

Amyotrophic Lateral Sclerosis can Mimic Orthopedic Disease - Cases Report

Bárbara Aymé Hernández*

Department of Clinical Neurophysiologist, Cuban Neuroscience Center, Cuba

Submission: October 09, 2015; **Published:** November 02, 2015

***Corresponding author:** Bárbara Aymé Hernández, Department of Clinical Neurophysiologist, Cuban Neuroscience Center, Cuba; Tel: (53)7-2610659; Email: barbara@cneuro.edu.cu

Abstract

Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disorder of unknown cause. It is characterized by diffuse involvement of cortical, bulbar and spinal motor neurons. The diagnostic of this disorder is based in demonstrating of abnormalities of upper motor neurons with involvement of low motor neuron. In Europe and the United States, ALS affects about 2 people per 100 000 habitants per year. Neurophysiological studies are a basic tool for ALS diagnostic; they are useful to differentiate ALS from other neurological diseases. Image studies, especially Magnetic Resonance Image (MRI) is very useful to differentiate ALS from others diseases, like cervical spondylosic myelopathy, medullar tumor, syringomyelia, that look like ALS. We have wanted to report three cases of ALS patients who had short time illness evolution, they had orthopedic symptoms initially, and images studies show orthopedic abnormalities but them didn't justify clinical symptoms, electrophysiological studies support ALS diagnosis. Motor and Sensory nerve conduction studies, Electromyography and Magnetic Resonance Image were done to them. In conclusion orthopedic disease can coexist with ALS and it could be the reason of consult of the patients. Doctors have to confirm them to doing electrophysiological and image studies; they have been repeated at six months period in order to confirm progression of the disease.

Keywords: Amyotrophic Lateral Sclerosis (ALS); Electrophysiological Studies; Nerve Conduction Studies; Electromyography; MRI.

Abbreviations: ALS: Amyotrophic Lateral Sclerosis; ICD: International Statistical Classification of Diseases; MRI: Magnetic Resonance Image; EMG: Electromyography; LMN: Evidence of Lower Motor Neuron; UMN: Evidence of Upper Motor Neuron

Introduction

Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disorder of unknown cause. It is classified as G12.2 in International Statistical Classification of Diseases and Related Health Problems (ICD-10) of 2015. Every 90 minutes someone is diagnosed with amyotrophic lateral sclerosis. Most people with ALS live 2-5 years after their first signs of disease. About 10% of people with ALS survive at least 10 years [1-3]. It is characterized by diffuse involvement of cortical, bulbar and spinal motor neurons. The diagnostic of this disorder is based in demonstrating of abnormalities of upper motor neurons (hyperreflexia, hypertonia, clonus and Babinski sign) with involvement of low motor neuron (muscular atrophy, muscular weakness and fasciculation) [1-4]. The criteria diagnosis for ALS was defined in the World Federation of Neurology Meeting at Escorial, in 1994 [5] (Appendix).

In Europe and the United States, ALS affects about 2 people per 100 000 habitants per year. In Cuba there isn't exactly statistic about incidence and mortality by ALS, but highest

hospitals institution that dedicate to study neurological diseases like National Neurological Institute reports 2-4 cases at week, 8-16 monthly and 200 patients by year [1-4]. Neurophysiological studies are a basic tool for ALS diagnostic; they are useful to differentiate ALS from other neurological diseases. Image studies, especially Magnetic Resonance Image (MRI) is very useful to differentiate ALS from others diseases, like cervical spondylosic myelopathy, medullar tumor, syringomyelia, that look like ALS [6-11]. At present there is not an effective treatment to stop the disease [1]. We have wanted to report three cases of ALS patients who had short time illness evolution, they had orthopedic symptoms initially, images studies show orthopedic abnormalities but them didn't justify clinical symptoms, electrophysiological studies support ALS diagnosis.

Method

Three patients were evaluated at Electrodiagnosis department of "Frank País" orthopedic hospital. They consult by orthopedic symptoms, Magnetic Resonance Image (MRI) confirm orthopedic abnormalities, but they didn't justify all of clinical

symptoms, electrophysiological studies results supported Motor Neuron Disease.

Nerve conduction study

By segment motor nerve conduction study of Median, Posterior Tibial and Deep Peroneal nerves was done. Sensory nerve conduction of Median and Sural nerves was also done. Neuronica 5 equipment of Neuronica S.A was employed to do electrophysiological tests; we use conventional parameters for these tests [8-10].

Electromyography

Needle EMG were done to all patients. We evaluated Deltoides, Finger Common extensor, First Interosseus dorsal, Lateral Vastus, Anterior Tibial, Gastrocnemius and facial muscles of both side [8-10].

Magnetic resonance image (MRI)

MRI of cervical spine cord was done to all patients, with equipment of 0,35 Tesla. We analyzed sagittal view in T1 and T2, with 10 cm between the slices. MRI of dorsal and lumbar spine cord was done too to patient 1.

Case Reports

Case 1

Age: 52 years old.

Sex: male.

Evolution time: 3 months

He attempted by: Weakness of left lower limb, lumbar pain.

Antecedents: Trauma of left lower limb.

Presumptive diagnosis: Lumbar disc herniation with compression of lumbar roots.

Physical Exam: Fasciculation in muscle of four extremities, Hyperreflexia, no muscular atrophy, no clonus, no Babinski's sign, normal sensibility, normal muscular tone.

Electrophysiological studies

Nerve Conduction Study

- i. Moderate myelinic damage of both motor Median and posterior Tibial nerves.
- ii. Axonal damage of both Deep Peroneal nerves.
- iii. Moderate myelinic damage of both sensory Median and Sural nerves.

Electromyography

- i. Fasciculation (+4) in all of the exam muscles of four extremities.
- ii. High Frequency Repetitive Discharges.
- iii. Potentials of Fibrillation in distal muscles of left arm.
- iv. Mark polyphasic Motor Unit Potentials (MUP) with normal amplitude and duration.
- v. Max contraction pattern: Isolate in all of evaluates muscle.

Image studies

Cervical Spine MRI: It showed C5, C6 and C7 disc were herniated. Cervical discs were dehydrated and with degeneration changes. Vertebral bodies normal. No abnormalities in the signal at cord.

Dorso-Lumbal Spine MRI:

- i. Dehydration of L4-L5 and L5-S1 discs.
- ii. Small posterior osteophytes at L4-L5 spaces, without compression signs of the dural sac.
- iii. No abnormalities of the medulla (Figure 1-6).

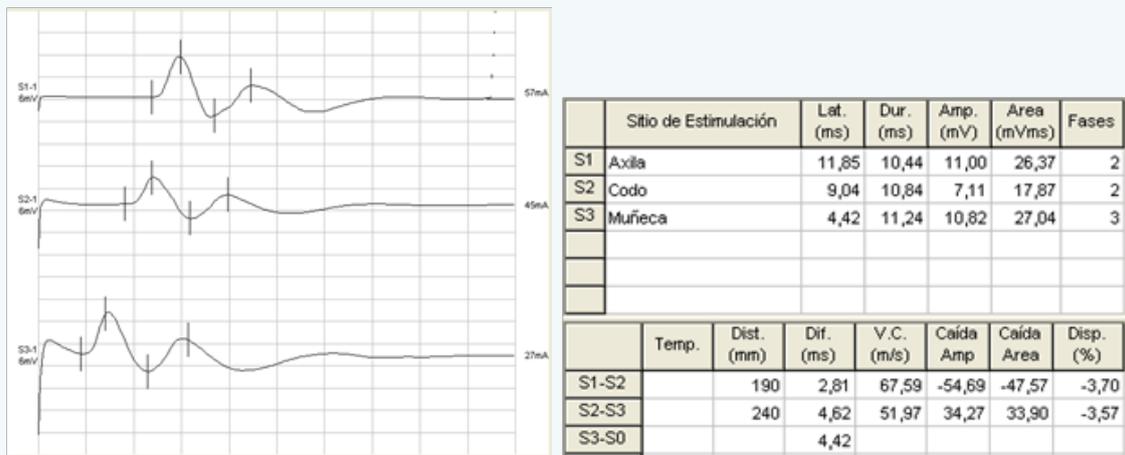
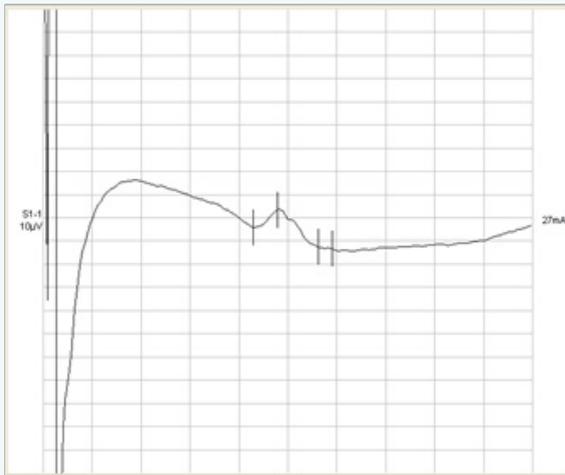


Figure 1: Motor nerve conduction study of right Median nerve of patient 1. Note slight enlargement of latency responses. Rest of parameters were normal.



	Sitio de Estimulación	Lat. (ms)	Dur. (ms)	Amp. (mV)	Area (mVms)	Fases	
S1	Debajo de la cabeza de...	10,24	12,05	0,63	2,22	1	
S2	Garganta del pie	4,42	10,24	0,17	0,38	4	
	Temp.	Dist. (mm)	Dif. (ms)	V.C. (m/s)	Caída Amp	Caída Area	Disp. (%)
S1-S2			5,82		-265,...	-482,...	17,65
S2-S0			4,42				

Figure 2: Motor nerve conduction study of right Deep Peroneal nerve of patient 1. Note diminish of amplitude response.



	Sitio de Estimulación	Lat. (ms)	Dur. (ms)	Amp. (µV)	Area (µVms)	Fases	
S1	Tercer dedo	4,30	1,61	7,48	6,38	2	
	Temp.	Dist. (mm)	Dif. (ms)	V.C. (m/s)	Caída Amp	Caída Area	Disp. (%)
S1-S0		180	4,30	41,89			

Figure 3: Sensory nerve conduction study of right Median nerve of patient 1. Note markedly enlargement of latency responses and diminish of nerve conduction velocity.



Figure 4: EMG of anterior Tibial muscle of patient 1 at slight contraction phase. Note increase of phase number and turns of the MUP.



Figure 5: MRI of cervical spine of patient 1 (T2 signal). Note slight disc herniation at C4-C5, C5-C6 and C6-C7.



Figure 6: MRI of dorsal and lumbar spine of patient 1 (T2 signal). Note small posterior osteophite at L4-L5.

Case 2

Age: 46 years old.

Sex: male.

Evolution time: 6 months.

Antecedents: Hypertension.

He attempted by: Weakness of right upper limb and cervical pain.

Presumptive diagnosis: Cervical disc herniation with compression of cervical roots.

Physical Exam: Weakness of four extremities, paralysis of right upper limb, Hyperreflexia in four extremities, changes in the voice, Babinski’s sign bilateral, Hypotonia in four extremities, Muscular atrophy in four extremities, predominantly in right upper limb, Fasciculation in muscle of four extremities, normal sensibility.

Electrophysiological studies

Nerve Conduction Study

- i. Axono-myelinic damage in both motor Median, Posterior Tibial and Deep peroneal nerves.
- ii. Moderate myelinic damage in sensory Median and Sural nerves.

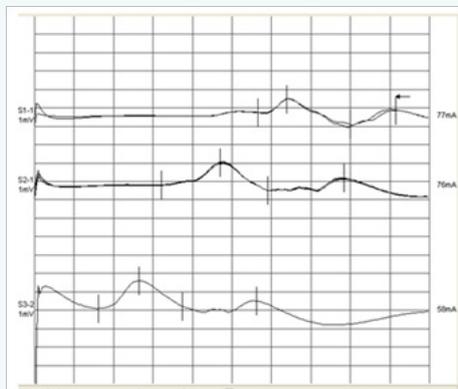
Electromyography

- i. Fasciculation in all evaluates muscles.
- ii. Fibrillation Potentials and Positive Sharp Wave in all of examined muscles.
- iii. MUP polyphasic with normal amplitude and duration.
- iv. Maxim contractile pattern: Isolated to simple oscillations.

Image studies

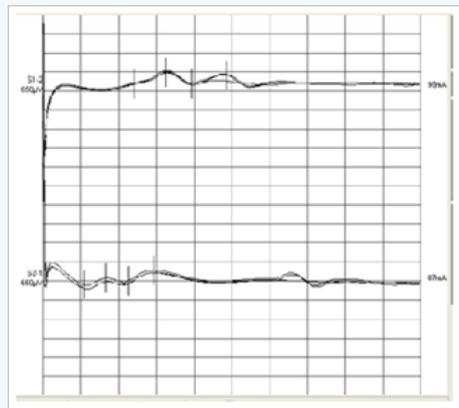
MRI of Cervical Spine

Disc Herniation at C5-C6 level (Figure 7-11).



Sitio de Estimulación	Lat. (ms)	Dur. (ms)	Amp. (mV)	Area (mVms)	Fases	
S1 Axila	16,99	10,48	0,71	1,45	2	
S2 Codo	9,64	13,86	1,20	3,55	3	
S3 Muñeca	4,82	12,05	1,51	4,70	1	
Temp.	Dist. (mm)	Err. (ms)	V.C. (m/s)	Calda Amp	Calda Area	Disp. (%)
S1-S2		7,35		41,00	59,20	-24,35
S2-S3	260	4,82	53,95	20,34	24,56	15,00
S3-S0		4,82				

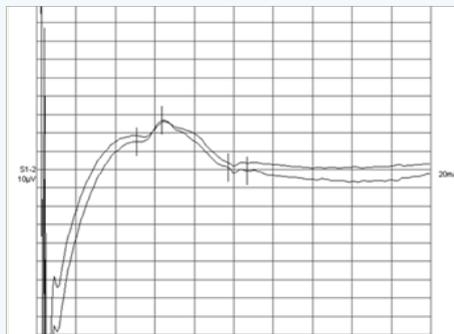
Figure 7: Motor nerve conduction study of right Median nerve of patient 2. Note moderate enlargement of latency responses and amplitude diminished.



Sitio de Estimulación	Lat. (ms)	Dur. (ms)	Amp. (mV)	Area (mVms)	Fases
S1 Debajo de la cabeza de...	12,05	12,25	0,40	3,79	1
S2 Garganta del pie	5,42	9,24	0,22	0,29	1

Temp.	Dist. (mm)	Dif. (ms)	V.C. (m/s)	Caída Amp	Caída Area	Disp. (%)
S1-S2		6,63		-79,79	-1,20...	32,61
S2-S0		5,42				

Figure 8: Motor nerve conduction study of right Deep peroneal nerve of patient 2. Note amplitude diminished notably.



Sitio de Estimulación	Lat. (ms)	Dur. (ms)	Amp. (µV)	Area (µVms)	Fases
S1 Tercer dedo	2,53	2,81	11,53	17,40	2

Temp.	Dist. (mm)	Dif. (ms)	V.C. (m/s)	Caída Amp	Caída Area	Disp. (%)
S1-S0	170	2,53	67,19			

Figure 9: Sensory nerve conduction study of right Median nerve of patient 2. Response is normal.



Figure 10: EMG of left anterior Tibial muscle at rest of patient 2. Note abundant Fibrillations potentials (A) and Fasciculations (B).

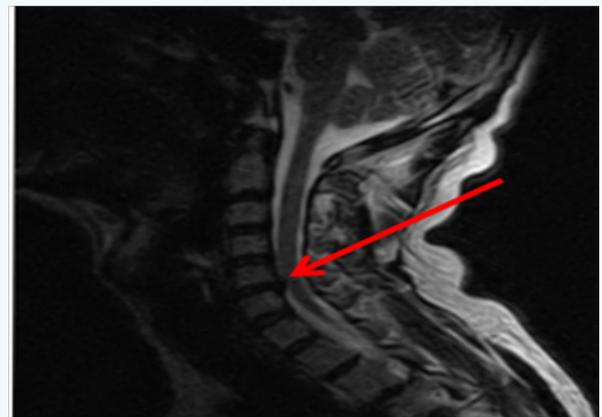


Figure 11: MRI of cervical spine of patient 2 (T2 signal). Note slight discal herniation at C5-C6.

Case 3

Age: 42 years old.

Sex: male.

Evolution time: 2 months

He attempted by fall down, trauma in right upper member.

Presumptive diagnoses: Arm trauma, Radial nerve trauma

Physical Exam: At first month he showed weakness of right arm and difficult to extend arm, at second month we start to show weakness in muscle of four extremities, Atrophy of distal muscle of both upper limbs, Fasciculation in muscle of four extremities, normal reflex, right Babinski's sign, normal sensibility.

Electrophysiological studies

Motor and Nerve Conduction Study: They were normal.

Electromyography

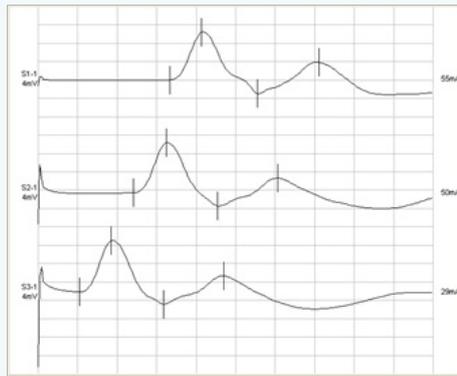
- i. Fibrillation potentials, Positive Sharp Wave and Fasciculations in all of examined muscles.
- ii. MUP polyphasic, amplitude increase and normal duration.

iii. Maxim contractile pattern: Isolated in four extremities.

Image studies

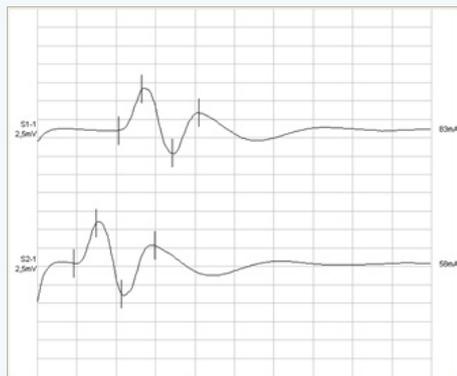
MRI of cervical spine:

It showed an osteophyte at vertebrae C7. No compression of the medulla (Figure 12-16).



Sitio de Estimulación	Lat. (ms)	Dur. (ms)	Amp. (mV)	Area (mVms)	Fases	
S1 Axila	10,00	11,33	10,41	24,93	3	
S2 Codo	7,23	10,96	10,85	26,09	3	
S3 Muñeca	3,13	10,96	11,20	26,71	1	
Temp.	Dist. (mm)	Dif. (ms)	V.C. (m/s)	Caída Amp	Caída Area	Disp. (%)
S1-S2	200	2,77	72,17	4,08	4,45	3,30
S2-S3	240	4,10	58,59	3,12	2,33	0,00
S3-S0		3,13				

Figure 12: Motor nerve conduction study of right Median nerve of patient 3. Responses are normal.



Sitio de Estimulación	Lat. (ms)	Dur. (ms)	Amp. (mV)	Area (mVms)	Fases	
S1 Debajo de la cabeza de...	10,24	10,24	5,65	16,05	2	
S2 Garganta del pie	4,62	10,24	5,52	14,35	3	
Temp.	Dist. (mm)	Dif. (ms)	V.C. (m/s)	Caída Amp	Caída Area	Disp. (%)
S1-S2	320	5,62	56,91	-2,31	-11,83	0,00
S2-S0		4,62				

Figure 13: Motor nerve conduction study of right Deep peroneal nerve of patient 3. Responses are normal.



Sitio de Estimulación	Lat. (ms)	Dur. (ms)	Amp. (µV)	Area (µVms)	Fases	
S1 Tercer dedo	2,73	2,17	10,04	10,83	2	
Temp.	Dist. (mm)	Dif. (ms)	V.C. (m/s)	Caída Amp	Caída Area	Disp. (%)
S1-S0	150	2,73	54,93			

Figure 14: Sensory nerve conduction study of right Median nerve of patient 3. Response is normal.



Figure 15: EMG of left Deltoid muscle at maximum contraction phase of patient 3. Note isolated pattern.



Figure 16: Right hand of patient 2. Note atrophy of First dorsal muscle .

Discussion

ALS is one of the most common neuromuscular diseases worldwide, and people of all races and ethnic backgrounds are affected. It is more common among white males. In 90 to 95 percent of all ALS cases, the disease occurs apparently at random with no clearly associated risk factors. Individuals with this sporadic form of the disease do not have a family history of ALS, and their family members are not considered to be at increased risk for developing it. About 5 to 10 percent of all ALS cases are inherited. The familial form of ALS usually results from a pattern of inheritance that requires only one parent to carry the gene responsible for the disease. Mutations in more than a dozen genes have been found to cause familial ALS. About one-third of all familial cases (and a small percentage of sporadic cases) results from a defect in a gene known as “chromosome 9 open reading frames 72,” or C9orf72. The function of this gene is still unknown. Another 20 percent of familial cases result from mutations in the gene that encodes the enzyme copper-zinc superoxide dismutase 1 (SOD1).

The age of our patients go from 42-52 years old. Most of the authors are agree with ALS is more frequent between 40 and 70 years old, although it could be appear in some young people; in which hereditary variant is suspect. It is most common among persons over age 60. All of our patients were male. ALS is more frequent in male than female, in a 2-3:1 proportion. ALS symptoms in the early stages of the disease can be similar to those of a wide variety of other, more treatable diseases or disorders like: Spondylotic Myelopathy, Multiple Sclerosis, Syringomyelia, Cord tumor, Disc herniated and others. All of

these differential diagnoses could be treated, whereas ALS does not have an effective treatment.

All of these diseases have an insidious start. In all of them patients have sensory abnormalities and pain. In ALS there is not clinical sensory abnormalities and pain either. Even though case 1 and 2 had pain, for that reason doctors thought in orthopedic disease, case 3 had right arm trauma antecedent; in this case doctors thought at first right radial nerve trauma, but in a month period, patients showed weakness of his left arm too [1-4,6].

Spondylotic Myelopathy appears in over 60 years old people, it is very similar to ALS, except Spondylotic Myelopathy patients have an intensive radicular pain, both diseases could be coexisting, electrophysiological studies are very similar too but MRI can confirm cervical spondylitis and cord compression. Multiple Sclerosis is a most diffuse diseases, it affect vision, audition, posture, eyes movement, balance, sphincter function. Nerve conduction studies show normal responses, EMG shows a central motor nervous system disturbance and MRI show demyelinating plaques in brain or in cord. Syringomyelia has most localize symptoms; it affects sensibility more than motor function, nerve conduction study could be normal or with predominance of sensory abnormalities, EMG shows localize abnormalities in relation of syringomielic cavity localization. MRI confirms the cavity topography at the cord. Cord tumor has localized symptoms in relation with topography of the tumor. Nerve conduction studies show normal responses; EMG shows localized abnormalities. MRI confirms tumor in the cord. Disc Herniation is a localized disease; it could show sensory and motor symptoms. Pain is very intense. Nerve conduction studies

are normal. EMG shows localized abnormalities in relation with root that are compressed by the herniated disc. MRI shows the disc herniation and the intensity of cord compression. In all of our cases MRI showed cervical spine abnormalities but them didn't justify the clinical finding examination [4].

Neurophysiological studies have suspected defined ALS in all cases. Neurophysiological evaluation is very important to confirm nervous system segment abnormality and could differentiate ALS of other diseases [11,12]. Álvarez Fiallo and col cited the Kothari study on 1998, in which was showed that neurophysiological studies results changed the initial diagnosis on 37% of ALS evaluated patients. Neurophysiological evaluation must be repeated at 6 months to show evolution of disease [13,14]. Motor nerve conduction study showed moderate abnormalities, fundamentally in lower limb nerves, in case 3 motor and sensory nerve conduction studies were normal. According to Gutiérrez Rivas nerve conduction studies could be normal in ALS if dying back degeneration has not occurred yet. In almost cases motor nerve conduction study show axonal damage, this is expression of dying back motor neuron degeneration, which appears after neuronal body degeneration [8].

Sensory nerve conduction studies showed moderate myelinic damage of upper and lower nerves. Some years ago some authors insured that sensory nerve conduction studies must be normal to ALS diagnosis. This planning has been relaxed because post-mortem studies in ALS have showed that there is degeneration of sensory nerves too, the most affected neurons are in dorsal root ganglia and in posterior horn. This degeneration is most slow and less intense that in motor neurons. This sensory abnormality doesn't express clinically, but it could be detect by electrophysiological tests [15]. Sensory function in patients with amyotrophic lateral sclerosis (ALS) is thought to be normal; however, there is convincing morphologic evidence that sensory systems are affected in addition to motor systems. Shefner and collaborator in 1991 reported nine of 18 patients had abnormally reduced minimum conduction velocity, even when peak-to-peak amplitude and maximum conduction velocity (calculated from the latency to the initial positive peak) were normal. Only 3 of 18 patients showed abnormalities in peak-to-peak amplitude [16].

Pugdahl et al. [17] in 2007 reported that of 41 sensory nerves examined in the ALS patient group, 32 (78%) nerves had abnormal sensory nerve conduction parameters 10 had decreased sensory nerve conduction velocity, 14 had decreased sensory response amplitude and 2 nerves did not elicit any response. Morphological findings of sensory nerve biopsies from patients with ALS have shown axonal degeneration, segmental demyelination and a reduced number of large neurons of the L5 dorsal root ganglion [17,18]. Electromyography was abnormal in all of patients. It showed neurogenic pattern in 3 of anatomical areas and it made evident upper and lower motor neuron damage. It is a most sensitive electrophysiological test in ALS

diagnosis and let to sit the stage of disease and to prognosis the evolution [8,11,12].

All of patients that we evaluated consulted the doctor because orthopedic symptoms like cervical and lumbar pain; it was accompanied by weakness and/or muscle atrophy of one member. Physical examination, electrophysiological and images techniques reject orthopedic diseases like cause of patient's symptoms. Cervical spondylotic myelopathy is the most important orthopedic disease to be considerate to ALS differential diagnosis. In some occasions electrophysiological test is very similar in both diseases, with exception that Spondylotic myelopathy respect bulbar muscle. Some authors like Yamada in 2003 have reported that in occasions coexist both diseases (Spondylotic Myelopathy and ALS); in these occasions ALS is frequently complicated by cervical Spondylotic myelopathy. When the presence of ALS is missed, cervical spinal surgery is performed followed by progressive worsening of the motor symptoms despite successful treatment of cervical spondylosis [19,20,21].

Conclusion

In conclusion orthopedic disease can coexist with ALS and it could be the reason of consult of the patients. Doctors have to confirm them to doing electrophysiological and image studies; they have been repeated at six months period in order to confirm progression of the disease.

References

1. Kiernan MC, Vucic S, Cheah BC, Turner MR, Eisen A, et al. (2011) Amyotrophic lateral sclerosis. *Lancet* 377(9769): 942-955.
2. McDermott CJ, Shaw PJ (2000) Diagnosis and management of motor neuron disease. *BMJ* 336(7645): 658-662.
3. Dugdale DC, Hoch DB, Zieve D (2010) Amyotrophic lateral sclerosis. A.D.A.M. Medical Encyclopedia.
4. Rooper AH, Brown RH, Adams, Victor's (Eds.), (2005) Principles of neurology. McGraw-Hill, New York, USA, pp. 120-158.
5. Brooks BR (1994) El Escorial World Federation of Neurology criteria for the diagnosis of amyotrophic lateral sclerosis. *J Neurol Sci* 124(Suppl1): 96- 107.
6. García Gálvez P, Cambón RS, Higes P, Sánchez Migallón MJ, Yustazquierdo A (2011) Enfermedad de la motoneurona. *Medicine* 10(77): 5200-5208.
7. Fernández-Lerones MJ, de la Fuente-Rodríguez A (1999) Diagnóstico electrofisiológico. In: Mora JS (Ed.), Esclerosis lateral amiotrófica. Una enfermedad tratable Barcelona: ProusScience, S.A pp. 157- 175.
8. Gutiérrez Rivas E, et al. (Eds.), (2000) Manual de Electromiografía Clínica. Barcelona: ProusScience, S.A, pp. 177-186.
9. Behnia M, Kelly JJ (1991) Role of electromyography in amyotrophic lateral sclerosis. *Muscle Nerve* 14(12): 1236-1241.
10. Santos Anzorandia CM (2000) El Abecé de la Electroneuromiografía clínica. La Habana: Ciencia Médicas, pp. 183-187.
11. Espadaler Gamissans JM (1996) Exploración electrofisiológica en las

- enfermedades de la motoneurona. *Neurologia I I(Supl 5)*: 20-28.
12. Cornblath DR, Kuncel RW, Mellits ED, Quaskey SA, Clawson L, et al. (1992) Nerve conduction studies in amyotrophic lateral sclerosis. *Muscle Nerve* 15(10): 1111-1115.
 13. Álvarez Fiallo R, Medina Herrera E (2004) La neurofisiología en el estudio de las enfermedades neuromusculares, desarrollo y limitaciones actuales. *Rev Cub Med Mil* 33(3): 15-18.
 14. Fiallo CRA, Anzorandia CCS, Herrera CEM (2007) Diagnóstico electromiográfico de las enfermedades neuromusculares. *Rev Cub Med Mil* 36(1): 1-5.
 15. Pavlovic S, Stenic Z, Kectkarevic D, Rakocevic-Stojanovic V, Lavrnic D, et al. (2003) Sensory abnormalities in patients with amyotrophic lateral sclerosis. *Muscle and Nerve* 12(Suppl): 145.
 16. Shefner JM, Tyler HR, Krarup C (1991) Abnormalities in the sensory action potential in patients with amyotrophic lateral sclerosis. *Muscle Nerve* 14(12): 1242-1246.
 17. Pugdahl K, Fuglsang-Frederiksen A, De Carvalho M, Johnsen B, Fawcett PRW, et al. (2007) Generalized sensory system abnormalities in amyotrophic lateral sclerosis: a European multicentre study. *J Neurol Neurosurg Psychiatry* 78(7): 746-749.
 18. Iglesias C, Sangari S, El Mendili MM, Benali H, Marchand-Pauvert V, et al. (2015) Electrophysiological and spinal imaging evidences for sensory dysfunction in amyotrophic lateral sclerosis. *BMJ Open* 5: e007659 doi:10.1136/bmjopen-2015-007659.
 19. Yamada M, Furukawa Y, Hirohata M (2003) Amyotrophic lateral sclerosis: frequent complications by cervical spondylosis. *J Orthop Sci* 8(6): 878-881.
 20. Yoshor D, Klugh A 3rd, Appel SH, Haverkamp LJ (2005) Incidence and characteristics of spinal decompression surgery after the onset of symptoms of amyotrophic lateral sclerosis. *Neurosurgery* 57(5): 984-989.
 21. Hernández Hernández A (2013) Rol de los estudios neurofisiológicos en el diagnóstico diferencial entre la esclerosis lateral amiotrófica y la mielopatíaespondilótica cervical. *Acta Neurol Colomb* 29(1): 53-59.

Appendix

El Scorial Criteria for ALS diagnosis

1. Presence of:
 - i. Evidence of lower motor neuron (LMN) degeneration by clinical, electrophysiological or neuropathologic examination.
 - ii. Evidence of upper motor neuron (UMN) degeneration by clinical examination.
 - iii. Progressive spread of symptoms or signs within a region or to other regions, as determined by history or examination.
2. Absence of:
 - iv. Electrophysiological and pathological evidence of other disease processes that might explain the signs of LMN and/or UMN degeneration.
 - v. Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs.

Diagnostic Categories

Clinically Definite ALS: Is defined on clinical evidence alone by the presence of UMN, as well as LMN signs, in three regions.

Clinically Probable ALS: is defined on clinical evidence alone by UMN and LMN signs in at least two regions with some UMN signs necessarily rostral to (above) the LMN signs.

Clinically Probable - Laboratory-Supported ALS: Is defined when clinical signs of UMN and LMN dysfunction are in only one region, or when UMN signs alone are present in one region, and LMN signs defined by EMG criteria are present in at least two limbs, with proper application of neuroimaging and clinical laboratory protocols to exclude other causes.

Clinically Possible ALS: is defined when clinical signs of UMN and LMN dysfunction are found together in only one region or UMN signs are found alone in two or more regions; or LMN signs are found rostral to UMN signs and the diagnosis of Clinically Probable - Laboratory-supported ALS cannot be proven by evidence on clinical grounds in conjunction with electrodiagnostic, neurophysiologic, neuroimaging or clinical laboratory studies. Other diagnoses must have been excluded to accept a diagnosis of clinically possible ALS.

Clinically Suspected ALS: it is a pure LMN syndrome, wherein the diagnosis of ALS could not be regarded as sufficiently certain to include the patient in a research study. Hence, this category is deleted from the revised El Escorial Criteria for the Diagnosis of ALS.