

Format: Abstract

Zhonghua Er Ke Za Zhi. 2017 Feb 2;55(2):95-99. doi: 10.3760/cma.j.issn.0578-1310.2017.02.008.

[Peripheral nerve injury in LAMA2-related congenital muscular dystrophy patients].

[Article in Chinese; Abstract available in Chinese from the publisher] <u>Liang XP</u>¹, <u>Wang S, Zhang W, Yuan Y, Ding J, Chang XZ, Wei CJ, Liu JY, Xiong H.</u>

Author information

Abstract in English, Chinese

Objective: To explore the injury pattern and features of peripheral nerve in congenital muscular dystrophy patients caused by LAMA2 gene mutation. Method: Seventeen patients genetically or molecular pathologically diagnosed as LAMA2-related congenital muscular dystrophy were recruited in Peking University First Hospital between 2002 and 2015. All the patients received nerve conduction velocity (NCV) and needle electromyography tests. Clinical and laboratory examination data of the patients was retrospectively analyzed. The correlation between the NCV and disease course was determined by Pearson correlation analysis. Additionally, one patient underwent a sural nerve biopsy. **Result:** Among these 17 identified patients (13 male and 4 female), all of them were diagnosed as congenital muscular dystrophy, and all of them underwent electrophysiological examination at ages between 1 month to 6 years. Electromyogram indicated seventeen patients of myogenic damage, of whom 10 cases were complicated with reduced NCV. Twenty-six of 95 analyzed nerves showed NCV slower than the normal average of contemporary in 17%-47%. Correlation analysis between NCV and the disease course indicated that NCV of median nerves, ulnar nerves, tibial nerves and common peroneal nerves were negatively associated with the disease course (r=-0.737, -0.771, -0.540 and -0.682, respectively; all P<0.05). Sural nerve biopsy revealed peripheral neuropathy changes of myelin. Conclusion: There is peripheral nerve injury in LAMA2-related muscular dystrophy patients. It mainly manifests as demyelinating lesions. Moreover, the NCV of peripheral nerve will decrease with the increase of the course of the disease.

KEYWORDS: Muscular dystrophies; Myelin sheath; Neural conduction; Neuromuscular diseases; Peripheral nerves

PMID: 28173645

[PubMed - in process]

Publication Types

PubMed Commons

PubMed Commons home

0 comments

How to join PubMed Commons