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Concentric macro EMG role in electrodiagnostic evaluation of inflammatory myopathies

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Abstract

Inflammatory Myopathies (IM) encompass a diverse group of systemic autoimmune muscle disorders, not easy to diagnose. Concentric-macro EMG is supposed to reflect electrical activity of the entire motor unit. It could help in electrophysiological characterization of inflammatory myopathies and recognizing earlier myopathic changes. Conventional EMG in our IM patients showed myopathic changes in all the cases, so it could be a golden tool for diagnosis. The degree of abnormalities was variable in different muscles in concentric macro EMG. Myopathic changes using concentric macro EMG was not so advanced as those seen in chronic stable relapsing phase. Pooling up the results of the conventional EMG, muscle enzyme tests, muscle ultrasound , and ConMac will give optimal results for diagnosis and could help in follow up of patients with IM.
Key Words: inflammatory myopathies; conventional EMG; concentric macro EMG.

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Inflammatory Myopathies (IM) encompass a mixed group of systemic autoimmune inflammatory disorders. They are characterized with rather symmetrical proximal muscle weakness, muscle pains, elevated serum levels of enzymes derived from skeletal muscle, electrophysiological changes consistent with inflammation, and muscle histopathology of non-suppurative inflammation, sometimes extra muscular manifestations involving the skin, heart, joint and lung may occur.[1] The most recognized subtypes are polymyositis, and dermatomyositis. They may be associated with other conditions such as connective tissue diseases and malignancy.[2] The early identification of these patients is important as they may often have a poor prognosis and quality of life.[3]

Although clinical history and physical examination remain important aspects of disease assessment, other methods are often required to improve diagnostic certainty. EMG plays an important role along with clinical assessment, laboratory tests, serum antibody testing, and muscle biopsies. In addition, EMG may have additional value in the diagnosis as it is not invasive such as muscle biopsy, and it can detect the affection in more than one muscle. The EMG characteristics of IM include increased insertional activity, fibrillations, and early recruited, short spiky polyphasic Motor Unit Action Potentials (MUAPS). In more chronic IM, complex MUAPs of increased duration may be seen.[4]

Macro EMG is a well-accepted technique for estimating the electrical size of a motor unit inside the muscle. It makes use of a large recording surface that makes it possible to sample the majority of muscle fibers of a motor unit, in contrast to the 10–20 muscle fibers sampled by the tip of a concentric needle electrode.[5,6]

Materials and Methods

This case-control analytical study was performed during the period from August 2021 to May 2023 in Cairo University Hospitals.

The study was carried out on a total of 46 subjects, with an age range from 18 to 65 years. 26 adult patients fulfilled the criteria for the diagnosis of inflammatory myopathy by clinical examination and laboratory investigation. Those patients were compared to 20 healthy controls matching in age and sex. The sample size was calculated to be 20 patients, as the minimum expected sensitivity and specificity are expected to be 95%. Using a power 80% and 5% significance level.

Patients with other identifiable neurological disorders, such as spinal diseases, neuropathies, different types of myopathies other than inflammatory ones, such as hereditary myopathies, chronic medical conditions causing myopathy, like endocrine disorders, and patients on medications causing muscle problems, like steroids or statins, were excluded.

Written informed consent was obtained from each patient. The research ethics committee of the Faculty of Medicine-Cairo University (MD-211-2021) approved on 10-8-2021. The study protocol followed the ethical guidelines of the 1975 Declaration of Helsinki and its later amendments (as

revised in Seoul, Korea, October 2008) with good clinical practice guidelines as reflected in a prior approval by the institution's human research committee. With respect to the patients' confidentiality, patients were represented in the study by code numbers and all personal data were hidden.
All selected patient were clinically assessed through complete history taking and full neurological examination with special stress on assessment of muscle power using MRC grading" muscle strength grading system by the Medical Research Council" which extends from grade one as no contractions to grade six as normal muscle power.[7,8] The distribution of the weakness, duration of the disease and medications such as steroids and immunosuppressant were also investigated. General examination, including other system involvement or characteristic rash in face, neck, and sun-exposed areas, arthralgia, Raynaud's phenomenon, and fish mouth presence was fulfilled. The laboratory tests were performed routinely for all cases included muscle enzymes levels (CPK, ALT and AST), inflammatory markers (ESR and CRP), virology screening while myositis specific antibodies were done only for selected number of cases due to limited availability and according to the positive antibody they were investigated, but none of the cases discovered malignancy at the time of examination. Muscle ultrasound was also done for all cases, and none showed the characteristic ground glass appearance of hereditary myopathy.
According to clinical assessment and history taking, the patients were classified into group I which had a rapidly progressive course, and group II which had a slowly progressive course, and according to the duration of the disease, the cases were subdivided into acute cases (new cases <1 year) and chronic cases (>1 year).
Needle EMG examination was performed using a Nicolet EDX system and Synergy software program (Natus Medical, Inc., San Carlos, California, USA) with a concentric needle electrode with standard filter settings of 10Hz -10 KHz. Several manipulations were needed to get the best morphology of MUP and ensure the persistence of the abnormality. Conventional EMG was evaluated first by assessment of spontaneous activities (Fibrillation potentials, positive sharp waves "PSWs" and chronic repetitive discharges "CRDs") during rest as well as the analysis of a run of motor unit potential configuration with regards to duration, amplitude and number of phases, analysis of firing rate relative to the action for interference pattern and recruitment judgment as the patient slowly increases the force of contraction to maximal effort. One proximal and one distal muscle were examined in each limb, as well as paraspinal muscles.
Concentric Macro EMG was carried out using a 2-channel EMG Viking machine, where channel 1; a concentric MUAP is recorded from the electrode's active recording tip, with the cannula used as reference. This concentric MUAP is used to trigger a time-locked potential on channel 2, where the needle's cannula is now the active recording area and a surface electrode nearby is used as a reference, as shown in Figure 1. Con-Mac was performed for the right Tibialis Anterior (TA), Biceps Brachii (BB) and Deltoid (DD) during slight voluntary activation of the muscle with strict instructions to the patient. Con-Mac potential amplitude, area and duration were calculated. Con-Mac potential gives global information not regional about MUP and motor unit size estimation. The values were compared to controls matching with age and sex during the same slight force of contraction on the same machine with the same settings.
Statistical analysis was carried out using the Statistical Package of Social Science Software program (SPSS) version 27. Data was presented using mean and Standard Deviation (SD) for quantitative variables, frequency, and percentages for qualitative variables. Comparisons between two groups were conducted using the "Student t-test" for quantitative variables, and the "Chi" test for qualitative variables. Comparisons between more than two groups were done using the ANOVA test for quantitative variables. Paired comparisons of frequencies (nominal data) were carried out using McNemar's Chi-Square test. Comparisons of sensitivity and specificity between different variables were presented using Area Under Curve (AUC), calculated from ROC curve. Correlations (association between quantitative variables) were carried out using Pearson's test.

Results

This case-control study included 26 inflammatory myopathic patients and 20 controls. Of these patients, 38% had polymyositis, 23% had dermatomyositis, 23 % had overlap syndrome, 16% rare subtypes such as IBM (3.84%), viral myositis (3.84%), IMNM (3.84%), PM associated with malignancy (3.84%), according to ACR/EULAR criteria.[9]
The age of the patients and controls ranged from 18 to 65 years with a mean of 36 years. There were 18 females (70%) and 8 males (30%) patients and 14 females (70%) and 6 males (30%) controls. The patients and controls were matched in age and sex. The duration of the disease ranged from 1 to 36 months with a mean duration of 13.5±11.4 months. The study was done for acute and chronic cases. Acute cases constituted 57.7% of the patients, while chronic cases represented 42.3% of the patients.
The clinical parameters were analyzed as follows. The grade of weakness was classified according to MRC grading of muscle power.[7] We graded the hip and shoulder joints to evaluate the grade of the proximal muscle weakness affecting the upper and lower limbs which generally occurs in myopathy. Muscle power of the proximal upper and lower limbs graded from 1-3 on MRC scale was interpreted as severe weakness while grades 4, 5 were considered as mild weakness. *Supplementary Table 1* shows that the study cohort had different grades of weakness independently on the duration of the disease. The proportion of severe weakness was predominantly in acute cases while the distribution of mild weakness was approximately the same among acute and chronic cases.
There was also no significant difference in CPK level between acute and chronic cases as presented in *Supplementary Table 2* shows that shows that the highest percentage of the patients had CPK level ranged from 1000 to 5000.
The highest percentage of fibrillation potentials and positive sharp waves were seen in acute cases, whereas the highest percentage of CRDs were noted in chronic cases, while only less than two-thirds of patients showed spontaneous activities. The absence of spontaneous activities in the acute phase was found in patients who received IVIG, as shown in *Supplementary Table 3*.

There was a statistically significant difference between cases and controls regarding con-Mac variables (area, amplitude, and duration) in tibialis anterior and biceps brachii, while the deltoid muscle showed a significant difference only in amplitude and area, as presented in *Supplementary Table 4*.

ROC curve results used to detect the cut-off point, sensitivity, and specificity of Con-mac in the diagnosis of inflammatory myopathy are displayed in *Supplementary Table 5*.

The most sensitive parameter of Con-Mac in the diagnosis of inflammatory myopathy was Tibialis Anterior (TA) muscle amplitude (μv) which had 88.46% sensitivity, followed by the area and amplitude of biceps brachii (BB) muscle (μv) and the duration (msec) of TA muscle with 84.62% sensitivity whereas the area (mm^2) of the deltoid muscle was the most specific parameter for the diagnosis with 100% specificity.

Supplementary Tables 6 and 7 show a significant positive correlation between the presence of spontaneous activities and CPK level, and there was a significant negative correlation between the presence of spontaneous activities and the power of the upper limb, which means that as the distribution of spontaneous activities increases, the grade of weakness increases. In addition, it was noted that CPK did not correlate well with the power examination of the patient.

The patients, who had spontaneous activities at the time of examination, have decreased MUP parameters of Con-Mac (amplitude, area, and duration) in all examined muscles with statistically significant differences from the patients who had no spontaneous activities, as shown in *Supplementary Table 8*.

Also, there was a highly significant negative correlation between the Con-Mac amplitudes of the tibialis anterior, biceps brachii & deltoid muscles and CPK level, as presented in *Supplementary Table 9*.

The Con-Mac area of the biceps brachii muscle showed a statistically significant decrease in patients with a rapidly progressive course compared to those with a progressive course, as presented in *Supplementary Table 10*.

In addition, there was a statistically significant decrease in the area and amplitude of Con-Mac of the biceps brachii muscle in acute cases in comparison with chronic ones, as shown in *Supplementary Table 11*.

Discussion

The current study confirmed the high diagnostic sensitivity of Con-Mac EMG for diagnosing inflammatory myopathy. A significant decrease in MUP parameters (amplitude, area, and duration) was observed in all examined muscles, while the amplitude of tibialis anterior muscle showed superior significant sensitivity.

Jabre (2017) reported that Con-Mac could be more helpful than conventional EMG in mild and early myopathic cases. Clearly abnormal or nearly flat pattern of Con-Mac was observed in polymyositis patients.[5] Another study was done by Fuglsang-Frederiksen (2006) showed that Macro EMG has the same sensitivity of concentric needle EMG in the diagnosis myopathy in general.[8]

On the Contrary, Barkhaus *et al.* (1990) studied macro-EMG of biceps brachii muscle in 10 patients with inflammatory myopathy. They found a decrease in macro-EMG amplitude in only 2 patients. This discrepancy between our and Burkhaus's results may be due to the number of the patients included as well as the muscles being studied (the small sample size).[4]

The current study showed a strong negative correlation between CPK level and Con-Mac amplitude, and also a strong positive correlation was found between the presence of CPK level and spontaneous activities as both reflect the inflammatory process "degree of damage in the muscle". No correlation was found between CPK level and the clinical data of the patients (MRC power grading).

Our results showed a significant decrease in Con-Mac MUP parameters in patients who had spontaneous activities in conventional EMG, while the cases with no spontaneous activities and had mild weakness (grades 4, 5 by MRC power grading) at the time of examination, their Con-Mac data parameters of all examined muscles (area, duration, and amplitude) were towards the normal range. This could be a sign of regeneration.

Conclusions

We concluded that Con- Mac EMG examination could be helpful as an objective dynamic sensitive test for follow up the degree of damage and improvement of those patients. This needs a long prospective follow-up study to be achieved.

List of abbreviations

Con-Mac EMG, Concentric macro electromyography.
IM, inflammatory myopathies.
MRC, Medical Research Counsil.
CPK, Creatin phosphokinase
ALT, Alanine transaminase
AST, Aspartate transaminase

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Availability of data and materials

The data that support the findings of this study are available on request from the corresponding author.

Conflict of interest

The authors declare that they have no conflict of interest.

Ethics approval and consent to participate

This study had the approval of the Ethics Committee for research of faculty of Medicine-Cairo University on 10-8-2021 with a code (MD-211-2021). A written consent was taken from each subject.

Contributions

EA achieved the methodology, shared in writing the manuscript, AF plotted the hypothesis, was the research manager, helped in general revision of the manuscript, LA revised the practical and theoretical inputs at each step of the research, helped in discussion generation, SA was responsible for proper selection of the patients, and progress of the clinical part of the research, EE organized the results segments, EAM and MS helped in organization of the results, writing and editing the manuscript.

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Disclaimer

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Online supplementary material:

Table 1. Summarization of the grade of weakness among the cases.

Table 2. CPK level grading among the subjects.

Table 3. Proportion of patients with spontaneous activities recorded by EMG.

Table 4. Con-Mac EMG Data of MUP analysis of 3 muscles (Tibialis Anterior (TA), Biceps brachii (BB), and Deltoid (DEL) muscles for the 2 study groups.

Table 5. Roc curve results to detect cut off point, sensitivity, and specificity of Con-mac in diagnosis of inflammatory myopathy.

Table 6. Correlation between the presence of spontaneous activities, CPK level, and power in upper and lower limbs as measured by MRC grading and age.

Table 7. Correlation between CPK level, and power in upper and lower limbs as measured by MRC grading:

Table 8. Comparison of Con-Mac data between the patients who had spontaneous activities during rest and who had not.

Table 9. Correlation between Con-Mac EMG variables, and CPK level.

Table 10. Disease progression and Con-Mac data.

Table 11. Comparison between acute and chronic cases regarding Con-Mac Data.