

SHORT REPORT

Motor neuron disease after electric injury

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Abstract

The occurrence of motor neuron disease after electrical injury in six patients is reported and compared with patients from the literature. The patients were five men with spinal onset and one woman with bulbar motor neuron disease after electric shock. Two patients were struck by lightning and four by industrial electric shock. For all six of them, the disease started at the site of the electrical trauma. The mean delay for onset of motor neuron disease was 44 months. In four of the spinal patients the disease progressed slowly with mild handicap after several years. For the fifth patient, improvement was noted progressively. The patient with bulbar disease died 26 months after onset. A link between electric shock and motor neuron disease is likely, given the homogenous profile of the patients both in the five spinal cases presented here and in the literature. Bulbar onset has not been reported to date. However, in this patient the long delay between the electrical injury and motor neuron disease, together with the rapid evolution may suggest a chance association.

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Motor neuron disease is a rapidly progressive and fatal degenerative disorder involving either upper or lower motor neurons, or both.¹ The aetiopathogeny of the disease is unknown. However, in a few cases the disease has been linked with genetic abnormalities.² Several reports have pointed out the possibility of motor neuron disease after electrical injury such as industrial electric shock or lightning.³ As early as 1889, Charcot noted that the nervous system could be affected by an electrical injury.⁴ Delayed neurological sequelae after electrical injuries have been reported in the literature and different reports have focused on the occurrence of motor neuron disease after electrical injury. The neurological sequelae after being struck by lightning are polysymptomatic and those of industrial injuries are more homogenous.⁵ Spinal cord damage is the most common of the permanent delayed sequelae of electrical injury and occurs when the path of the current is either from arm to arm or from arm to leg. It has been stressed that permanent

spinal cord sequelae are usually seen when weakness develops within days or years after the shock.⁶ The spinal forms resemble either lower motor neuron syndrome, amyotrophic lateral sclerosis, or transverse myelitis. Sensory changes in the affected limb are common.⁷ Onset of the disease is at the site through which the shock entered, and may spread or remain localised.⁶ The interval between the electrical injury and the onset of motor neuron disease has no limits (days to decades).³ Recovery, when it occurs, is usually slow and incomplete. We report six cases of motor neuron disease after electrical injury, and compare them with 65 cases from the literature.

Case reports

All patients underwent comprehensive biological and radiological investigations to rule out differential diagnosis of degenerative motor neuron disease. They all had complete electroneuromyography. In all cases, a denervation pattern was found corresponding with motor unit potentials (MUPs) with increased duration and amplitude associated with a high firing rate (>15 Hz) and decreased recruitment.

PATIENT 1

A 56 year old man was struck by lightning in the right hand while using an ice axe during a hiking trip in the mountains. The current made its exit through his right foot. Six months later he complained of weakness in his right upper and lower limbs. Two years later there was diffuse amyotrophy and weakness of all muscle groups of the right upper limb without fasciculations. Deep tendon reflexes were normal. The plantar responses were both flexor. Pain and paraesthesia appeared immediately after the trauma and were still present in his right hand. There was sensory loss for temperature on the right upper limb. Neurography was normal except for a decreased amplitude for the compound muscle action potentials (CMAPs) on motor nerves on the right upper and lower limbs. Myography disclosed a denervation pattern on all four limbs. He was diagnosed as having progressive lower motor neuron disease, which after 3 years had not progressed.

PATIENT 2

Seven months after having experienced a direct electric shock (380 V) to the left hand by turning the door knob of a door which was touching a short circuit in the wall, a 42 year old man

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complained of fasciculations and weakness of the left hand. Twenty two months after the trauma there was severe amyotrophy of the left hand associated with muscle weakness in the left upper limb rated 1/5 on distal and 4/5 on proximal muscles (Medical Research Council (MRC) scale). Fasciculations were present on the tongue. Deep tendon reflexes were brisk on all four limbs. Plantar reflexes were flexor. Sensory examination and neurography were normal. Myography disclosed a denervation pattern on all four limbs. He was diagnosed with probable amyotrophic lateral sclerosis/motor neuron disease. After more than 3 years the patient's course showed slow progression of muscle weakness, but with a mild handicap (ALS functional rating scale 35/40).

PATIENT 3

Ten days after he had been shocked by 380 V from an electric cable on the right hand, a 68 year old man complained of weakness in his right upper limb. Three months after the trauma physical examination disclosed muscle weakness in the right hand rated 3/5 on the MRC scale. Deep tendon reflexes were absent on all four limbs but the plantar responses were both extensor. There were fasciculations on the right deltoid. Sensory examination and neurography were normal and myography showed a denervation pattern on all four limbs and on the abdominal muscles. He was initially diagnosed as having amyotrophic lateral sclerosis, but his condition progressively improved. After 12 years the patient had no complaints and physical examination showed a discrete weakness in the right hand rated 4+/5 on the MRC scale. We considered him to have an almost complete remission of a motor neuron disease after 12 years of follow up.

PATIENT 4

A 50 year old man had an electric shock to his right hand in April 1999, after coming into contact with a 380 V cable. Immediately after the trauma he complained of pain and paraesthesia of the right hand. Sixteen months later, there was a progressive motor deficit of the right arm associated with amyotrophy and fasciculations. Deep tendon reflexes were present in the atrophied muscles of the right arm and were brisk elsewhere. Plantar responses were both flexor. The patient described distal paraesthesia on the right hand, without objective sensory loss. Electroneuromyography confirmed lower motor neuron involvement with a severe denervation pattern on the right upper limb and on the legs. He was considered as having probable amyotrophic lateral sclerosis/motor neuron disease.

PATIENT 5

Three years after he had had an electric shock to his left foot by a 380 V cable a 26 year old man complained of walking difficulties. Physical examination showed a motor deficit in dorsal flexing the left foot, amyotrophy of the left extensor digitorum longus, and amyotrophy of the lateral peroneus muscles. There were no

fasciculations. Deep tendon reflexes were normal. The left plantar response was extensor. Electroneuromyography showed a denervation pattern on all four limbs. He was considered as having a possible amyotrophic lateral sclerosis/motor neuron disease. The motor deficit progressively extended and the patient died of respiratory failure after 107 months.

PATIENT 6

A 67 year old woman presented with dysarthria and dysphagia. Eighteen years earlier she had been struck by lightning while talking on the phone, holding the receiver to her left ear with her left hand. The electric current came out of her left thigh, causing superficial burning of her skin. Physical examination showed brisk deep tendon reflexes on her four limbs, and also a brisk jaw jerk. Plantar responses were both extensor. She had dysarthric speech, dysphonia, and dysphagia, associated with tongue atrophy and fasciculations. Neurography was normal. Myography showed a denervation pattern on all four limbs, on the abdomen, and on the tongue. She was considered as having definite amyotrophic lateral sclerosis/motor neuron disease, with bulbar onset. She died in her sleep 26 months after the onset.

Review of the literature

A 6 year old boy injured by a 220 V power line who developed amyotrophy of the traumatised upper limb and shoulder 15 months later, was reported on by Hoel.⁶ Physical examination disclosed "lively" deep tendon reflexes, and a stiff gait. The patient was considered as having a typical clinical picture of amyotrophic lateral sclerosis, which continued to advance over several years.

Farrell and Starr described a 67 year old man who was injured in his right upper limb by 18 000 V from a high tension line. Immediately after the trauma, the patient complained of pain in the right hand and left foot, where the current had made its exit. Two years later he complained of weakness and numbness of his left leg. Physical examination showed amyotrophy, fasciculations, and deficit of the left quadriceps. The left patellar tendon reflex was absent. There was decreased sensation to pin, touch, temperature, and vibration on the left thigh. Neurography was normal, whereas myography showed a denervation pattern on the left upper and lower limbs. Almost 3 years after the electrical trauma the patient's course had remained unchanged.⁷

Panse reported 17 cases from the literature and one personal case of electrical injury caused by lightning. Sex and age of the patients and the site of the trauma were not specified for all the cases. The interval between the lightning and the onset of disease was within months. They all had amyotrophy, some patients presented with spasticity, there was sensory involvement in two patients, and one patient had bladder and rectum paralysis. Four of them improved within a few years. The personal case that he qualified as "typical" was a 26 year old soldier struck by lightning in the back. Two months later, he developed paralysis

of the left arm with amyotrophy and absent deep tendon reflexes. He also had a spastic right leg. Twelve years later his clinical state remained unchanged.⁵

Panse also reported on 20 patients considered as having spinal atrophy after industrial electrical injury. Sex and age were not specified. The delay to onset was between 2 days and 1 year. The onset was on the traumatised limb in all but one, the exception being the occurrence of the first symptoms on the leg where the current made an exit. Two patients had sensory involvement and one had urinary incontinence. Two of them recovered.⁵

Panse reviewed eight cases of upper and lower motor neuron involvement considered as having amyotrophic lateral sclerosis. They were mostly men under the age of 56. The source of the shock was industrial electricity. The delay to onset was between a few weeks to a few months. The evolution was variable (death between 6 months and 7.5 years).⁵

Sirdofsky and Rollin reported on a 42 year old man who received an electric shock (110 V) which entered through his left arm and made an exit from the left leg. Within 2 weeks the patient developed cyanosis of the left hand, coolness, loss of nail growth, and fasciculations. Three months later physical examination showed weakness, amyotrophy, and fasciculations on the left upper limb. The left plantar response was extensor and deep tendon reflexes on the left side were brisk. There was decreased sensation to pinprick on the same side. Neurography disclosed low median and ulnar CMAPs on the left upper limb. Myography showed denervation of the left arm. The symptoms progressed and the patient died 33 months after the electrical trauma. Repeated MRI showed cervical cord atrophy. Neuropathology showed classic changes of amyotrophic lateral sclerosis.⁶

Gallagher and Talbert reported on 17 patients presenting with motor neuron disease after electric shock. All were men and younger than 45. They had all experienced industrial electrical injury. Five developed the disease within 36 months of the electrical accidents and the others not until decades later. In eight of the patients the first signs appeared in the hand; in seven in a lower limb; in one in the shoulder; and in one both in a leg and an arm. All were diagnosed as having amyotrophic lateral sclerosis. All but one were alive at the time of the publication.³

Discussion

We report on one woman and five men presenting with motor neuron disease after electrical trauma. Two patients were injured by lightning and four by industrial electricity. Mean age of the patients was 51.5 years. The woman had a bulbar onset and the five men had a spinal onset. For all of them the disease started at the site of electrical trauma. The delays for onset of motor neuron disease were 10 days, 21 days, 6 months, 7 months, 33 months, and 18 years. One patient recovered after 9 years and the other spinal patients had slow evolution. The follow up periods were 24, 26, 40, 41, 107 months, and 12 years. Two of

the patients had sensory signs in the traumatised limb. None of them had dysautonomic abnormalities. Two patients died, 26 and 107 months after the onset of the disease. They all had a motor neuron syndrome. One had predominant lower motor neuron syndrome, one had a cord syndrome with recovery, three had a profile compatible with possible amyotrophic lateral sclerosis/motor neuron disease, and one had definite amyotrophic lateral sclerosis/motor neuron disease. Interestingly spasticity was absent in all of our patients

The patients described in the literature were all males whereas our series included one female patient. The onset of the symptoms was always at the traumatised site both in our series and in the literature except for two patients reported by Panse and Farrell, where the beginning of the disease was at the exit point.^{5,7} For the interval between the trauma and the occurrence of symptoms, we noticed the same variable period as in the literature (days to years).^{5,7} Sensory symptoms were present for two of our patients. Both had immediate pain after the trauma and one of them had an objective sensory deficit. It has been reported that the immediate pain and paraesthesia may fade in a few days or be associated with sensory loss.⁶ Finally, both in the literature and in our series all spinal patients seemed to have a slow evolution with mild handicap. The fact that in the literature all the reported patients were male raises the question of a possible genetic factor. However, not all men who receive an electric shock develop motor neuron disease, which would reinforce the possibility of a combination of genetic and environmental factors.

The bulbar patient differs from the literature data in five ways: her sex; the very long delay between the electrical injury and the onset, the source of the electric shock (lightning); the bulbar onset; and the faster evolution compared with the spinal patients. Thus it is more difficult for this patient to suggest a relation between the electrical trauma and the motor neuron disease.

Although rare, electrical trauma should be more often considered as a possible cause of motor neuron disease. Even if the pathogenic relation between the electrical trauma and motor neuron disease is difficult to ascertain, it is noteworthy that in these cases the disease started at the site of the electric trauma, and that patients have a mild handicap after several years. This has already been reported in the literature and the evidence from our five spinal patients supports this idea.

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