CASE REPORT

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Intramedullary Spinal Cord Lipoma Extending from the Cervicomedullary Junction to the Upper Thoracic Cord

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A case of intramedullary spinal cord lipoma is presented. A one month-old male infant presented with irritability and weakness on his upper extremities. A magnetic resonance(MR) image of the cervical spine demonstrated a well defined, high signal intensity lesion on both T1 and T2-weighted MR images and suppression on the fat saturation sequence. The tumor mass extended from the foramen magnum to T2 vertebra level. Ventral displacement of the spinal cord with kinking of the cervico-medullary junction was evident on the T2-weighted sagittal image. Partial resection of the tumor mass through laminoplastic laminotomy from C1 to T2 resulted in improved motor weakness on his upper extremities.

KEY WORDS : Intramedullary · Lipoma · Laminoplastic laminotomy.

Introduction

Spinal cord lipomas are rare lesions that constitute less than 1% of all intraspinal tumors[1,4-6,8-10,15]. Most spinal lipomas are extradural and associated with spinal dysraphism[2,6]. Pure intramedullary lipomas are even more infrequent in infants[3,7,8]. We report a case of spinal cord lipoma extending from foramen magnum to T2 vertebra, which was not associated with spinal dysraphism.

Case Report

This 36-day-old boy, who had been delivered by cesarian section due to a breech presentation, presented with a ten day history of irritability and progressive quadriparesis that was more pronounced in the upper extremities, especially in his right side. Physical examination revealed tenderness on his posterior neck and right shoulder. There was no bulging on the fontanelle. Skin stigmata such as hair, dimple or mass were not found.

Electrophysiological study revealed C8 and T1 radiculopathy. Magnetic resonance(MR) imaging of the spine demonstrated that a high signal intensity lesion was seen in the right posterolateral portion of the spinal canal and the spinal cord was displaced left ventrolaterally on axial T1-weighted fast spin echo image, which extended from the foramen magnum to the T2 vertebra on sagittal T2-weighted fast spin echo image. Cervicomedullary kinking was evident at the level of craniovertebral junction. The mass also showed a reduced signal on the fat saturation sequence (Fig. 1). It was not enhanced after administration of gadolinium. The mass was compatible with fat tissue. Progressive motor weakness made us to perform decompressive surgery.

Under general anesthesia, the patient was placed in the prone position. After a midline skin incision was made, exposure of the laminae were performed from C1 to T2. A midline splitting of the laminae from C1 to T2 was made following partial drilling of the laminae along the medial border of the facet joints (Fig. 2). The exposed spinal dura was moderately tense. The yellowish tumor mass bulged out after the dura opening. We attempted to debulk the tumor using CUSA (Aloka, SUS-201D) and bipolar electrocautery, but complete resection of tumor mass was difficult due to an indistinct margin. The tumor mass was removed only partially, and then split laminae were reconstructed from C1 to T2.

Pathologically, the tumor mass was composed of mature adipose tissues (Fig. 3). Postoperatively, the patient had uneventful recovery with slight improved motor weakness on

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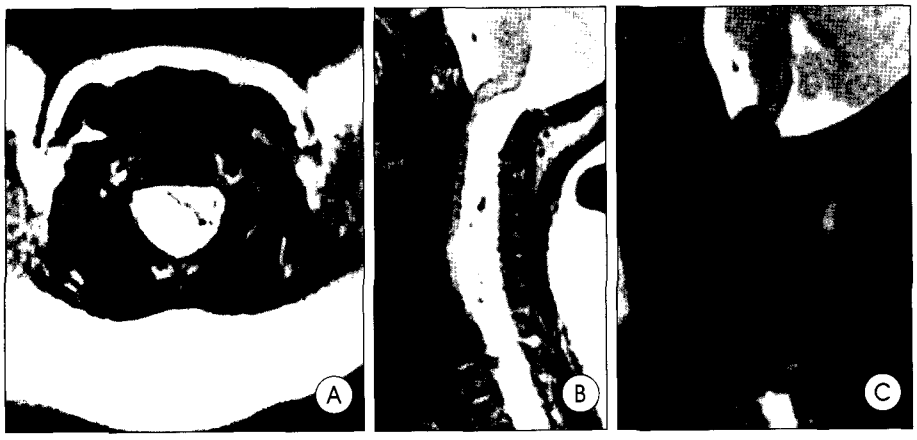

Fig. 1. A : The magnetic resonance im-aging of the cervical spine demonstrat-ing the dorsal intramedullary lipoma be-tween foramen magnum and T2 verte-bra. The tumor mass shows high signal intensity on both T1 (A) and T2 (B) weig-hted fast spin echo images, and reduc-ed signal intensity on T2 weighted fat sa-turation sequence (C). Spinal cord was severely compressed and displaced to left ventrolaterally by large lipoma.

Fig. 2. Intraoperative photograph showing the bulging yellowish tumor mass following dura opening.

his upper extremities. At one year follow up after surgery, the patient was doing well without cervical spinal instability.

Discussion

Spinal lipomas are rare lesions that occur more commonly at the lumbosacral spine, conus medullaris, and filum terminale and they are frequently associated with spinal dysraphism such as spina bifida occulta[5,6]. Cervical intramedullary lipoma in infancy is rare and its intracranial extension is extremely rare[3,7,8]. Unlike lumbosacral lipomas, cervical spinal lipomas are not associated with spinal dysraphism.

The pathogenesis of intramedullary lipomas remains unclear. Some authors believe the cause to be an embryologic inclusion of misplaced cell types during neural tube closure[3,5,10]. Ammerman, et al.[1], consider it to be of hamartomatous origin. Histologically, intramedullary lipomas consist of mature fat cells, separated by delicate connective tissue and admixed nerve bundles are often located at the periphery, suggestive of secondary entrapment of adjacent nerve roots[1].

Clinical manifestations of intramedullary spinal cord lipoma usually result from direct compression of the spinal cord rather than cord invasion by the tumors. Spinal axis pain, dysesthetic sensory changes, gait disturbance, weakness, numbness, and incontinence are the common presenting symptoms[1].

Concerning the timing of symptom onset, intradural lipomas have been noted to become symptomatic primarily during the first five years of life, or during the second and 3rd decades of life, and during the fifth decade of life[1,15]. Lee, et al.[10], in a series of six intramedullary lipomas, determined a mean age of 27years. Occurrence of symptoms largely depend on the degree of cord compression due to the growing lipoma. If an initial mass is large, then symptoms will present at an early age. Spinal lipomas without associated dysraphism have only

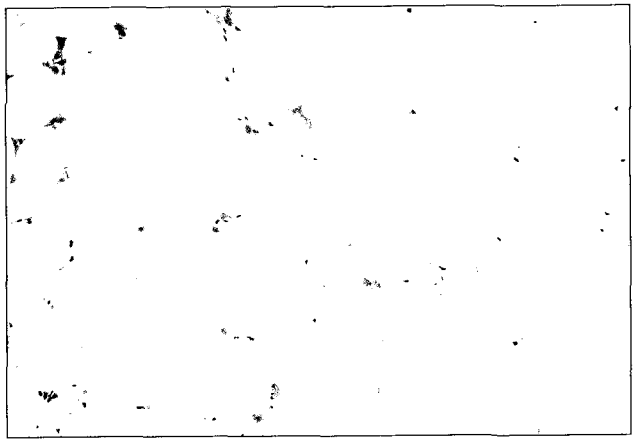

Fig. 3. Photomicrograph showing mature adipose tissue and intervening connective tissue (H & E, X100).

a small space for expansion, and thus an early presentation of the symptoms[8].

In our case, very early onset of symptoms can be explained by an exhausted physiological reserve of spinal cord probably due to the size of the lipoma and negative association to spinal dysraphism.

Diagnosis of spinal lipoma can be done by CT scan, but the most accurate diagnostic tool is known to be the MRI[10,14-16]. Lesions usually show high signal on T1 and low signal on T2-weighted images, but with the use of fast spin echo technique that is used to reduce scan time, the fat showed high signal intensities on both T1 and T2 weighted images. And with the use of fat suppression technique on T2 weighted image, the fat shows reduced signal intensity. MRI also has the advantage of visualizing the lipomaspinal cord interface well.

Management of intramedullary lipoma is controversial. Although McLone, et al.[12], propose early prophylactic intervention in only selected asymptomatic patients, because surgical intervention after onset of symptoms is unfavorable due to significant postoperative residual disability. But most authors postulate surgical decompression for symptomatic lipomas only. They recommend partial resection or only simply dural decompression by bone removal and dural grafting, because the infiltrative character of the tumor made total removal impossible[9,10,13]. Decompression of the spinal canal following debulking of the tumor mass using CUSA, can improve the surgical outcome. Total excision of lipoma may cause damage to the spinal cord and result in neurologic deficit. In our case, we could achieve neurological improvement after partial removal of the lipoma.

Kim, et al.[8], reported three cases of intramedullary lipoma, one of which had intracranial extension similar to our case. They postulated that laminoplastic laminotomy is an appropriate approach for decompression of an intramedullary lipoma because their patient was young and spinal stability and cosmetic result was important. In our case, we also performed laminoplastic laminotomy using midline splitting of laminae. Cervical spinal instability didn't appear at the last follow up of one year.

Pathology of lipoma shows mature adipose tissue, without evidence of malignancy or dedifferentiation[10]. Cells appear large, regular, polygonal, reticulated and optically empty with delicate cytoplasmic membranes and unremarkable eccenteric nuclei consistant with mature fat[11]. Macroscopically, they are soft, yellow, fusiform tumors with indistinct intraparenchymal margins[11]. In our case the tumor showed typical mature adipose tissues with intervening connective tissue compatible with lipoma.

Conclusion

We report a case of spinal intramedullary lipoma extending from the cervicomedullary junction to upper thoracic cord. Spinal cord decompression including partial removal of lipoma and duroplasty through laminoplastic laminotomy might be an appropriate surgical approach to avoid postoperative spinal instability.

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