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Original research article

Clinicopathologic features of sporadic inclusion body myositis in China

Ke Li [a], Chuanqiang Pu [b,*], Xusheng Huang [b], Jiexiao Liu [b], Yanling Mao [b], Xianghui Lu [b]

[a] Department of Geriatric Neurology, Chinese PLA General Hospital, Beijing, China
[b] Department of Neurology, Chinese PLA General Hospital, Beijing, China

ARTICLE INFO

Article history:
Received 20 October 2014
Accepted 8 June 2015
Available online 17 June 2015

Keywords:
Sporadic inclusion body myositis
Rimmed vacuole
Inclusion body granule
Chinese population

ABSTRACT

This study is to investigate the clinical and pathologic features of sporadic inclusion body myositis (sIBM) in China. We retrospectively evaluated the clinical and pathological features of consecutive patients in our department between January 1986 to May 2012. Total 28 cases of sIBM (20 males, 8 females, mean age was 56.93 ± 8.79) were obtained by review of all 4099 muscle biopsy reports. The proportion of sIBM was 0.68% (28/4099) in China. Muscle weakness of quadriceps appeared 100% in 28 cases, while conspicuous atrophy of quadriceps appeared only in five cases (17.86%). Creatase values of 28 patients with sIBM were normal or mildly elevated. Muscle biopsies showed that atrophic fibers resembled more frequent in small angular and irregular shape (82.14%), less common in small round shape (17.86%). Rimmed vacuoles resembled crack (67.86%) and round (32.14%) shape. Mononuclear cell invasion into necrotic muscle fibers (35.71%) was more frequent than non-necrotic muscle fibers (7.14%). sIBM was still a rare disease in China compared to other countries. There were some certain specific pathological characteristics existed in Chinese sIBM patients.

1. Introduction

Sporadic inclusion body myositis (sIBM), a progressive inflammatory skeletal muscle disease, occurs mainly in over 60-year-old patients [1]. The clinical and pathological features of sIBM were available in many countries, but did not reported in China. Since technique of enzyme-histochemistry method in muscle biopsy was not popularly for myopathy, few sIBM cases were studied and reported in China [2]. Meanwhile, there was also rare epidemiological data of sIBM in the world. The prevalence rate was reported differently among different countries and ethnic groups [3]. And the epidemiological data of sIBM in China was not available yet.

To study the clinical and pathological features of sIBM in China, we investigated the clinical manifestations, serological examination, electrophysiology, and muscle pathology in 28 sIBM cases. In addition, we surveyed all suspected myopathy patients who were subjected to muscle biopsy in our department. And we calculated an indirect epidemiological prevalence rate of sIBM in China.

* *Corresponding author at*: Department of Neurology, Chinese PLA General Hospital, 28 FuXing Road, 100853 Beijing, China. Tel.: +86 10 66939721; fax: +86 10 66939721.
E-mail address: pucq30128@sina.cn (C. Pu).
http://dx.doi.org/10.1016/j.pjnns.2015.06.004

2. Patients and methods

2.1. Patients

From January 1986 to May 2012, a total of 4099 patients with suspected myopathy around China were subjected to muscle biopsies in the neuropathology laboratory of our hospital. 28 patients were diagnosed as sIBM. All their medical records, electromyography (EMG) reports and muscle histopathology were collected. The muscle biopsies sites in this study were quadriceps and/or biceps. All 28 patients did not have dysphagia. The diagnose depended on 2011 European Neuromuscular Centre diagnostic criteria for inclusion body myositis [4].

2.2. Data collection

Study protocol data included: (1) detailed general history: gender, age at onset, initial symptom or sign, clinical course, family history; (2) muscle power: manual muscle testing (extremities, facial muscles, oculorotatory muscles), pharyngeal portion muscles, distribution and degree of muscle weakness and atrophy, progression pattern; (3) laboratory data: serum creatine kinase, nerve conduction velocity, muscle biopsy and other examination.

2.3. Histopathologic study

Muscle samples were frozen in liquid nitrogen immediately after removal and stored at −80 °C. Transverse serial frozen muscle sections were stained with hematoxylin and eosin (H&E), periodic acid-Schiff (PAS), oil red O (ORO), Gomori trichrome stain, nicotinamide adenine dinucleotide dehydrogenase (NADH), nonspecific esterase (NSE) and adenosine triphosphatase (ATPase) staining after incubation in pH 4.3, 4.5 and 10.6. Morphometric evaluation of muscle specimens was performed by light microscopy. Ultrathin sections were prepared in epon-embedded material, stained with osmic acid and examined by an electron microscope.

2.4. Statistical analysis

Dichotomous variables were analyzed by chi-square test; continuous variables were presented as mean ± SD and analyzed by Mann–Whitney *U*-test. All statistical tests were two-tailed. $P < 0.05$ was considered as statistically significant. In this study, all statistical analysis was performed by SPSS13.0 statistical software.

3. Results

3.1. Prevalence and onset age

We analyzed 28 patients with sIBM in our department from January 1986 to May 2012. The results showed that the proportion of sIBM was 0.68% (28/4099) in this group.

Among all 28 patients with sIBM, there were 20 male patients (71.43%) and 8 female patients (28.57%). The onset age ranged from 38 to 71 years (51.5 ± 7.3 years). The diagnosis age ranged from 41 to 78 years (56.93 ± 8.79 years). In detail, three patients were 40–49 years old (10.71%), 16 patients were 50–59 years old (57.14%), 6 patients were 60–69 years old (21.43%), and 3 patients were older than 70 (10.71%). And the time course from onset to diagnosis ranged from 1.5 to 14 years (5.54 ± 3.15 years), which was 5.15 ± 3.56 years for men and 6.5 ± 1.51 years for women, respectively.

3.2. Clinical features

The clinical course of 28 patients with sIBM was chronic and progressive. As for the onset symptom of total 28 patients with sIBM, 17 cases presented both lower limbs weakness (60.71%), 5 cases for both upper limbs (17.86%) and 6 cases for four limbs weakness (21.43%). More clinical details were listed in Table 1.

3.3. Laboratory examination

Serum creatase of 28 patients with sIBM were normal or mildly elevated. The mean serum CK value of 28 patients with sIBM was 397.38 ± 295.06 (79–1384) u/L. The mean serum LDH value of 28 patients with sIBM was 218.21 ± 64.32 (120–374) u/L. The mean serum ALT and AST value was 30.21 ± 18.41 (12–97) u/L and 27.67 ± 13.85 (11–56) u/L, respectively.

The sIBM patients' creatase values between different disease duration groups were listed as supplemental data. CK values of patients with sIBM for less than 5 years duration were significantly higher than that of more than 5 years duration (544.82 ± 376.34 vs. 272.62 ± 109.78) u/L ($P < 0.05$). Also, ALT values were higher in patients with <5 years duration than that in patients with ≥5 years duration (38.82 ± 21.79 vs. 22.92 ± 11.32) u/L ($P < 0.05$).

EMG reports showed that there were 21 cases with myogenic lesion, while seven cases with both myogenic lesion and neurogenic lesion.

3.4. Myopathological features

Pathological features of patients with sIBM were summarized in Table 2. Atrophic fibers presented small angularity or irregularity (82.14%, 23/28) (Fig. 1A) and small round shape (17.86%, 5/28) (Fig. 1B). Rimmed vacuoles resembled crack (Fig. 2A) (67.86%, 19/28) or round (Fig. 1C) (32.14%, 9/28).

Table 1 – Clinical symptom of 28 patients with sIBM.

Clinical manifestation	Cases	Percentage (%)
Onset symptom		
Upper limbs weakness	5	17.86
Lower limbs weakness	17	60.71
Four limbs weakness	6	21.43
Clinical symptoms		
Facial muscle weakness	2	7.14
Dysphagia	2	7.14
Neck muscle weakness	9	32.14
Proximal upper limb weakness	22	78.57
Distal upper limb weakness	23	82.14
Proximal lower limb weakness	28	100
Distal lower limb weakness	20	71.43
Quadriceps atrophy	5	17.86