complete intramedullary spinal cord lipomas without spinal dysraphism are very rare and have been reported to be only 1% of all spinal cord lipomas. They are usually considered congenital inclusion tumors.

Most spinal lipomas originate in the dorsal intramedullary region in the spinal cord. However, the embryologic defect that leads to the development of these tumors is unknown. Several hypotheses have been proposed to explain how spinal cord lipomas arise. The most widely accepted theory is that such developmental malformation occurs during the formation of the neural tube and leads to inclusion of embryonic crests of fat cells. The mechanism starts from the premature separation of cutaneous ectoderm from neuroectoderm during the process of neurulation. This premature separation permits the surrounding mesenchyme to enter the ependymalized central canal of the neural tube, which has not yet closed. Moreover, the mesenchyme entering the central canal then differentiates into fat. It seems to develop into fat either by receiving signals promoting adipocyte production or through blockage of signals that prevent adipocyte production.

The fat of the lipoma behaves metabolically just like the rest of normal body fat and changes their size according to alteration of body fat. Growth of a lipoma has been observed mainly in patients with an increase in body fat.

These clinical symptoms are related to mass effect and secondary compressive myelopathy. Presenting symptoms include spinal pain, dysesthesia sensory changes, gait difficulties, weakness, and incontinence. Most patients have progressive impairment of the lower extremity, bladder, or bowel functions. In addition, spinal dysraphism shows cutaneous lesions. Cutaneous markers in a high percentage accompany spinal malformations aiding the clinician in further diagnostic and therapeutic work. Spinal cord tethering resulting from occult spinal dysraphism causes progressive neurological deficits, pain, and spinal deformity. However, most patients with spinal cord lipomas in non-dysraphic states may have normal spinal function. In patients with intramedullary spinal cord lipoma, spinal cord function varies because intramedullary lipoma tissue replaces the normal spinal cord tissue during the developmental process.

MRI is the most useful diagnostic imaging modality. MRI not only confirms the fat component of the tumor as CT does but also delineates its relationship to the adjacent neural structures, which is important for surgeon. It has been proposed that neither CT nor myelography was necessary.
before operation other than MRI\(^7\). High signal intensity lesions on T1-weighted images are quite characteristic of lipomas due to its very short relaxation time of the fat\(^8\)\(^9\). Lipoma tissue demonstrates a low signal on T2-weighted images. Plain film of spine usually shows dorsal midline fusion defects.

Some authors conclude that patients with intradural spinal lipoma who present with significant neurological compromise have a very poor prognosis with regard to neurological function and generally show no improvement with surgical resection\(^10\). Prophylactic surgery with asymptomatic patients is not advisable\(^9\). Surgical decompression is indicated for cases in which neurological symptoms are progressive. However, total excision has generally been shown to achieve a lesser degree of postoperative improvement, and in the literature, some cases have been reported to have a significant worsening of symptoms\(^11\). Therefore, aggressive complete removal of this tumor is not recommended because these lesions do not have a clear-cut margin\(^11\). The goal of surgery for this tumor is not complete resection of the lesion, but decompression of the adjacent neural structures, even if it entails incomplete resection. Excision with the help of a carbon dioxide laser can preserve the neurologic function. Because of the high water content of lipoma, the laser vaporizes the fatty tissue without physical manipulation of the neural tissue\(^8\). Additionally, in multi-level involved spinal lipomas, laminoplasty with preservation of intervertebral joint is sometimes recommended because of the possible spinal deformity or instability, especially in young or middle-aged patients\(^9\).

CONCLUSION

We report two cases of intradural lipomas without spinal dysraphism. The goal of surgery is decompression of intradural lipoma. It is suggested that a careful and limited decompression of the affected spinal cord through a partial tumor removal may result in a neurological improvement.

References