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MUSCLE BIOPSY CORRELATED WITH ELECTROMYOGRAPHY

STUDY OF 100 CASES

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SUMMARY — To find what the correlation is and verify if is possible to avoid extensive electromyographic examination, studying only *one muscle*, 100 patients with neuromuscular disorders (58 primary myopathies, 32 neurogenic disorders and 10 myotonic dystrophies) were submited to quantified electromyography (EMG) and muscle biopsy (MB) with fresh-frozen section plus histochemistry in the same muscle, but on the opposite side. The EMG was abnormal in 98% and MB in 93% of the cases. EMG and MB had a concordance of 84.3% in the neurogenic disorders and 84.77% in the primary myopathies. A correlation of 80% was obtained between all MB and EMG (including the cases of myotonic dystrophies), regarding the origin of the pathogenic process ($p<0.01$). The EMG had 5% inconsistences and the MB 11%, with respect to the pathogenic process. When the myotonic dystrophy was separated from the primary myopathies and from the denervation disorders, a complete concordance was found in all MB and had only 3.4% inconsistences in the denervation disorders and 3.1% in the primary myopathies.

Correlação entre biópsia muscular e eletromiografia: estudo de 100 casos.

RESUMO — A fim de verificar qual a correlação entre uma biópsia muscular e a eletromiografia quando um único músculo é estudado, bem como verificar a possibilidade de evitar eletromiografias extensivas e dolorosas em determinadas doenças, foram investigados 100 pacientes com doenças neuromusculares (58 com miopatias primárias, 32 doenças que determinam desinervação e 10 distrofias miotônicas). Todos os casos foram submetidos a eletromiografia quantificada (EMG) e a biópsia muscular (BM), utilizando técnicas de colorações a fresco e pela histoquímica, realizadas no mesmo músculo, mas em lados opostos do paciente. A EMG foi anormal em 98% e a BM in 93% dos casos. A EMG e a BM tiveram concordância de 84,3% nas doenças neurogênicas e 84,77% nas miopatias primárias. Foi obtida correlação de 80% entre a EMG e a BM em todos os casos, com relação à patogenia do processo ($p<0,01$). A EMG teve 5% de inconsistências e a BM 11%, em relação ao diagnóstico patogênico. Quando a distrofia miotônica foi separada das miopatias primárias e dos processos que determinam desinervação, uma concordância completa foi encontrada entre todas as BM e a EMG mostrou inconsistências somente em 3,4% das doenças que determinam desinervação e 3,1% das miopatias primárias, sugerindo que a distrofia miotônica deva ser classificada em um grupo separado (neuromiopatias). Os autores discutem os dados encontrados em relação à literatura existente e concluem que, dependendo da avaliação clínica, é possível submeter os pacientes a somente um dos procedimentos (geralmente a BM), utilizando ambos os procedimentos somente nos casos em que não foi possível chegar a um diagnóstico, sendo então examinados diversos músculos na EMG.

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Since the demonstration of the value of electromyography (EMG) to differentiate the primary muscular disorders from the diseases of neurogenic origin by Buchthal and Clemmensen [12], several authors [10,11,32,34,37] helped to consolidate the EMG showing the value in the diagnosis of myopathies and disorders resulting from lesions in the lower motor neurons and nerves without, however, allowing a nosological diagnosis [11,34]. The first open muscle biopsy (MB) carried out by Griesinger and Billroth [33], induced Duchenne [22,23] to study the muscle of patients to firm an exact diagnosis and prognosis of muscular diseases. Erb [29] defined the essential alterations in dystrophic muscles, that were extended and used untill now [1,18,21]. With the introduction of histochemistry in the analysis of MB, the histological diagnosis of neuromuscular disorders was expanded and improved [7,8,21,28,58]. Several studies in the past were done, trying to confront the results of MB with EMG [4,9,13,35,37,43,53,56], sometimes reaching conflicting results, favouring at times the EMG [13,35,53] or in other occasions the MB [43], due to different techniques of investigation (diseases with simultaneous involvement of muscles and nerves [37]; lack of specific date about MB [37]; lack of report in which cases the concordance happened between both procedures [9,37]; absence of histochemical techniques [9,35,37,53,56]; heterogenous and rare group of diseases in the analysis [9,13,37]; study of several muscles by the EMG and only one muscle in the MB [4,13,35,37,53,56]; several EMG in different times and only one MB [13,35,53,56]; small number of patients [9]; MB in the same place of previous EMG [9,53]; study only of myopathies [43] or only denervation disorders [56]).

To correlate the MB processed by fresh-frozen section stains and histochemistry with EMG, in relation to the pathogenic diagnosis and verify it is possible to avoid extensive and painful EMG studies when only one muscle was studied in both procedures, this present study was undertaken using specific pre-stablished criteria to avoid the above difficulties.

MATERIAL AND METHODS

We selected patients with the diagnosis of Duchenne muscular dystrophy (DMD), limb-girdle muscular dystrophy (LGMD), facio-scapulo-humeral dystrophy (FSHD), myotonic dystrophy (MD), dermato-polymiositis (DP) without neoplasia or associated to connective tissue disorders, amyotrophic lateral sclerosis (ALS), infantile spinal muscular atrophy (ISMA) and peripheral neuropathy (PN) who had typical clinical and laboratory findings, as well as electromyographical and histological-histochemical data available to meet the criteria described below. The confrontation of 600 MB with 1500 EMG were carried out from 1975 to 1983, looking for cases which had both procedures, and which met the following criteria: 1) symmetrical pathology, with clinical involvement on both sides of the body; 2) EMG and MB should have been on an identical muscle but on the opposite side; 3) cases where an EMG had been done on both sides previously, would not be included, to avoid the «needle myopathy» (*5,6,16,24,47,52*); 4) EMG and MB should have been done at the same period (within 3 weeks), based on studies which evaluate the regeneration and alterations in volunteers after an EMG (*62*); 5) the EMG should have been quantitative, having registered the insertion activity, spontaneous activity during muscular relaxation, average duration of potentials, register of long and short polyphasic potentials, recruitment of efforts and pattern of severe efforts, according to criteria which will be defined; 6) the MB should have been adequate, in quality and number of muscle fibers, containing information on the proliferation of connective tissue, adipose tissue infiltration, necrose, phagocytosis, diffuse inflammatory infiltrate, perivascular inflammatory infiltration, internal nucleus, splitting fibers, whorls, moth-eaten, snake coils, ring fibers positive fibers and increased of the acid phosphatase in the interstice, positive fibers and increase of the alkaline phosphatase in the interstice, type I and II fiber atrophy, type I and II fiber hypertrophy, predominance of type I and II fibers, type I and II fiber deficiency, dark angular atrophic fibers in the NADH-tetrazolium reductase, dark angular fibers in the non specific esterase, targets, grouping of type fibers, fascicles atrophy and perifascicular atrophy, such as the criteria defined in other studies (*1,7,8,13,21,28,58*); 7) the muscle studied should be degree 4 (MRCM) (*42*), to avoid the selection of very severe affected cases.

Electromyography — The EMG were done with a MEDIC equipment, with frequency response between 15Hz and 10KHz, using normal concentrical needle electrodes and the ground electrode was put at the root of the examined limb. Room temperature was between 20-30ºC, with patients extremities kept warm. The exam procedure was normal, according to techniques described by several authors (*10,30,32,38,39*). During the procedure, each one of

the parameters below was evaluated, according to the criteria described. 1) Insertion activity: abnormal if the duration was above 300ms (*32,38*). 2) Fibrillation: duration from 1-5ms, amplitude varying from 20-300 uV, usually biphasics, detected outside the area of the motor end-plate, with the first component being positive (*14,20,32,39*). 3) Fasciculation: registered during muscular relaxation, without a fixed frequency, amplitude, duration or morphology (*32,39*). 4) Positive waves: quick positive potential, followed by a long negative phase, with a voltage varying between 50uV to 1mV, with a duration of 20-200ms (*14,32,39*). 5) High frequency discharge: identical frequency for each muscular fiber group, synchronous, with an amplitude of 50uV to 1mV and an average duration of 150ms (*38*). 6) Myotonia: continuous and rythmical discharges, which increase and diminish the voltage, for long periods after having stopped the initial stimulus, being able to assume the shape of positive waves or fibrilations (*38*). 7) Average duration of action potentials: approximately 20 different potentials were analysed, obtained during slight contraction and a mean voltage was obtained; the result was compared to the values proposed by Buchthal (*10*), being considered increased if the deviation was over 20% and diminished if it was under 20%. 8) Average voltage of voluntary potentials: identical analysis to the previous potentials was made, but the only potentials considered were those which the positive deflexion peak to highest negative was not above 200us; there are no agreement in the literature about the normal values — 300uV to 5uV (*32*), 100uV to 2mV (*39*), higher than 2mV and lower than 4mV (*13*), 100 to 2000uV (*53*), important when it exceeds 6000uV(*55*), some hundreds of uV to under 3mV (*56*) — we use the voltages found in normal individuals from our laboratory, which vary from 500 to 3000uV (unpublished data); they were considered increased if above 3000uV and diminished if under 500uV. 9) Polyphasic potentials: voluntary potentials which present more than 4 phases and are considered abnormal if exceed 12% of potentials examined (*10,15,32*), but we consider abnormal in our laboratory, only if they exceed 25%; if the total duration was the same or lower than the average duration for the age, they were called short: if the duration was higher than the average for the age, they were called long (*32,37*). 10) Effort recruitment: when the individualization of several motor units in the interference pattern with reduced duration during weak contraction was possible, it was considered increased; if the total number of motor units was diminished, allowing verification of areas without potentials, it was considered reduced (*39*). 11) Effort pattern: during a weak, medium and strong contraction, the effort pattern was analysed, being classified as BSAP if the potentials were of reduced duration, low voltage and plentiful on effort (*19,25*), PMUP if the motor unit presented polyphasic potentials, which were individualized on the interference pattern, SMUP if the potentials presented reduced duration and normal voltage, GMUP if there were potentials of high voltage, usually over 5mV, MUP if they presented individualized motor units of normal duration and voltage and MIXED if by temporo-spacial somation of the discharges it was impossible to distinguish any individualized potentials, as it occurs in the normal muscle. 12) Electromyographic diagnosis: depended on the addition or absence of some kinds of potentials found, being divided into specific and non specific, for cases of primary muscular involvement (myopathies) or for denervation (secondary muscle atrophy or neurogenic) (*38*); were necessary to make a diagnosis, to have 3 types of specific criteria in the same category; the non specific criteria were not considered, unless they appears with high intensity and frequency; when an exam had at same time potentials belonging to myopathies or denervation categories, it was placed in the category of mixed involvement (Table 1).

Muscle biopsy — After choosing the muscle which will be examined, usually degree 4 (MRCM) (*42*) and which also had been tested on EMG but on the opposite side, a biopsy with stains for haematoxilin-eosin, modified Gomori trichrome by Engel & Cunningham, PAS, oil red O and cresyl violet was done. The histochemical reactions were also processed for the alkaline ATPase pH 9.4, acid ATPases pH 4.3 and 4.6, NADH-tetrazolium reductase, non specific esterase, succinic dehydrogenase, myophosphorilase, acid and alkaline phosphatase, according to techniques used in our laboratory (*58*). To systematize the diagnosis, the main histological alterations were grouped according to previous reports in the literature (*21,58*) and the examiner (LCW) did not know in this time the clinical diagnosis. These histological alterations were then catalogued under the following items. 1) Myopathies: if the MB had proliferation of the connective tissue, adipose tissue infiltration, internal nucleus, necrose, phagocytosis, excessive diffuse or perivascular inflammatory infiltration, basophilic fibers, increased acid phosphatase in the fibers and in the interstice, increase of alkaline phosphatase in fibers and interstice, fiber splitting, whorls, moth-eaten, snake coils, ring fibers, predominance of type I fiber atrophy and predominance of type II fibers hypertrophy. 2) Denervation: if the MB had very rare fibers with necrose and phagocytosis,

Specific criteria	Non specific criteria
Primary muscle involvement (Myopathies)	
Diminished duration of potentials	Increased insertion activity
Reduced average voltage	Fibrillation
Increased recruitment	Positive waves
Excess of short polyphasic potentials	High frequency discharges
Myotonia	BSAP or SMUP pattern
Secondary muscle atrophy (Denervation or neurogenic)	
Increased duration of potentials	Increased insertion activity
Increased average potentials voltage	Fibrillation
Reduced recruitment	Positive waves
Fasciculation	High frequency discharges
Excess of long polyphasic potentials	MUP or PMUP pattern
GMUP pattern	
Mixed involvement (Myopathic and denervation components)	
Combinations of the specific criteria of the first and secnd group.	

Table 1 — Electromyographic diagnosis criteria.

rare internal nucleus, frequent dark atrophic angular fibers in the non specific esterase and NADH-tetrazolium reductase, targets, fiber type groupings, presence of type I and II fiber hypertrophy and predominance of type II fiber atrophy, atrophy of large groups of fibers or involving the whole fascicle. 3) Mixed: when the MB had elements found in myopathies and denervation as described above. 4) Acessories diagnosis: when only type I or II fiber atrophy as the only abnormality found.

RESULTS

Fullfilling the adopted criteria we study 100 cases, according to table 2. There were 98 EMG and 93 MB abnormal in the 100 cases (Table 3).

Number of cases	Neuromuscular disorders	Age (years) mean	Sex		Muscle studied			
			M	F	Quadr	Bic	Gastr	Delt
25	Pseudo-hypertrophic muscular dystrophy	9.1	25	0	24	1		
14	Limb-girdle muscular dystrophy	21.5	8	6	10	3	1	
9	Facio-scapulo-humeral dystrophy	28.6	5	4	3	6		
10	Myotonic dystrophy	34.2	8	2	3	7		
10	Dermato and polymyositis	28.2	6	4	5	4		1
10	Amyotrophic lateral sclerosis	49.7	8	2	9	1		
9	Infantile spinal muscular atrophy	4.6	4	5	9			
13	Peripheral polyneuritis *	34.9	12	1	11		1	

*Table 2 — Muscle studied, sex and age in 100 cases of neuromuscular disorders. * Etiology of peripheral polyneuritis: chronic insecticide intoxication, 9 cases; Dejerine-Sottas disease, 2 cases; intermittent acute porphyria 1 case; chronic recurrent polyneuritis, 1 case. Sex: M, male; F, female. Muscle: Quadr, quadriceps; Bic, biceps; Gastr, gastrocnemius; Delt, deltoid.*

	PHMD	LGMD	FSHD	D&P	MD	ALS	ISMA	PP
Number of cases →	100	25	14	9	10	10	10	9	13
Electromyographies:								
Myopathic	52	25	13	6	4	3		1	
Denervation	38			2		7	10	8	11
Mixed	8				6				2
Normal	2		1	1					
Muscle biopsies:								
Myopathic	56	25	14	7	8	2			
Denervation	32					6	10	9	7
Mixed	3			1	1	1			
Type II fiber atrophy	2								2
Normal	7			1	1	1			4
Accordance between EMG and MB	80	25	13	5	5	5	10	8	9
	80%	100%	98.85%	55.55%	50%	50%	100%	88.88%	69.23%
χ^2	96.41 ($P < 0.01$)								

Table 3 — Results of electromyographies and muscle biopsies grouped according to the pathology: PHMD, pseudo-hypertrophic muscular dystrophy; LGMD, limb-girdle muscular dystrophy; FSHD, facio-scapulo-humeral dystrophy; D&P, dermato and polymyositis; MD, myotonic dystrophy; ALS, amyotrophic lateral sclerosis; ISMA, infantile spinal muscle atrophy; PP, peripheral polyneuritis.

Incompatible EMG with definitive diagnosis — In the group of 58 myopathies (DMD, LGMD, FSHD and DP) incompatible results were found in the EMG of 2 FSHD (EMG of denervation) and 2 other cases with normal EMG, one being of LGMD and another of FSHD, with a total of 4 cases (6.89%). In the group of 38 denervation cases (ALS, ISMA and PN), one case of ISMA had EMG suggestive of myopathy (3,12%). The myotonic dystrophies were separated in this classification, as it will be explained later. In our overall results, there were 5 disagreeing cases of EMG with the pathogenic diagnosis (Table 4).

Number of cases	Type of pathology	Electromyographic diagnosis				
		Myop.	Denerv.	Mix.	Norm.	Incomp.
	Myopathies:					
25	Pseudo-hypertrophic musc. dystr.	25				
14	Limb-girdle musc. dystrophy	13			1	1
9	Facio-scapulo-humeral dystr.	6	2		1	3
10	Dermato and polymyositis	4		6		
58		48	2	6	2	4 (6.89%)
	Denervation:					
10	Amyotrophic lateral sclerosis		10			
9	Infantile spinal muscle atrophy	1	8			1
13	Peripheral polyneuritis		11	2		
32		1	29	2		1 (3.12%)
10	Myotonic dystrophy	3	7			
100	Total	52	38	8	2	5 (5.0%)

Table 4 — Results of electromyographies. Electromyographic diagnosis: Myop, myopathic; Denerv, denervation; Mix, mixed; Norm, normal; Incomp, incompatible.

Incompatible MB with definitive diagnosis — In the group of 58 primary myopathies, two had normal results, a FSHD case and another of DP, and none incompatible with the pathogenic proposition (3.44%). In the group of 38 denervation cases, four had a normal MB (PN cases). In the 10 MD cases, one was normal. In our overall results, seven cases were normal (7%), with no cases of incompatibility between the basic pathology and MB. Some pathologies can have mixed alterations, and in this way, cannot be catalogued then as incompatible, either in the EMG or MB results. The MD presented most cases with typical alteration for primary muscle involvement and denervation, which will permit a separate analysis (Table 5).

Concordance between EMG and MB — There were an agreement of 80% between the MB and EMG. The agreement between the two methods was 82.75% in the primary myopathies (100% in DMD, 92.85% in LGMD, 55.55% in FSHD and 50% in DP) and 84.37% in the denervation disorders (100% in ALS, 88.88% in ISMA and 69.28% in PN). The myotonic dystrophy presented only 50% of accordance between the two methods. The chi-square test gave a value of 96.41 ($p<0.01$) (Table 3).

Number of cases	Type of pathology	Anatomo-pathological diagnosis					
		Myop.	Denerv.	Mix.	Type II Fib. Atr.	Norm.	Incomp.
	Myopathies:						
25	Pseudo-hypertrophic musc. dystr.	25					
14	Limb-girdle musc. dystrophy	14					
9	Facio-scapulo-humeral dystrophy	7		1		1	1
10	Dermato and polymyositis	8		1		1	1
58		54		2		2	2 (3.4%)
	Denervation:						
10	Amyothophic lateral sclerosis		10				
9	Infantile spinal muscle atrophy		9				
13	Peripheral polyneuritis		7		2	4	4
32		26			2	4	4 (12.4%)
10	Myotonic dystrophy	2	6	1		1	1 (10.0%)
100	Total	56	32	3	2	7	7 (7.0%)

Table 5 — Results of muscle biopsies. Anatomo-pathological diagnosis: Myop, myopathic; Denerv, denervation; Mix, mixed; Type II Fib Atr, type II fiber atrophy; Norm, normal; Incomp, incompatible.

Incompatibility between EMG and MB — The cases were grouped under diseases of primary myopathic origin, disorders of neurogenic origin and myotonic dystrophy, having in mind the controversy regarding the pathogenesis of myotonic dystrophy. Witn this division there was no incompatibility in the MB regarding the pathogenesis (no denervation diagnosis in primary myopathies and no myopathic diagnosis in denervation). In the EMG there was 3.4% incompatibility in primary myopathies (2 cases of primary myopathy with diagnosis of denervation) and 3.1% incompatibility in denervation disorders (1 case of denervation with diagnosis of primary myopathy (Table 6).

	Primary myopathies		Denervation disorders		Neuromyopathic disorders	
	Pseudo-hypertrophic musc. dystr. Limb-girdle musc. dystr. Facio-scapulo-humeral dystr. Dermato and polymyositis		Amyothophic lateral sclerosis Infantile spinal muscle atrophy Peripheral polyneuritis		Myotonic dystr.	
Number of cases	58	100%	32	100%	10	100%
Abnormal biopsies	56	96.6%	28	87.5%	9	90%
Abnormal EMG	56	96.6%	32	100%	10	100%
Muscle biopsies:						
Myopathics	54	93.7%	0		2	20%
Denervations	0		26	81.3%	6	60%
Mixed	2	3.4%	0		1	10%
Type II fiber atrophies	0		2	6.2%	0	
Normals	2	3.4%	4	12.5%	1	10%
Electromyographies:						
Myopathics	48	82.8%	1	3.1%	3	30%
Denervations	2	3.4%	29	90.7%	7	70%
Mixed	6	10.4%	2	6.2%	0	
Normals	2	3.4%	0		0	

Table 6 — Incompatibility between electromyographies and muscle biopsies.

COMMENTS

Comparing our EMG data with those of current literature, we notice that some authors obtain lower indexes of abnormalities [4,9,13,37,43,53]. Only Hausmanowa-Petrusewicz & Jedrzejowska [35] obtained a higher abnormality rate (98 5%). Comparing our MB results with those reported, there was also a variation, where some obtained lower indexes [4,13,35] and others higher one [37,43], where Micaglio & col. [43] obtained 98.6%, but only studied primary myopathies, without including denervation cases.

The presence of inconsistencies varies according to the studies. Hausmanowa-Petrusewicz & Jedrzejowska [35] felt the lack of contribution of EMG for the diagnosis of myopathies in 2.2% and 1.1% in the cases of neurogenic origin; Schwartz & col.[53] refered 18.7% EMG not compatible or disagreed with the clinical diagnosis; Buchthal & Kamieniecka [13] felt lack of contribution in 2.65% of myopathies and 1.65% in the neurogenic disorders. For MB, Hausmanowa-Petrusewicz & Jedrzejowska feel lack of contribution in 10% of primary myopathies and 15.4% in neurogenic disorders; Buchthal & Kamieniecka [35] felt lack of contribution in 3.72% of myopathies and none for the biopsies of neurogenic origin. We were able to obtain an adequate correlation between the EMG and MB in 80% of the cases, studying *only one muscle*, similar to other published reports, who studied several muscles in the EMG and only one MB, such as Humphrey & Shy [37] with 85%; Schwartz & col.[53] with 79% and Black & col.[4] with 90%. Buchthal & Kamieniecka [13] found a concordance of 77% in myopathies and 91% in disorders of neurogenic involvement. This difference, compared with our results (Table 6), possible is due to the methods of study, since these authors examined several muscles during the EMG, and eventually this exam was repeated later [13].

Since Steinert [55] and Batten & Gibb [2] initial descriptions, the myotonic dystrophy was catalogued as a myopathy and classified among the muscular dystrophies. From

1966 onwards, several authors, studying histological and histochemical aspects [26,27], as well electrophysiological ones [41,48], suggested the existence of a denervation process, although study in the peripheral nerve failed to demonstrate structural alterations [49]. The investigation of phosphorilation in frozen erythrocytes and using techniques of magnetic resonance in erythrocytes [51] of patients with myotonic dystrophy, showed abnormalities suggesting alterations in the membrane of the cells of the organism. Several publications showed evidences of histological, physiological and biochemical abnormalities in myotonic patients, such as the heart [31,57], gastrointestinal smooth musculature [40], respiratory system with hypoventilation [3], ocular system with involvement of the crystalline, ciliary body and retina [36], abnormal production of immune-globulins [17], endocrine and metabolical disturbances [44,45,46], alterations of leucocyte membrane [54], baldness, cranial hyperostosis, mental retardation and abnormal behaviour [59]. This involvement could be at the stage of cell membranes, which would also be present in the muscle fibers, peripheral nerves and motor neurons, determining the muscle lesion, as well as in membranes of many different tissues [50]. Based on the above works, we think it is justified to take the myotonic dystrophy from the group of primary muscle disorders and also from the disorders which determine only muscular denervation. We think it should be catalogued in a separate group (neuromyopathies?), taking under consideration the mixed involvement found in the histological and electromyographical evidences, as we demonstrate in table 6.

Concluding, we think that the electromyography and muscle biopsy, using the parameters previously described and when only one muscle is examined, present almost the same differentiation indexes between the process of neurogenic and myopathic origin, avoiding some times long and painfull EMG tests. There was an agreement in 80% between the muscle biopsy and electromyography when only one muscle was studied, in relation to the nature of the diagnosis of the pathogenic process ($p<0.01$). This data permits us to select patients to be submitted to only one of the procedures (usually the MB), because permits us to induce a pathogenic and frequently a nosological diagnosis (the EMG rarely is able to do this). In the cases where only one of the procedures plus the clinical and laboratory findings unable us to reach a diagnosis, then both procedures are done and several muscles are examined in the EMG.

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