CASE REPORT

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Intramedullary Spinal Cord Lipoma without Spinal Dysraphism

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Extradural lipomas have been frequently reported in the literature, but intramedullary lipomas are far rarer, constituting only approximately 2% of total intramedullary tumors. Intramedullary lipomas are also commonly associated with spinal dysraphism. Lipomas which are not associated with spinal dysraphism are present in only about 1% of spinal lipoma patients. Here, we report a rare case of a patient suffering from an isolated intramedullary lipoma without evidence of spinal dysraphism.

KEY WORDS : Lipoma · Intramedullary cord tumor.

Introduction

Spinal cord lipomas without spinal dysraphism are extremely rare lesions, which account for only 1% of all spinal masses, with only 100 cases ever having been reported[1,6]. Furthermore, true intramedullary spinal cord lipomas are even rarer, and are represented in the literature only as scattered individual case reports[14] .

Here, we report a case of intramedullary lipoma which was not associated with spinal dysraphism, and have reviewed the characteristics of this rare disease entity.

Case Report

History

A 57-year-old man presented to our institution suffering from left dominant paraparesis, right leg hyperesthesia, and lower back pain. The patient's symptoms had progressively gotten worse for the previous 2 months. His bowel and bladder functions were normal. The patient denied having any history of either trauma or systemic disease.

Examination

Upon physical examination, the patient exhibited none of the typical features of either spinal dysraphism or cutaneous abnormalities. The motor power of the patient's left leg was scored as grade IV-, and his right leg was scored as grade IV+, with brisk deep tendon reflexes. He complained of decreased temperature sensation in his right lower extremities, and also experienced reduced vibration sensation and proprioception in his left lower extremities, both of which are consistent with Brown-Sequard syndrome. The Babinski reflex could be evoked with the patient's left foot, but his bowel and bladder functions were normal, as stated earlier. Magnetic resonance imaging(MRI) revealed an intramedullary mass located in the lower thoracic spinal cord, extending from T9 to T12. T1-weighted and T2-weighted images both revealed an ovoid region of increased signal intensity, which exhibited no enhancement of the mass with gadolinium, whereas the signal intensity is diminished when the fat saturation technique was exploited (Fig. 1).

Operation

There was no evidence of spinal dysraphism after the exposure of the lamina. The dura was opened along the midline, and the entire tumor was observed to be covered with a pale pia, which adhered to the arachnoid membrane. The tumor mass was soft and yellow, without clear margins (Fig. 2). A conservative, subtotal removal was conducted, and the excised tissue was immediately sent to the laboratory for pathological assessment. The intimate relation of the lipoma to the nerve roots, as well as the absence of a distinct plane between the

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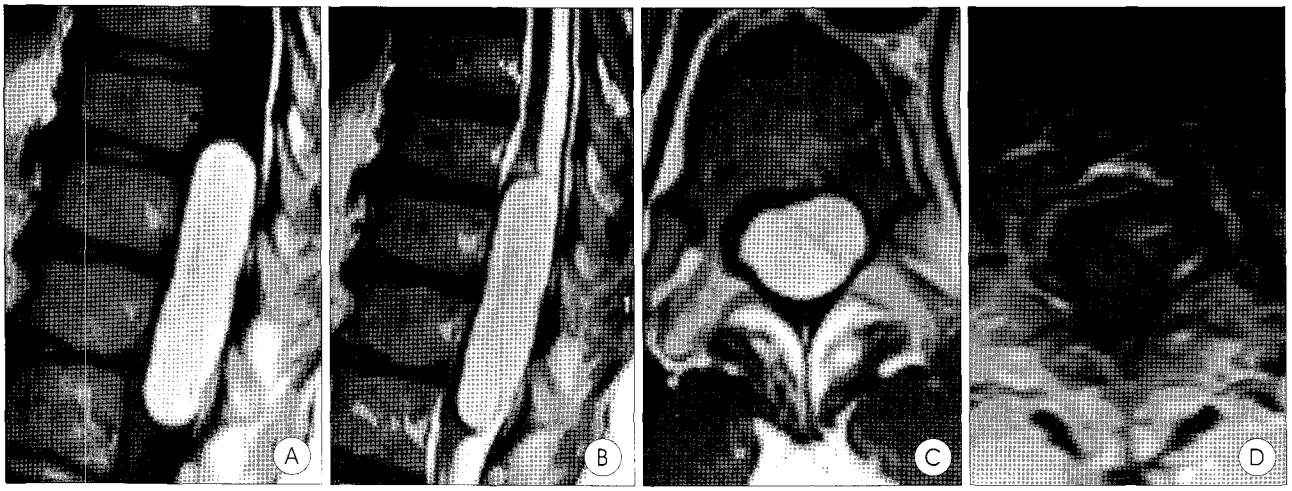

Fig. 1. Sagittal nonenhanced T1-(A) and T2-weighted magnetic resonance(MR) images (B) show an elongated intramedullary mass at the T9 through T12 level. The high signal intensity is consistent with fat. T1-weighted axial image (C) demonstrates a mass at the level of T10. Precise delineation of the spinal cord is not possible because the intrinsic lesion deforming it. Signal of the fatty components of the mass is suppressed by a T2 fat suppression MR sequence (D).

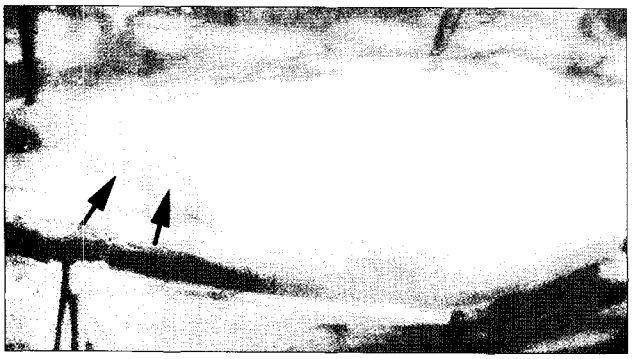

Fig. 2. Intraoperative photograph reveals a yellowish fatty tumor (arrows) protruding from the subdural space.

Fig. 3. Histopathologic examination reveals that the tumor is composed of mature adipose tissue and fibrous septa without evidence of malignancy or dedifferentiation (H&E, X200).

tumor and the spinal cord, made the complete resection of this tumor impossible. Somatosensory-evoked potentials did not change substantially during the operation.

A histopathological examination revealed mature adipocytes, intermixed with adjacent mesenchymal fibrous tissues (Fig. 3).

Postoperative course

The patient's postoperative recovery was uneventful. His motor weakness improved markedly, from grade IV- to grade IV+, and the back pain and the dysesthesia/numbness of the patient's right leg had also improved after surgery.

Discussion

Lipomas account for less than 1% of all spinal tumors in adults and for 5~7% of all spinal tumors in children[2,10]. These tumors are most frequently extradurally located, and are normally associated with spinal dysraphism[4,5]. Sacral or lumbosacral defects which communicate with the subcutaneous lipoma are also quite commonly discovered[14].

Spinal lipoma are often classified as lipomas with spinal dysraphism or without spinal dysraphism[2]. The first major group of these, which occur mostly within the pediatric population, consists of lipomas located at the lumbosacral junction, between the conus medullaris and the filum terminale. This group of lipomas is generally associated with spinal dysraphism (spina bifida, myelomeningocele), and/or cutaneous abnormalities (subcutaneous lipoma, fistula, pilosity). Over 90% of all spinal lipomas are classified into this group[2]. The second group is far less frequently observed, and tends to occur principally in the adult population. This group is comprised of intraspinal subdural lipoma, which are normally localized in the cervical, thoracic, and cervico-thoracic regions, and are usually not associated with spinal dysraphism or with cutaneous abnormalities[2].

Although the pathogenesis of spinal lipoma associated with

spinal dysraphism is generally believed to represent the result of a dysfunctional developmental process, metaplasia, or to be of harmatomatous origin[9,11], the actual pathogenesis of intramedullary lipoma remains still unknown[17]. However, many investigators consider the embryonic malformation neural tube theory to be one of the most plausible hypotheses advanced[3,14,19]. According to this theory, the formation of spinal intradural lipomas results from the separation of the primitive ectoderm into the neuroectoderm, neural crest, and cutaneous ectoderm, the simultaneous fusion of the resulting neural plate edges in a posterior direction, and the consequent disruption in the formation of the overlying edges of the cutaneous ectoderm. A brief disjunction in the timing of the process occurs. The cutaneous ectoderm closes just before the neural tube, and a transient gap is left before the neural tube, through which paraxial mesenchyme are able to migrate. This mesenchyme becomes, for the most part, fat under of inductive influences exerted by the inner ependymal surface of the placode, although tissue of mixed embryonic origin, such as bone, cartilage, smooth or striated muscle, collagen, renal epithelia, and gastric mucosa may also persist within these lipomas to some degree[21].

With regard to the evaluation of spinal lipomas, MRI has proven to be superior to CT in the sagittal plane, whereas CT clearly retains the edge over MRI in the axial plane[6,16]. The relaxation times of fat on T2-weighted images tend to be quite variable, and can appear hyperintense, isointense, or hypointense, as compared to normal neural parenchyma[18]. However, high signal intensity on T1-weighted images, due to the very short relaxation times of fat on T1-weighted images, is a clear characteristic of lipomas[10,20]. This tissue can be confirmed to be fat via the performance of a fat suppression MRI[7,18]. It is important, in these cases, to differentiate between epidural lipomatosis and intradural or intramedullary lipomas[8]. Epidural lipomatosis refers primarily to spinal lipomas within the epidural space, whereas the term "intramedullary lipoma" refers to an intradural or a subpial lipoma, which is usually found to have merged with the medullary substance of the spinal cord, making its total excision almost impossible, without inflicting permanent neurological damage. MRI scans normally prove helpful in the identification of the locations of these tumors[8].

Although the appropriate treatment of intramedullary lipoma is a matter of some controversy, surgical decompression is indicated in cases in which the symptoms progress. However, total excision has generally been shown to effect a lesser degree of post-operative improvement, and in some cases in the literature, appears to have induced a significant worsening of the patient's symptoms[10]. Therefore, the aggressive removal of this tumor is generally contraindicated. Moreover, the outcomes of surgery for the treatment of this condition normally turn out fairly poorly[15]. This might be due to the fact that the lipoma replaced neural tissue during embryonic development, resulting in the formation of fewer redundant spinal tracts. This would mean that the spinal cords of patients with this condition suffer from a congenital lack of physiological reserve[11,14]. Therefore, the decision of surgical treatment for spinal lipoma should be one which has been given very careful consideration. Surgical decompression is indicated for symptomatic lipomas[12]. However, if the patient is symptomatic, tumor debulking tends to be sufficient only to improve or stabilize the patient's neurological dysfunctions[11]. The primary purpose of surgery in cases of lipoma, then, is not total removal, but rather the decompression of the adjacent neural structures. The principal role of surgery in the treatment of intramedullary lipomas might be to stabilize and temporize the patient's clinical course[13]. As the majority of symptomatic patients do not improve after surgery, it might be advantageous for them to undergo early surgery, before the course of their disease debilitates them to a substantial degree[4,14,18,19].

Conclusion

Intramedullary spinal cord lipoma without spinal dysraphism can manifest in adulthood, usually as myelopathy and back pain. Here, we report a rare case of intramedullary lipoma without spinal dysraphism, and believe that this case report supports the notion that the meticulous microsurgical decompression of the intramedullary lipoma, under neurophysiological monitoring conditions, may prove effective with regard to the amelioration of the patient's neurological dysfunctions.

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