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ORIGINAL ARTICLE

Clinical and electrophysiological study of peripheral and central neuromuscular changes in connective tissue diseases in children

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KEYWORDS

Juvenile connective tissue disease;
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Abstract *Introduction:* Children with juvenile connective tissue diseases (JCTDs) may have a wide variety of clinical features ranging from fever or a simple arthritis to complex multisystem autoimmune diseases.

Aim of the work: To study clinical and electrophysiological peripheral and central neuromuscular changes in children with connective tissue diseases.

Patients and methods: Thirty children with different JCTDs were enrolled. Clinical and neurological examination and laboratory investigations were done. Electrophysiological evaluation was performed and included: peripheral nerve conduction studies, late responses, somatosensory evoked potential and electromyography.

Results: Twenty patients had juvenile idiopathic arthritis (JIA) (66.7%), 8 patients had juvenile systemic lupus erythematosus (JSLE) (26.7%), one patient had juvenile systemic sclerosis (JSScl), and one patient had juvenile overlap syndrome (JSScl and polymyositis). Clinical neurologic abnormalities were present in 3 patients (ulnar neuropathy, median neuropathy and polymyositis). Electrophysiological abnormalities were detected in 18 patients (clinical in 3 and subclinical in 15 patients) and included ulnar entrapment neuropathy, median axonal neuropathy, demyelinating sensory motor polyneuropathy, deep peroneal nerve entrapment at the ankle (anterior tarsal tunnel syndrome), prolonged posterior tibial somatosensory evoked potential latency and prolonged H reflex latency not explained by peripheral neuropathy, increased H/M ratio and myopathic motor units. The most common electrophysiological abnormalities were present in patients with JSLE.

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Conclusion: Clinical neurological abnormalities are not common in JCTDs whereas subclinical neurological abnormalities are common findings. Juvenile systemic lupus erythematosus had the most common abnormalities among JCTDs. Polyneuropathy in JIA is commonly of demyelinating type. Entrapment neuropathy is less frequent than in adults.

1. Introduction

Children with juvenile connective tissue diseases (JCTDs) may have a wide variety of clinical features ranging from fever or a simple arthritis to complex multisystem autoimmune diseases [1]. Juvenile connective tissue diseases (JCTDs) are autoimmune multisystem inflammatory disorders including juvenile idiopathic arthritis (JIA), juvenile systemic lupus erythematosus (JSLE), juvenile systemic sclerosis (JSScl), juvenile dermatomyositis (JDM) and polymyositis, juvenile mixed connective tissue disease (JMCTD), juvenile Sjögren's syndrome (JSS) and vasculitis [2]. Different organs other than the musculoskeletal system may be involved including skin, kidneys, cardiopulmonary system, gastrointestinal and nervous systems [3].

Neurologic manifestations of rheumatic disorders can arise in both primary and secondary fashion [4]. That is, the antibodies or cellular immune elements responsible for the underlying disease can directly attack and injure nerves, muscle, brain, spinal cord, and sensory organs. On the other hand, innocent bystander effects of such rheumatic disease accompaniments as the hypercoagulable state, inflammation of the blood vessel wall, immune complex deposition and side effects of medications used in the treatment of rheumatic disease also take their toll on the nervous system [5]. Electrophysiological studies including nerve conduction studies (NCSs), electromyography (EMG) and somatosensory evoked potentials (SEPs) are most often used to diagnose disorders of the peripheral nervous systems (PNS) and central nervous systems (CNS) and provide valuable information about the underlying pathology [6,7].

The aim of this study was to study the clinical and electrophysiological peripheral and central neuromuscular changes in children with connective tissue diseases (CTDs) and to describe the neurologic complications of childhood rheumatic disease.

2. Patients and methods

Thirty children with different JCTDs (JIA, JSLE, JSScl, JDM and polymyositis, juvenile overlap syndrome and JSS) attending the pediatric rheumatology outpatient in Al-Shatby children university hospital were enrolled. Exclusion criteria included the presence of other neurological diseases, endocrinal, chronic infection, malignancy and heritable connective tissue diseases. Clinical and neurological examination and laboratory investigations were done. Thirty healthy children of matched age and sex were included as a control group. The study was approved by the local ethics committee of the Faculty of Medicine, Alexandria University and the patients gave an informed consent before inclusion in the study.

Electrophysiological evaluation was performed and included:

1. Peripheral NCSs: Nerve conduction studies were carried out using NIHON KOHDEN (Neuropack 2) electrophysiologic apparatus. All recordings of action potentials were carried out by surface electrodes (8 mm) in diameter and a ground electrode was placed between the stimulating and recording electrodes. Stimulation was carried out using bipolar stimulator having a production current ability of 50 mA. The filter setting was between 2 Hz and 10 kHz. The following nerves were studied unilaterally; sural nerve (sensory study), posterior tibial and deep peroneal nerves (motor study), radial nerve (sensory study), median and ulnar nerves (motor and sensory studies). Involvement of one peripheral nerve only was defined as mononeuropathy. Abnormality in 2 or more nerves was defined as a peripheral neuropathy electrophysiologically [8].
2. Late responses: *F* wave of median and ulnar nerves and the soleal H-reflex were performed unilaterally.
3. SEPs: SEPs of median and posterior tibial nerves with cortical recording were done.
4. EMG: Standard concentric needle EMG of gluteus medius and tibialis anterior was performed unilaterally.

Statistical analysis: Student *t* and Mann–Whitney tests were used to determine statistical differences between patients and controls as regards the values of NCSs and SEPs. Normal values for our laboratory were obtained from control children and abnormal values were defined as 2 standard deviations above/below the normal mean. Significant values were considered at $p < 0.05$.

3. Results

Twenty-two patients were girls (73.3%), and 8 patients (26.7%) were boys. Their age ranged from 6 to 16 years with a mean of 12.37 ± 2.92 years. There was no statistically significant difference between patients and controls as regards age ($p = 0.35$) and sex ($p = 0.27$). Twenty children had JIA, 8 had JSLE, one had JSScl and another had juvenile overlap syndrome.

Patients had involvement of various systems; 29 patients (96%) had articular manifestations, and 22 (73%) developed extra-articular extra-neurological manifestations during the course of disease (skin manifestations, subcutaneous nodules, fever, headaches, hypertension, diabetes mellitus, abdominal pain and fatigue). Only 3 patients (10%) developed neurological abnormalities; two of them had JSLE associated with lupus nephritis (one with ulnar mononeuropathy who had decreased sensation on ulnar nerve distribution of the right