

Case Report / Olgu Sunumu

MRI appearance of epidural and paraspinal lipomatosis in a child with hemihypertrophy: case report

Hemihipertrofisi bulunan bir çocukta epidural ve paraspinal lipomatozisin MRI bulguları: olgu sunumu

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Abstract

Diffuse lipomatosis is a rare condition characterized by overgrowth of mature adipose tissue that grows in an infiltrative manner. It most commonly involves large portions of a limb or the trunk. In this report, we present the magnetic resonance imaging findings in a 14-year-old boy with diffuse infiltrating lipomatosis, epidural lipomatosis and lower extremity hypertrophy.

Keywords: Diffuse lipomatosis, epidural lipomatosis, hemihypertrophy, magnetic resonance imaging

Özet

Diffüz lipomatozis matür adipöz dokunun infiltratif şekilde aşırı büyümesi ile karakterize nadir görülen bir durumdur. Yazımızda sağ torakolomber bölgede infiltratif tipte intramüsküler lipomatozis, epidural lipomatozis ve alt ekstremitelerde hemihipertrofi saptanan 14 yaşındaki erkek hasta, manyetik rezonans görüntüleme bulguları eşliğinde sunmaktayız.

Anahtar sözcükler: Difüz lipomatozis, epidural lipomatozis, hemihipertrofi, manyetik rezonans görüntüleme

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Atalar et al. Lipomatosis.

142

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Introduction

Lipomatosis, a classic example of an infiltrating fatty tumor, is a rare condition characterized by diffuse infiltrating overgrowth of mature adipose tissue with ill-defined borders. It usually involves a large portion of an extremity or the trunk. In some cases, there are distant lipoma or angiomas, or hypertrophy of subjacent bone [1, 2]. It can be associated with obesity, steroid ingestion, or Cushing's syndrome or may be idiopathic [3]. Hemihypertrophy is best defined as asymmetry between the two sides of the body to a greater degree than can be attributed to normal variation. It is a congenital entity caused by an asymmetrical overgrowth of a body part. It can occur as idiopathic or as a part of several recognized clinical syndromes that have other visible cutaneous or vascular anomalies, including Proteus syndrome, Klippel-Trenaunay syndrome, neurofibromatosis, and Beckwith-Wiedemann syndrome [4, 5].

In this report, we present the magnetic resonance imaging findings in a 14-year-old boy with diffuse infiltrating lipomatosis of thoracolumbar region of back, epidural lipomatosis involving the entire thoracolumbar spine and lower extremity hemihypertrophy without infiltrating lipomatosis as an unusual example to infiltrating lipomatosis.

Case

A 14-year-old boy presented with the complaint of an increase in the size of swelling in his back and bending of his spine. His family reported that his right leg from thigh to foot has been larger compared to the left leg since his birth, and that lipomatosis was previously diagnosed with a biopsy of mass in his back. On examination, a soft mass involving thoracolumbar region was palpable. The boundaries of the lesion were ill defined and it was non-pulsatile. A scoliosis with a spinal curve to left was observed. Right leg was larger than left leg. There was no abnormality in other clinical and laboratory tests. His mental and motor development was normal. There is no relevant family history. No history of chronic steroid use, obesity, Cushing's syndrome, and other endocrine disorders was found.

MRI on a 1.5-T scanner showed a nonenhancing, ill-defined, infiltrative, inhomogeneous, hyperintense lesion (isointense to fat) on T1W images in the subcutaneous, muscular, and intermuscular planes of the thoracolumbar region; the lesion appeared isointense to fat on T2W images. A radiological diagnosis of congenital infiltrating lipomatosis of the trunk was made.

MRI scanning revealed diffuse subcutaneous lipomatosis involving thoracolumbar area. Paraspinal muscles, especially the erector spinae group, had diffuse fatty infiltration. MRI scanning also showed epidural lipomatosis involved the entire thoracolumbar spine. Conus medullaris terminated at L5 level and there were findings of tethered cord syndrome. On right side, there were hypertrophic changes in the posterior parts of the thoracolumbar vertebrae. On MRI of the pelvis and legs, there was no abnormality in fat tissue and vascular and bony structures except hypertrophy in the muscles of right pelvis and extremity.

Atalar et al. Lipomatosis.

143

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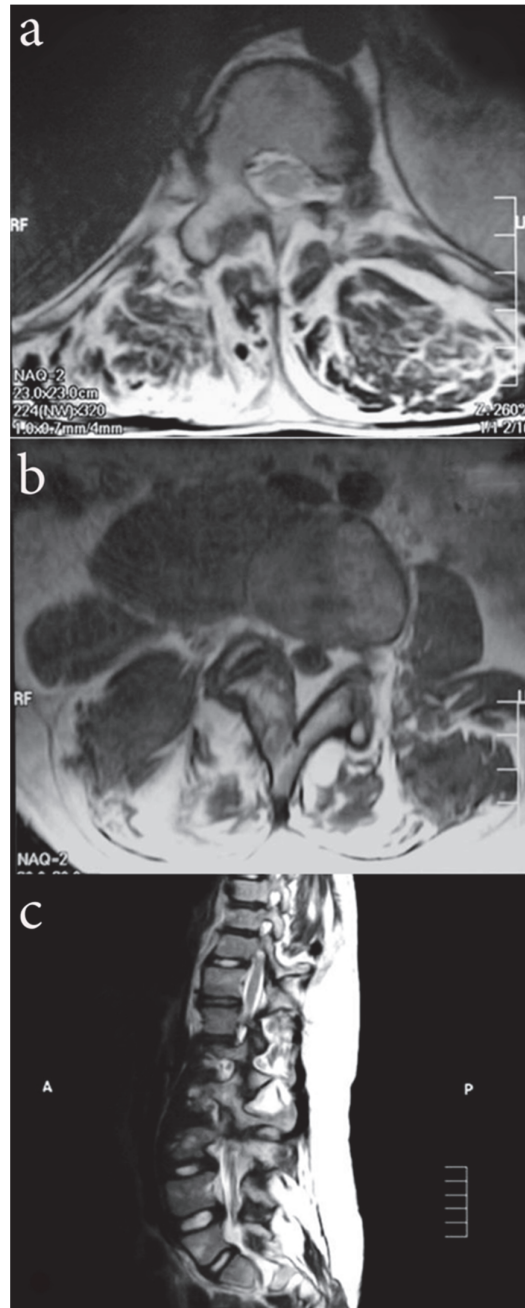


Figure 1. Axial T1W images show fat-intensity in paraspinal muscles and epidural region and hypertrophy of right psoas muscle and vertebral posterior elements (a, b). Sagittal T1W image shows lipomatosis in thoracolumbar subcutaneous region with stretched spinal cord and rotoscoliosis (c).

Atalar et al. Lipomatosis.

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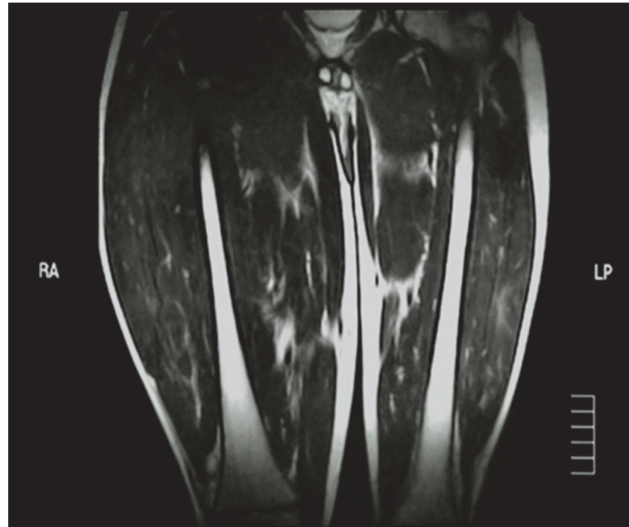


Figure 2. Coronal T1W image shows prominent hypertrophy of soft tissues of right thigh compared to left side.

Discussion

There are several types of lipomatous lesions presenting quite commonly as the largest single group of mesenchymal tumors. Diffuse lipomatosis is a rare subtype of lipomatous lesions and characterized by diffuse overgrowth of mature adipose tissue infiltrating through the adjacent soft tissues of the portion of the affected extremity or body trunk. Microscopically, this lesion is indistinguishable from lipoma [5, 6]. The radiological findings in lipomatosis include the presence of fatty tissue diffusely distributed within and between the involved muscles. Diffuse lipomatosis is generally diagnosed during the 2 years of age and generally involves trunk and limbs but it can be located any place in the body such as thyroid gland, tongue, heart,

Spinal epidural lipomatosis is a rare condition that can present with the hypertrophy of adipose tissue located in the spinal epidural space. It is most commonly associated with long-term steroid use but can be observed in patients with a number of other conditions. As the adipose tissue enlarges, it encroaches on the spinal canal and compresses the neural elements [7]. There are a few reported cases with similar imaging findings to our case [8-10].

Radiological findings is in accordance with the pathologic process, there are diffuse increase and infiltration of adipose tissue in the involved area. The tumoral tissue in diffuse lipomatosis is not well-circumscribed and homogenous. In affected areas, there is diffuse swelling of the soft tissues and osseous overgrowth and deformity may be observed.

The differential diagnosis often included are the Proteus syndrome (postnatal growth of multiple tissues such as linear epidermal naevus, vascular malformations, and lung cysts),

Atalar et al. Lipomatosis.

145

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encephalocraniocutaneous lipomatosis (brain abnormalities), hemihyperplasia-multiple lipomatosis syndrome and Bannayan syndrome (microcephaly, postnatal retardation, and mesodermal hamartomas may be found).

In conclusion, diffuse lipomatosis is an unusual clinical condition with atypical clinical presentations. In addition, based on the findings in this particular case, we think that the association between diffuse lipomatosis and hemihypertrophy emphasizes the requirement of MRI scanning in the diagnostic workup of in children with diffuse lipomatosis. MRI scanning provides detailed information about the involved structures and organs and developed complications. These lesions can be diagnosed based on the clinical and imaging features; however, the definitive diagnosis of diffuse infiltrating lipomatosis is established by histological examination, as a number of other conditions can simulate the clinical presentation.

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