Muscle pathological features in a patient with a hypokalemic periodic paralysis/dermatomyositis "double trouble"

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Objectives

To describe myopathological findings in a patient affected with dermatomyositis and hypokalemic periodic paralysis.

Patient and methods

A 66-year-old woman presented with hand and face erythematous rashes, progressive proximal limb weakness and dysphagia with a subacute onset. Serum CK levels were elevated 3-4 times the normal values and EMG analysis showed a diffuse chronic myogenic pattern. About six months after symptom onset a cholangiocarcinoma was diagnosed.

In the past, she received a clinical diagnosis of hypokalemic periodic paralysis.

Deltoid muscle biopsy was performed.

Results

Deltoid muscle biopsy showed variability of fiber size, many fibers with internal nuclei, some fiber splitting, rare cell necrosis with phagocytosis, some small perivascular and perimysial inflammatory infiltrates, multifocal perifascicular atrophy and few COX-negative fibers.

Several fibers presented pseudo-vacuolations that appeared optically empty or containing some amorphous material on routine histological staining. They usually had curving or undulating borders and a clearly defined limiting membrane. Some of them were connected with the extracellular space as an intra-cytoplasmic sarcoplasmic invagination. Diffuse MHC I sarcolemmal positivity was observed also involving the pseudo-vacuolation limiting membranes.

Discussion

Muscle biopsy confirms a singular «double trouble» in this patient. On one hand, such pathological pattern is suggestive for proliferation, regeneration and dilation of components of the T tubular system and the sarcoplasmic reticulum as often observed in dyskalemic periodic paralysis; on the other hand, presence of perifascicular atrophy, cell necrosis and inflammatory cells support the clinical diagnosis of dermatomyositis.