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# Sporadic amyotrophic lateral sclerosis. A clinical analysis

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**ABSTRACT**

**Objective.** To illustrate that sporadic amyotrophic lateral sclerosis (ALS) may be caused by ischemia in the intraparenchymal territory of the anterior spinal artery (ASA) and/or anterior-ventral spinal arteries (AVSAs). **Case report.** A 56-year-old woman presented clinical data of spinal and bulbar forms of ALS. Previously she was attended in several neurological centers. **Results.** In 2002 the patient began with paresthesias of the fingers in both hands. Three years later, she presented fasciculations in her left hand and later on, in the right hand, ascending to the forearm, arm and shoulder girdle muscles. Since October 2008, she presented fasciculations in thighs and legs, and finally, bulbar symptoms. The examination revealed dysarthria, weak voice, fasciculations and paresis in the tongue. Spastic tetraparesis and muscular atrophy, predominantly in the upper limbs. The superficial and deep sensory signs were normal. **Conclusions.** The onset and clinical course of the symptoms in this patient, it suggests that sporadic ALS is of ischemic origin in the intraparenchymal territory of the ASA and/or AVSAs, secondary to vascular anomalies and atherosclerosis.

***Key words:*** *Amyotrophic lateral sclerosis, anterior spinal artery, anterior sulcal arteries, anterior-ventral spinal arteries, vertebral arteries, anterior radicular arteries.*

**RESUMEN**

**Objetivo.** Ilustrar que la Esclerosis Lateral Amiotrófica (ELA), puede ser causada por isquemia en el territorio intraparenquimatoso de la arteria espinal anterior (AEA) y/o en las arterias espinales antero-ventrales (AEAVs). **Caso clínico.** Mujer de 56 años de edad, presentó datos clínicos de las formas espinal y bulbar de ELA. Previamente, fue atendida en varios centros neurológicos. **Resultados.** En 2002, la paciente empezó con parestesias en los dedos de las manos. Tres años después, presentó fasciculaciones en la mano izquierda y luego en la mano derecha; ascendiendo a los antebrazos, brazos y hombros. Desde octubre 2008, presentó fasciculaciones en muslos y piernas. Y finalmente, síntomas bulbares. El examen neurológico reveló disartria, voz apagada, fasciculaciones y paresia de la lengua. Tetraparesia espástica y atrofia muscular, predominantemente en los miembros superiores. La sensibilidad superficial y profunda fueron normales. **Conclusión.** El comienzo y curso clínico de los síntomas en la paciente, sugieren que la ELA esporádica, es de origen isquémico en el territorio intraparenquimatoso de la AEA y/o en las AEAVs, secundario a anomalías vasculares y aterosclerosis.

***Palabras clave:*** *Esclerosos lateral amiotrófica, arteria espinal anterior, arterias sulcales anteriores, arterias espinales antero-ventrales, arterias vertebrales, arterias radiculares anteriores.*

## INTRODUCTION

Up to date, all the researchers inform that the specific cause of amyotrophic lateral sclerosis (ALS) is not known. But, a recently report patient[1,2] suggest that the primary cause of the bulbar form of ALS can be of ischemic origin, in the intraparenchymal territory of the anterior-ventral spinal arteries (AVSAs ).[2-4]

In this paper, I analyzed the clinical case of a woman with spinal and bulbar forms of ALS who was admitted to the Hospital for diagnosis and therapeutic management. An omental transplantation was proposed to the patient and her family, but the surgery was not performed.

## CASE REPORT

A 56-year-old woman was attended by tetraparesis and bulbar symptoms. In 2002, the patient began with paresthesias of the fingers in both hands. The onset of the sensory disorders was insidious and course undulating (periods of clinical improvement alternated with those of worsening) during almost 4 years and then, disappeared. Since 2005, she started with fasciculations in the interosseous muscles of her left hand, followed by other muscles of the hand, forearm, arm and shoulder girdle region. Two

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years later, she began with fasciculations in the thenar eminence and interosseous muscles of the right hand, ascending gradually to the forearm, arm and shoulder girdle muscles. Simultaneously she presented weakness and muscular atrophy in the upper limbs. Besides this, since October 2008 she began with fasciculations in the muscles of the thighs and legs. The onset of this motor deficit was insidious, course undulating and progressive Finally,15 months before her admission she presented weak voice and progressive difficulty in pronouncing words, swallowing and breathing (Figure 1). Sometimes she presented fits of apnea for a few seconds. She never presented bladder and rectal disorders.

During these years of disease, she was attended in several neurological centers and in all of them, she was diagnosed of ALS. She received multiple treatments and rehabilitation. In 2007, several electrodiagnostic tests revealed poor or absence of motor neuroconduction in the cubital, median and radial nerves. Likewise, a computerized tomography scans showed to the encephalon and cervical cord normal.

### *Past and family history*

The patient was a cigarette smoker since she was 25 to 40 years old and then, was exposed to organic solvents. There was no family history of similar diseases.

### *Examination (April 2010)*

The neurological examination revealed slight dysarthria, moderate weak voice, moderate impairment of the nausea reflex, fasciculations and paresis in the tongue. Respiration was superficial and of the abdominal type. Spastic tetraparesis: upper limbs in degree 1 to 3 and lower limbs in degree 4 to 5.Moderate muscular atrophy in the upper extremities, especially in the small muscles of the hands (Aran-Duchenne´s hands), and slight muscular atrophy in the lower limbs. The atrophy was asymmetrical, predominantly in the left upper limb and with winged scapulae. Hyperreflexia in the upper and lower extremities. Bilateral Hoffmann sign and by contrast, absence of Babinski sign. The superficial and deep sensory signs were normal. Her blood pressure was 120/75 mmHg, hemoglobin 14.3 gr%, blood glucose 110 mg% and creatinin 0.90 mg%.

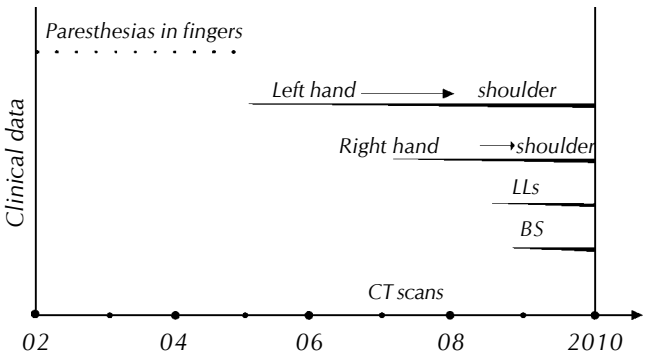


***Figure 1.*** *Clinical course in years. The motor deterioration was initiated in the left hand and then in right hand, lower limbs (LLs) and finally, bulbar symptoms (BS). Computed tomography (CT) scans.*

## DISCUSSION

There is no doubt that this patient began with a spinal form (progressive spinal muscular atrophy, PSMA) of ALS and six years later, was incorporated a bulbar form (Figure 1). For this reasons, to clarify the pathogenesis of this disease, I wish to comment the relation between the clinical course of the symptoms and the affected nervous tissue.

At the C5-T1 level, normally the anterior horns and medial portions (constituted by pyramidal axons of the upper extremities) of the lateral corticospinal tracts are supplied by two small branches, the anterior sulcal arteries with a mean diameter of 0.21 mm (range 0.06 to 0.40 mm)[5 6] originated from the anterior spinal artery (ASA).Often can exist common anterior sulcal trunks that after a short distance are bifurcated in right and left anterior sulcal arteries. Each anterior sulcal artery supplies only its own half of the cord in the vast majority of cases.[6]

In the cervical cord, the ASA with an average caliber of 0.75 mm (range 0.34 to 1.12 mm) receives arterial blood from two origins.[3-6] First, by means of AVSAs originated from the V4 segments of the vertebral arteries and Second, through anterior radicular arteries arising from deep muscular branches of the thyrocervical and costocervical arteries.

Based on the above mentioned data and the clinical course in this woman, I believe that the lesion was initiated in the posterior horns (laminae VI to IV) of the cervical cord, and three years later, in the lateral end of the left anterior horn, with progression towards the medial end, and then, the same agent affected to the right anterior horn. Moreover and simultaneously, the medial portions of the lateral corticospinal tracts were also involved. In other words, cervical PSMA (a spinal form of ALS)[7 8] seems be caused by progressive ischemia in the intraparenchymal territory of the ASA, which affect to the pyramidal fibers of the upper extremities and lower motor neurons at cervical cord level (Figure 2).

Therefore, the spinal form of ALS is not degenerative but of microvascular origin; due to a deterioration of the blood flow through the anterior radicular arteries, seconda-

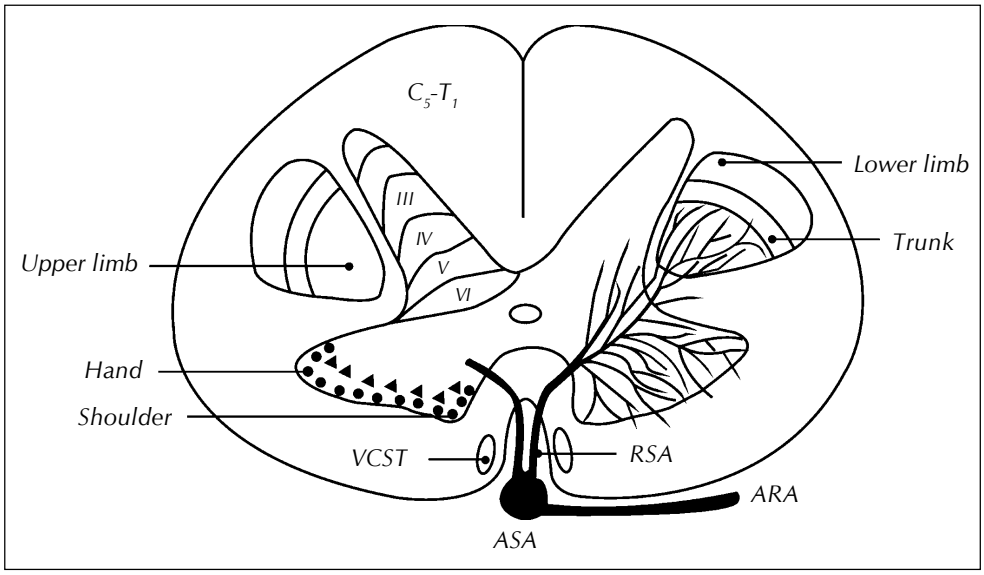


***Figure 2.*** *Diagram of the cross-sectional vascular anatomy of the cervical cord. ASA, anterior spinal artery. ARA, anterior radicular artery. RSA, right sulcal artery. VCST, ventral corticospinal tract.*

ry to vascular anomalies and atherosclerotic plaques located at the mouths of the thyrocervical and/or costocervical arteries originating from the subclavian arteries.[6 9] These arteries usually protect the vascularization of the cervical cord, when there are stenosis or occlusion at the origin of the V1 segments of the vertebral arteries. In contrast, in my opinion, an omental transplantation on the anterior, lateral and posterior surfaces of the cervical cord, it could improve this form of ALS; because, previous experiences have demonstrated that placing omental tissue on the injured spinal cord, it produced neurological improvement.[10,11]

In relation to the bulbar form of ALS, a report patient previously has showed[1 2] that the pyramids (constituted by pyramidal fibers destined to the upper limbs ,trunk and lower limbs) and motor nuclei of the cranial nerves XII and both of them nucleus ambiguous[12] were affected by progressive ischemia in the intraparenchymal territory of the AVSAs;[1 3,4] because in contrast to this, an omental transplantation on the anterior, lateral and posterior surface of the medulla oblongata produced functional recovery of the residual nervous tissue in the pyramids and motor nuclei in the medulla oblongata,[1] and in the manner of other "neurodegenerative diseases",[11,13] neurological improvement was better during the first days and weeks after the surgery than in the following months.[1,2] In this respect, the omentum is the best tissue for developing vascular connections with adjacent and underlying zones, and through the omental neovessels, the ischemic parenchyma receives an increase in blood flow, oxygen, neurotrophic factors, neurotransmitters, adipocytokines and omental stem cells.[2,11]

## CONCLUSIONS

Based on the neurological improvement obtained into patients with injured spinal cord and in a patient with bulbar form of ALS after an omental transplantation, I postulate that the cervical PSMA may also be stopped and improved with the same neurosurgical technique. However, future studies in large animal models will be necessary to further define the microvascular role in the pathogenesis of this disease.

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