ROLE OF MUSCLE BIOPSY IN DETECTION OF MUSCLE AFFECTION IN PATIENTS WITH SYSTEMIC SCLEROSIS AND SYSTEMIC LUPUS ERTHRYMATOUES

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Abstract

Muscle affection related to systemic sclerosis (Myo-SSc) and systemic erthrymatoues (Myo-SLE) is a disabling and unpredictable complication of SSc and SLE so the assessment of muscle according to the European Neuromuscular Centre (ENMC) criteria. The clinical, serological, laboratory as (serum aldolase, creatine kinase (CK), alanine transaminase (ALT), aspartate transaminase (AST) and C-reactive protein (CRP) and muscle biopsy features were compared between patients with and without histological evidence of myositis.

Keywords:muscle biopsy, systemic sclerosis, systemic lupus erthrymatoues.

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1. INTRODUCTION

Systemic sclerosis (SSc) is a systemic rheumatic disease characterized by three main pathologic mechanisms: altered immune function and inflammation, an obliterative vasculopathy and tissue fibrosis. Muscle involvement in SSc is a poor prognostic feature impacting survival (1) and has also been associated with cardiopulmonary complications and even sudden cardiac death (2). Myopathy in scleroderma can occur as a result of non-autoimmune etiologies such as malnutrition, disuse, or other neuromuscular disorders (3,4).

Skeletal muscle involvement occurs three to five times more frequently with diffused systemic sclerosis (dSSc) than with limited systemic sclerosis (lSSc ((5). The prevalence of skeletal myopathy in SSc varies from 5% to 96%(6,7) due to the lack of diagnostic criteria.

The EMG displays pathologic findings in the vast majority of SSc patients (>90%) (8) regardless of clinical muscle involvement, laboratory features or disease duration. The electromyographic features are similar to those of patients with polymyositis (8,9). Although distal electromyographic findings correlate with disease progression and severity in both SSc subsets, there is no established role of EMG as an outcome measure. There is also no association between EMG findings and CK or antibody levels (8,10).

Muscle biopsies reveal a variety of histopathology in SSc, such as fibrosis of the perimysium and epimysium, intimal proliferation of endomysial and perimysial vessels, perivascular infiltrates and muscle necrosis. A muscle biopsy is more likely to be useful to diagnose the disease and to serve as tools used to evaluate muscle weakness (10).

Systemic lupus erythematous (SLE) is a multisystem chronic inflammatory disease that can involve any organ system. The clinical manifestations have a wide spectrum (11). Skeletal muscle involvement is not uncommon in SLE and is seen in the form of proximal muscle weakness, myalgia and atrophy (12). The incidence of myositis in adult patients with SLE is low and varies from 4% to 16% (13,14) although not universally, accompanied by elevated muscle enzymes as ALT, AST and CK (15).

The following features on muscle histology in SLE: presence of atrophic, necrotic, degenerating and regenerating fibers, peri-fascicular atrophy, group atrophy/ type grouping, presence of inflammation and its localization, perivascular/ perimysial/endomysial invasion of non-necrotic

fibers, presence of vacuoles, fiber type abnormalities and vessel wall changes (16).

A spectrum of histological changes was described on biopsy/necropsy samples of muscle which included myositis, vasculitis, type 2 atrophy, vessel wall thickening, vacuolarmyopathy and neurogenic muscle injury. subclinical inflammatory involvement of muscle in SLE by needle biopsy. The lymphocytic vasculitis and myositis are potentially treatable. Hence the diagnosis is important (17).

A partially validated laboratory test to assess the activity of myopathies is to assay the muscle enzymes: creatine kinase (CK), alkaline phosphatase, transaminase enzymes including aspartate transaminase (AST), and alanine transaminase (ALT) and aldolase A, which present in muscles, other isomers (aldolase B and C) are present in brain or liver and are not found in plasma, except in liver diseases. Elevated serum or plasma levels of CK, transaminase enzymes or aldolase A were observed in patients with SSc with neither muscle weakness nor having engaged in any physical activities the days prior to the muscular enzymes assay.

On the other hand, in Myo-SSc, inflammatory cells and activated auto-reactive T cells contribute partly to muscle damage. Increased inflammatory markers, such as C-reactive protein (CRP), might also indirectly reflect inflammatory muscle injury and could be a potential predictive marker of subsequent Myo-SSc occurrence. Taken together, muscle enzymes, including aldolase A, CK, AST, ALT and CRP, could help identify patients who were going to have subsequent disabling Myo-SSc, but without proximal muscle weakness (18).

To show evidence of myositis in muscle biopsy specimen: myofibre atrophy and necrosis/regeneration; inflammation; fibrosis; micro-angiopathy; vasculitis involving small arteries; mitochondrial abnormalities and neuropathic changes. For each muscle biopsy information of four domains including inflammation, vascular, muscle, and connective tissue based on the international consensus proposed-score system for muscle biopsy (19) was recorded.

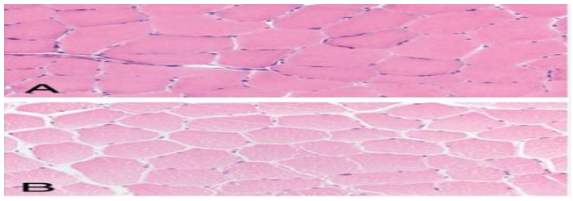

Fig. 1.- Normal muscle in transverse section (A, frozen: B, paraffin embedded). The fibers are typically polygonal, and the nuclei are located peripherally. Empty space between muscle fibers is a shrinkage artifact, which is typically more pronounced in paraffin sections (H&E)

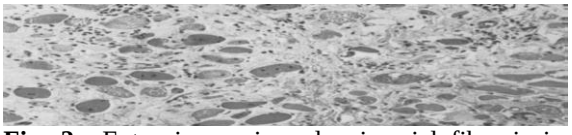

Fig. 2.- Extensive peri- and epimysial fibrosis in cross section in deltoid muscle; fibrosis mixed with scattered chronic inflammatory cells. Muscle fibers distorted and some with central nuclei. Hematoxylin and eosin, 270 x.

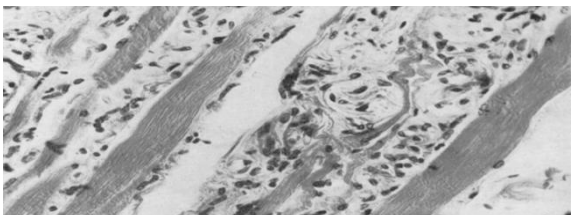

Fig.3-Fibrosis, muscle fiber atrophy and abortive regeneration in longitudinal section in quadriceps muscle. Hematoxylin and eosin, 270 x

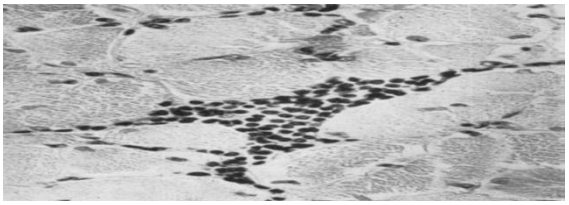

Fig. 4.- Interstitial collection of lymphocytes in quadriceps. Hematoxylin and eosin,270 x.

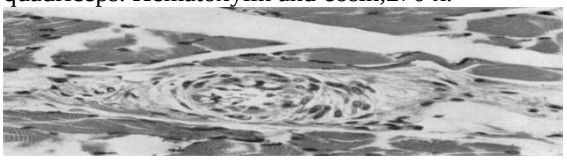

Fig. 5.- Perivascular fibrosis. Hematoxylin and eosin, 270 x.

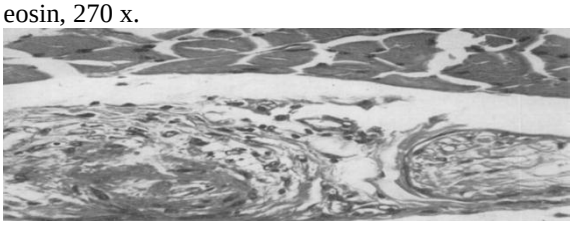

Fig. 6-Hematoxylin and eosin, 270 x.

In our study microscopic involvement of muscle fibers in SLE inflammation was present in 11(36.6%) of 30 cases, vasculitis was present in 10(33.4%) of 30 cases, atrophy was present in 16(63.3%%) of 30 cases and degenerative fiber was present in 15(50%) of 30 case so the most consistent abnormality in SLE was muscle atrophy. Microscopic involvement of muscle fibers in Systemic sclerosis inflammation was present in 14(46.7%) of 30 cases, vasculitis was present in 11(36.6%) of 30 cases, atrophy was present in 15(50%) of 30 cases and degenerative fiber was present in 15(50%) of 30 case so the most consistent abnormality in SSC was muscle atrophy and degenerative fiber.

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