Thoraco-lumbar nondysraphic intramedullary spinal cord lipoma with an unusual aspect on MRI: case report and review

Lipoma intramedular tóraco-lombar não associado a disrafismo espinhal com aspecto pouco usual à RM: relato de caso e revisão

RESUMO

Lipomas intramedulares são tumores raquidianos muito raros, sendo a maioria associado a disrafismos espinhais. Neste artigo relatamos o caso de um paciente oligossintomático, apresentando um lipoma intramedular tóraco-lombar não associado a disrafismo espinhal, com um sinal bifásico pouco usual na RM ponderada em T1, e submetido a biópsia microcirúrgica. Aspectos de origem, evolução, diagnóstico e tratamento são discutidos.

Palavras Chave: lipoma intramedular; RM.

ABSTRACT

Intramedullary spinal cord lipomas are very rare spinal tumors, most of them related to spinal dysraphism. We report the case of an oligosymptomatic patient, presenting with a thoraco-lumbar nondysraphic intramedullary spinal cord lipoma, with an unusual biphasic sign on T1-weighted MRI, and submitted to microsurgical biopsy. Features of origin, evolution, diagnosis and treatment are discussed.

Key Words: intramedullary lipoma; MRI.
INTRODUCTION

Intramedullary spinal cord lipomas are very rare entities, most of them linked to dysraphic spinal cord states. They comprise about 1% of all spinal cord tumors and the incidence of these tumors without association to spinal dysraphism is even smaller (0.6%). The first report was provided by Gowers, in 1876. The most common topography is the lumbosacral region, in which the lipoma is found as a component of a spinal dysraphic state. Most spinal lipomas originate in the dorsal juxtamedullary topography of the spinal cord. However, the embryologic defect that leads to the development of these tumors is unknown. The most common accepted theory is that a developmental malformation occurs during the formation of the neural tube and leads to inclusion of embryonic crests of fat cells. Lipomatous fat of these tumors is metabolically similar to systemic adipose tissue, so the onset of symptoms is associated with body weight gain. The most widely accepted classification divided spinal lipomas in dorsal, transitional, and terminal lipoma, defined with regard to the relationship with the cord, conus medullaris, or filum terminale. Regarding the treatment for intradural lipoma, there is controversy. Concerning surgery, few authors have advocated aggressive surgical removal of spinal lipomas. In this article, we report a nonsymptomatic thoraco-lumbar nondysraphic intramedullary spinal cord lipoma with an unusual biphasic sign on T1-weighted MRI, and literature is reviewed.

CASE REPORT

A previous healthy female patient, 42 years-old, complained of progressive lumbar pain with bilateral leg pain, paresthesias and occasional numbness in both legs. Neurological examination was normal. Thoraco-lumbar magnetic resonance imaging (MRI) disclosed a T12 to L3 biphasic intramedullary space-occupying lesion in T1-weighted sequences, slightly hyperintense in its greater extension, with a prominent hyperintensity in the superior part (fig. 1 and 2), and iso to hypointense in T2-weighted sequences (fig. 3). She was submitted to a T12-L1-L2 laminectomy and, after dural opening, a soft yellow tumor emerging from the spinal cord was found (corresponding to the hyperintense superior portion), and an inferior tissue slightly different from fatty tissue, which was biopsied using microsurgical technique. Histopathological examination revealed fatty cells tissue, characterizing a lipoma (fig. 4). In the follow-up, the patient remained without neurological deficits and was referred to rehabilitation care.
There is considerable controversy in the literature regarding intramedullary spinal cord lipomas terminology, and a proper classification is long overdue, because congenital spinal lipomatous malformations constitute a diverse group of lesions. These malformations constitute a wide spectrum of lesions ranging from relatively simple lipomas of the filum terminale to complex malformations, and differ from one another in their embryology, clinical presentation, operative strategies, complications, and prognosis. Failure to differentiate between the different forms of congenital spinal lipomatous malformations may lead to inaccurate assumptions regarding prognosis and inappropriate management. “True” intramedullary lipomas are defined as those not associated with spinal dysraphism. Its origin is unclear, but it is postulated that they arise from mesenchymal cells migration from the ectoderm, before closure of the neural tube during the embryonic period. These mesenchymal cells instead of differentiating themselves into duramater, differentiate into adipose tissue. However, apparently another differentiation mechanism is involved because, in these tumors, the duramater and the posterior elements of the spine are normal. Other entities, such as the filum-terminalis lipoma and caudal lipoma are formed by a similar mechanism to the intramedullary lipomas and, in addition, they also have dural and posterior elements integrity. Therefore, they were recently classified as Group I of Congenital Spinal Lipomatous Malformations. The neurological signs depend on size and localization of the tumors. The slow evolution with periods of clinical worsening, interspersed with periods of remission, seems to be related to metabolism of systemic body fat. Periods of excessive weight gain, or treatment with corticosteroids, with consequent systemic fat accumulation, appear to be associated with neurological worsening. Agraharkar et al. described a case of rapid growth of intramedullary lipoma (six weeks), in a patient submitted to renal transplantation, associated with the use of corticosteroids. Regarding location, the thoracic spine seems to be more involved. When located in the cervical region, it may extend into the intracranial space, and a case of massive intracranial extension with subsequent development of hydrocephalus was reported. There is also a report of migration of intramedullary lipoma in a patient presenting with symptoms of migratory myelopathy. As intramedullary lipomas present with nonspecific symptoms of compromise of the spinal cord, diagnosis can only be confirmed by MRI. Diagnostic characteristics of these tumors are hyperintense signal appearance in T1 and hypointense on T2, showing the typical signs of fatty tissue, usually confirmed by fat suppression sequence. In this case we found a biphasic aspect of MRI signal, so we indicated biopsy based on slightly hyperintense T2-weighted of the inferior portion of the tumor. Regarding surgical treatment, those patients clinically asymptomatic or oligosymptomatic should be monitored or submitted to biopsy for diagnosis, when MRI signal is...
suspicuous, according to this present case\textsuperscript{29}. Radical removal of spinal intradural lipomas is not recommended as attempts at complete excision carry an unacceptable risk of postoperative morbidity, and sufficient decompression with or without duraplasty generally provides a successful clinical outcome\textsuperscript{24}. A 6\% deterioration rate after surgical treatment is reported for symptomatic patients, and improvement rate of 44\%, but complete resolution of symptoms was observed in only 14.2\%: the reported low rate of postsurgical worsening indicates that surgery for spinal lipomas are safe, and operations performed after the onset of symptoms seldom cure the patients, so this results support early untethering for any kind of symptomatic spinal lipoma\textsuperscript{16}. Another study concluded that intramedullary tumors of the cervical spinal cord are amenable to total surgical removal and surgery is suitable when a patient presents with a moderate neurologic deficit, but proficient surgical technique for total tumor resection is necessary for good results and preoperative radiotherapy contributes to difficult surgery and poor prognosis, and is not recommended\textsuperscript{13}. However, this is usually impossible because the separation of the tumor from the neural tissue is associated with significant postoperative morbidity and the intimate relationship of the lipoma to the nerve roots and the absence of a distinct plane between tumor and spinal cord precludes a complete tumor resection to prevent further progression of symptoms and to offer the possibility of neurologic improvement is mandatory\textsuperscript{29}. Excisional surgical treatment should be reserved to the cases associated with myelopathy\textsuperscript{3}. In this latter case, the objective is not total removal, but decompression of neural structures. Long-term results of this kind of treatment is satisfactory\textsuperscript{15,23}.

**CONCLUSION**

Even though intradural spinal lipomas are not a frequent spinal space-occupying lesion, they are associated with varied neurologic deficits. For asymptomatic or oligosymptomatic patients, microsurgical biopsy is occasionally necessary for diagnosis, and for patients with neurologic deficits early surgical decompression without attempts for complete excision is an ideal therapeutic option. Serial MRI for follow-up is mandatory for all patients.
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Relato de caso


