Cervical Intradural Lipoma with Associated Hemivertebra Formation at C6 Level: A Case Report

C6 Düzeyinde Hemivertebra Oluşumu ile Beraber olan Servikal İntradural Lipom : Olgu Sunumu

ABSTRACT

INTRODUCTION and CASE DESCRIPTION: Intramedullary lipomas are rare tumours of the spinal cord and account for about 1% of all spinal neoplasms. These lesions can occur anywhere along the length of spinal cord, but are frequently localized to the lower thoracic and lumbosacral levels. The authors present a 18-year-old female with intractable shoulder and neck pain and progressive weakness in the upper extremities, harbouring a cervical intradural lipoma with intramedullary extension, along with concomitant scoliosis.

CONCLUSION: Despite its benign nature, surgical treatment of these lesions in symptomatic patients generally provides satisfactory relief of symptoms. Radical removal of spinal intradural lipomas is not recommended since attempts at complete excision carry an unacceptable risk of postoperative morbidity and sufficient decompression with or without duraplasty generally provides a successful clinical outcome.

KEY WORDS: Intradural lipoma, Benign, Intramedullary, Surgical decompression, Hemivertebra

ÖZ

GİRİŞ ve OLGU SUNUMU: İntramedüller lipomlar omuriliğin nadir tümörlerindendir ve tüm spinal tümörlerin yaklaşık %1'ini oluştururlar. Bu lezyonlar omurilik uzanımı boyunca her seviyede görülebilmelerine karşın sıklıkla alt torakal ve lumbosakral bölgeye lokalizedirler. Bu olgu sunumunda yazarlar, şiddetli omuz ve boyun ağrısı ve ilerleyen üst ekstremite güçsüzlüğü ile başvuran ve skoliozun eşlik ettiği intramedüller uzanımlı servikal lipomu bulunan bir olgu sunmaktadır. Sonuç: Selim gidişatlı olsalar da bu lezyonların semptomatik hastalarda cerrahi olarak çıkartılması genellikle semptomları giderme açısından tatminkar sonuçlar sağlamaktadır. Spinal intradural lipomların radikal olarak çıkartılması önerilmemektedir zira, total eksizyon girişimleri kabul edilemez cerrahi sonrası morbidite riski taşımaktadır ve duraplastili veya duraplastisiz yeterli dekompresyon genellikle başarılı klinik sonuç sağlamaktadır.

ANAHTAR SÖZCÜKLER: İntradural lipom, Selim, İntramedüller, Cerrahi dekompresyon, Hemivertebra

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Received: 07.01.2008 Accepted: 05.04.2008

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INTRODUCTION

Spinal cord lipomas are benign lesions, accounting for less than 1 % of all spinal tumours (11, 15). The most common site of involvement is the lumbosacral region, in which the lipoma is found as a component of a spinal dysraphic state (6, 8, 14, 21). Intradural spinal lipomas of the cervical and upper thoracic region are rarely encountered (6, 17, 21). Unlike lumbosacral lipomas, cervical spinal lipomas not associated with dysraphism are even rarer (9, 15). We report a patient harbouring an intradural lipoma with an intramedullary extension associated with scoliosis and hemivertebra formation. We present the clinical presentation, neurological examination, the diagnosis and treatment options for this rare entity.

CASE REPORT

An 18-year-old female was admitted to our department with a six-month history of neck pain and progressive weakness in her upper extremities, especially in finger adduction. Neurological examination revealed mild paresis in the upper extremities and increased deep tendon reflexes. Plain X-rays and magnetic resonance imaging (MRI) of the cervical spine revealed scoliosis and hemivertebra formation at the level of C6 (Figure 1). Cervical MRI of the spine showed an intradural lesion with an intramedullary extension at the C3 (Figure 2). The patient underwent decompression of the spinal cord via C3 laminectomy. Following the dural incision, a yellowish intradural mass extending into the spinal cord was detected. The greater bulk of the intradural mass showed typical features of a spinal lipoma, including its tight adherence to the spinal cord and lack of cleavage planes that prevented a clear separation of the lesion from the spinal cord. A subtotal microsurgical resection was performed with sufficient spinal canal decompression. The postoperative period was uneventful and the patient was discharged on the 3rd postoperative day without additional neurological deficit. The histopathological examination revealed that the lesion was uniformly composed of mature adipose tissue and the diagnosis was consistent with a lipoma. The patient was free of symptoms except for mild neck pain with a normal neurological examination at her 6-month follow up.



Figure 1: Coronal T-1 weighted cervical MRI section shows scoliosis and hemivertebra formation at the C6 level (arrow).

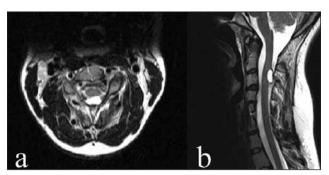


Figure 2: Axial (A) and sagittal (B) T-2 weighted MRI sections reveal an intradural spinal mass with intramedullary extension. Note the tissue plane between the spinal cord and the mass in figure 2b suggesting its intramedullary extension.

DISCUSSION

Intradural intramedullary lipomas are histologically benign neoplasms located mostly in the lower thoracic and lumbosacral levels (22). Those located at the cervical region are very rare. Excluding the lumbosacral spine, the distribution of the location of lipomas has been reported to be thoracic in 32%, cervicothoracic in 24% and purely cervical in only 13% (19). The peak incidence is in the first 5 years of life and another peak is seen in the fifth decade (13, 18).

The symptoms and signs depend on the location of the tumor. As they most frequently involve the posterior part of the spinal cord, sensory deficits and gait disturbances predominate in the clinical presentation (13, 14, 16). Localized pain and/or motor deficits in the extremities can also be the presenting symptoms (3, 14). Brown-Séquard syndrome and signs of long tract involvement, such as clonus, hyperreflexia and Babinski sign are also common but likewise nonpathognomonic (1). The symptoms may be long-standing and progression may be slow (13, 18). This is particularly true for the cervical area as it is reported that more than 80% of patients with a cervical intradural lipoma displayed symptoms for more than 10 years (12, 14). In our case, the patient presented with localized shoulder and neck pain and progressive motor deficit in both upper extremities. Interestingly, she had no sensory deficit. Moreover, her symptoms had started 6 months prior to her admission, a relatively short period when the literature is considered.

Similar to the majority of spinal lesions, MRI is the preferred method for radiological evaluation of intradural lipomas. On MRI, intradural lipomas have a hyperintense signal on T1-weighted images and a hypointense (1, 22) or hyperintense (17, 23) signal on T2-weighted images. Fat is distinguished by using the fat suppression effect from other lesions that are also hyperintense on T1-weighted images. In such cases, the high signal will characteristically drop when the fat suppression effect is added to the imaging sequence (5). Both computed tomography (CT) and MRI can reveal the fat component, but MRI is superior to CT in demonstrating the relationship with adjacent normal neural tissue (1, 5). CT of the spine with three-dimensional or high-quality multiplanar reconstructed images might be helpful for presurgical planning, especially in cases where there is associated spinal dysraphism or scoliosis (1, 7).

Although considered to be a primary intradural spinal tumor in most neurosurgical sources (5, 22), some authors believe that spinal lipomas may be classified under hamartomas rather than true neoplasms (16). The observation of an increase in the size of lipomas parallel to weight gain in some patients, and in pregnant women, and the frequent association with other malformations, such as spina bifida support this concept (16, 21). However, rapid lipoma growth despite meticulous diet control (4) contradicts these findings. Moreover, lipomas can coexist with lesions that may contain potentially neoplastic cells (teratomas, dermoid cysts) and

definite histopathological diagnosis is therefore important as the natural course and treatment strategies may differ in these subgroups (2).

The appropriate surgical management of intraspinal lipomas remains controversial (16, 21). The need for prophylactic surgery in asymptomatic patients, especially in pediatric cases, has been a matter of debate. It has been suggested that prophylactic surgery is warranted for lipomas of the filum terminale, but not for patients with lipomas attached to the conus medullaris (21). This assumption can also be applied for lipomas involving the thoracic and the cervical region (16). In our case, the patient had progressive neurological deficit presumably due to spinal cord compression and/or intramedullary extension of the lesion. The symptoms completely resolved following satisfactory decompression within 6 months postoperatively. We concur with other authors that prophylactic surgery for asymptomatic patients is not recommended for intradural spinal lipomas due to the benign course of such lesions. However, early surgery may prevent the development of a permanent neurological deficit in symptomatic patients.

The main purpose of surgery in spinal lipomas is to perform a decompressive procedure that decreases the bulk of the lipoma (4, 10, 11, 12, 16, 20). Aggressive surgical removal can be associated with significant postoperative morbidity and should not be attempted as these lesions usually do not have cleavage planes that clearly separate them from the spinal cord and nerve roots (3, 16, 20, 23). Duraplasty was not done in our case because a sufficient decompression of the spinal cord and nerve roots and free passage of the CSF were achieved by partial removal of the tumor.

CONCLUSION

Spinal lipomas are benign tumors that grow very slowly and may change size according to alterations in the body fat level. Although the symptoms are generally slowly progressive, the time interval between the appearance of the initial symptom and admission for treatment may be much shorter for cervical intradural lesions. Surgery should be performed in symptomatic cases and adequate decompression rather than total removal should be the main aim in spinal intradural lipomas.

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