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★ 2001 年 "百种中国杰出学术期刊"获奖证明

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★ 本刊 2003 年 "继教园地" 通知

本期 705～725 页文章为本刊 "继教园地"，项目编号：(2003) 中华医学会继教备字第 001 号。

自学时间：每期自学 6 学时，每一学习单元 12 期，需自学 72 学时。

学分授予方法：答题并经审卷及格者，可授予中华医学会继续医学教育Ⅱ类学分；每期 1 分，12 期 12 分为一单元。

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咨询电话：024－22714435；024－22714352　联系人：吕明超

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Value of MRI in the evaluation of lipid storage myopathy

Zheng Xianying 郑贤应, Murong Shenxing 慕容慎行, Li Yinguang 李银光, Ni Xihe 倪希和, Cai Jiangping 蔡江萍

Zheng Xianying, Murong Shenxing, Li Yinguang, Ni Xihe, Cai Jiangping, Intraneural Department, First Affiliated Hospital, Fujian Medical University, Fuzhou 350001, Fujian Province, China

Abstract

AIM: To investigate the magnetic resonance imaging (MRI) findings of lipid storage myopathy (LSM), evaluate the clinical value of MRI for LSM. **METHODS:** Eight cases with LSM biopsy-proved underwent MRI of pelvis and thigh, spin echo sequence, fast spin echo sequence and short tau inversion recovery (STIR) were utilized. **RESULTS:** The signal intensity of diseased muscle of 8 cases with LSM was hyperintense on T2-weighted images (T2WI) and T1-weighted images (T1WI), and hyperintense signal intensity on T2WI and iso-intense on T1WI was seen on 2 patients as well. **CONCLUSION:** MRI can provide objective data for clinical diagnosis, therapeutic evaluation and follow-up. It can also help to decide the accurate localizations for biopsies.

SUBJECT WORDS: lipidoses; muscular disease; magnetic resonance imaging

INTRODUCTION

Lipid storage myopathy belongs to inflammation of muscle cells caused by self immunity. (lipid storage myopathy, LSM). Domestic researches about its evaluation with MRI were unavailable. Our objective was to investigate MRI findings of LMI and its value in the evaluation of LSM.

MATERIALS AND METHODS

Materials

9 LSM patients admitted from April 1998 to April 2002 were included in our study. These patients include 5 men and 5 women aging 4－36 years (mean age: 24. 9 years). Course of disease varied from 9 months to 12 years, averagely 6. 8 years. 1 subject suffered from the first onset and the remaining were the recurrent cases. All patients had myasthernia clinically and were relived in 3 cases 2－4 years ago. Creating phosphokinase (CPK) were 217－3 297 IU/L, averagely 950 IU/L (reference value: 25－200 IU/L), lactic dehydrogenase (LDH) 303－3 080 IU/L, averagely 1 132 IU/L (reference value: 109－245 IU/L). Electromyography showed myogenic diseases. Diagnosis was confirmed pathologically.

Methods

Scanning instrument superconductive that is MRX machine Flexart 0. 5 T was provided by Toshiba Company. Positive alternating body and tail coils were used. All patients received axis scanning of pelvis, bilateral legs and thighs and paralled examination, no enhanced scanning was performed. Scanning sequences included self echo T1WI (TR 600 ms, TE 30 ms), fast self echo T2WI (TR 3 000 ms, 80 ms), short transverse recovery T2WI (TR 2 000 ms, TE 30 ms, TI 150 ms). Layer was 10 mm in thickness, and interval was 10 mm. Vision field was 35 cm × 35 cm, and matrix was 198 ×256.

RESULTS

Positive findings were observed in 7 of 8 patients receiving examination. T2WI and T1WI showed spot or slice like signal. Signal of high density in STIR sequence magnified as that of low density such as extensive lesions, unclear boundary, and symmetry. Posterior muscles of thigh and adductor muscle groups showed T1 and T2 signal suggesting necrosis of muscle fibers. No positive findings was observed in 1 recovered patient with course of disease of 2 years. Hip muscles, posterior muscles of thigh, and adductor muscles groups were all affected in 7 patients receiving examination.

Signal of the greatest gluteal muscle and biceps muscles of thigh were abnormal in 7 cases, signal of quadriceps was abnormal in 5 cases. No affected iliopsoas muscle was observed. musculus vastus intermedium was affected slightly, and the greatest gluteal muscle was affected seriously.

DISCUSSION

LSM is resulted from disorder of fat metabolism of muscular fibers, and hence leading to fat accumulation. It often showed acute or subacute onset and is characterized by myasthernia of proximal end of limbs with or without myopain, myogenic lesions confirmed by EMG, muscle enzyme increase, course of recurrence and relief. Definite diagnosis is available after pathological examination. MRI showed that signal of normal muscles was located between bone cortex and subcutaneous fat and is mediate in intensity. For normal muscles, T2WI showed signal of high intensity similar to strips representing connective tissue filling space, while T1WI showed arc like signal of high density representing the little amount of fat locating covering of muscles.

Muscles with LSM lesions showed 2 types of signals, short T1 + long T2, and T1 + T2, the former suggesting fat storage myopathy, and the latter suggesting necrosis of muscular fibers and magnifying as LSM. Necrosis is usually located in posterior muscles of thigh and adductor muscle groups and especially common among patients of acute phase. It is different from results of clinically pathological examination. Pathological examination found no necrosis of muscular fibers[1-2] and was probably due to differences in course of disease and tissue acquirement. Lesions of the greatest gluteal muscle, adductor muscles groups, and posterior muscles of thigh were especially severe.

PM is similar to LSM in clinical symptoms and should be differentiated from it. MRI showed two kinds of signals for PM, inflammation and edema signal of long T1 and T2 and fat singal of short T1 and long T. MRI is incapable of differentiating local necrosis and inflammatory infilatration. MRI findings of PM is characterized by inflammatory edema signal, fat lesions is only can be observed in advanced cases. MRI findings of LSM is characterized by fat accumulation, and local muscles fibers is only can be observed in acute patients. Fat accumulation is severe in greatest gluteal muscles, and moderate in posterior muscle thigh and adductor muscles, for LSM, and mild in quardriceps, and necrosis signal is observed in posterior muscles of thigh and adductor muscles in flammatory edema signal can be observed in all muscles and is extensive, and basolamina may be affected, some fat signal is shown in posterior muscles of thigh.

Patients with long course of disease showed atrophy, and slightly affected quadriceps. Lesion range and locus can be observed, inflammation, edema, and fatty degeneration be compared by using MRI. Distribution and percent of lesions will provide basis for diagnosis of LSM. In addition, MRI will provide intrstruction for muscle biopsy and EMG examination.

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