Case Report

Rehabilitation Challenge of Motor Neuron Disease Following Occupational Exposure to Magnetic Fields and Electric Shocks: Case Series

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Abstract

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disorder of motor neurons and their axons characterized by signs of upper and lower motor neuron dysfunction. ALS has been associated with exposures in so called electrical occupation. We have found two young patient who developed features of MND following electrocution within 1 year. Due to this great diversity of possible causing agents for ALS, new researches are necessary to elucidate possible etiologies for a better approach to the patients, promoting preventive programs for the disease, optimizing functions and improving the life quality of the patients. A number of studies have demonstrated cognitive performance deficits following electric injury observed cognitive symptoms and neurobehavioral defecit. As treatment approach rehabilitation is important part of treatment as pharmacological part didn't prove strong recovery. As MND causes high level of disability and dependency on caregiver causes gap in rehabilitation due to psychological issue. Neuropalliative rehabilitation is more important in these patient which is unavailable in our country.

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Introduction

Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disease that results in a constellation of problematic symptoms and a high patient caregiver burden.[1] Recognizing these disorders early is necessary in order to initiate specific treatments when available and appropriately counsel patients particularly in the setting of genetic disorders.[2] It is the most common form of motor neuron disease defined by progressive and degenerative compromise of the upper and lower motor neurons,resulting in progressive amyotrophy, fasciculation, paresis, and spasticity.[3-5] Respiratory muscles failure is generally the fatal event, occurring in 1-5 years after the first manifestations of the disease.[6] Its incidence is 1-3 cases/100,000 inhabitants, its prevalence is 3-5/100,000.The combination of asymmetrical weakness and wasting in the limbs associated with clinical evidence of corticospinal tract damage (increased tone, brisk reflexes, extensor plantars) typically comes on insidiously over months and accounts for about 85% of all cases of MND. The disease usually begins either in one limb (foot drop or wasting of the intrinsic hand muscles) or with a combination of bulbar and corticobulbar symptoms (dysphagia, dysarthria, tongue wasting, and a brisk jaw jerk). Clinically detectable sensory involvement should raise the suspicion of an alternative diagnosis such as an inflammatory neuropathy. Fasciculation one of cardinal and isolated symptom can exacerbated by anxiety, caffeine. Extraocular muscles and sphincter are usually spared. Treatment approaches comprises counselling, pharmacological options with rehabilitation program which includes structures individually according to symptom.

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Indoor rehabilitation followed by home based exercise helpful to improve ADL. Particular rehab programe designed according to muscle involvement with or without respiratory involvement. Submaximal strengthening exercise are more preferable and well accepted for avoid fatigue that can reduce functional activity and increases disease progression.

Case Report

First Case

Mr. Abdullah 21 years old, male, normotensive, non diabetic, non asthmatic, right handed, Madrasha student hailing from Madaripur got admitted into BSMMU with the complaints ofWeakness and wasting of left hand and forearm for 4 years following electrocution. weakness intensified in last 3 years and more marked in distal parts. He felt difficulty in performing activities of daily living and compromised hand grip function. On general examination patient is anxious, ill looking, cooperative, Pulse:80beats/min,BP:120/80mmHg and other vital parameters are within normal limit. On left upper limb examination, Wasting of thenar, hypothenar, forearm muscle, dorsal guttering present. Hyperextension of MCP joints, flexion of interphalangeal joint giving an appearance of complete claw hand. A contracture over palmer surface of left 4th MCP due to healed burn injury from electrocution. (Figure 1)

Temperature normal. radial ,brachial and axillary pulsation present, sensory intact. Weakness of all small muscle of left hand present. Pinch grip, hook grip, power grip of left hand is not possible. Ceiling test : positive, Card test : positive. froment test cannot perform due to weakness of both ulnar and median nerve. Other system examination reveals normal findings.Laboratory and radiological Investigations showed normal complete blood counts. CRP in normal range. SGPT, S. Creatinine normal. NCS and EMG showed Motor axonal polyneuropathy of distal portion of left upper extremity. (Table I)

Treatment includes Physical therapy: Neuromuscular Electrical Stimulation Therapy in the form of faradic current over Flexor digitorumsuperficialis and Flexor digitorumprofundus, Flexor pollicislongusand Abductor pollicisbrevis 10 min 2 times daily, Vibration therapy over intrinsic muscle of left hand for 10 min 3 times daily, Warm-moist compression of left hand before exercise 10 min 3 times daily. Therapeutic Exercises Includes: Passive stretching of intrinsic muscles and extensors of hand (5 repetitions 3 times daily), Tendon gliding exercise (2 repetitions 3 times daily), Intrinsic muscles of hand exercise like (Exercise with putty, Ball squeezing exercise), Hand dexterity exercise5 repetitions 3 times daily.(Figure 2)

Table I: NCS and EMG findings

Nerve	Amplitude(mV)	Latency(msec)	NCV(m/sec)
Median motor at wrist	830.00 uV(>4mV)	5.68ms(<4.2msec)	52.3 m/s(>45)
Median motor at elbow	1.21mV(>4mV)	10.46ms(<4.2msec)	52.3(>45)
Ulnar motor at wrist	(>3.5mV)	(<4msec)	(>45m/s)

Second Case

Md. Emon, 16 years, mason in occupation normotensive non-diabetic have one healthy brother and sister came with complaints of weakness of left upper limb for last three months. He has been facingdifficulty in holding object, weight lifting thus the work became complicated.Gradual weakness andwasting of right hand for 10 months following electrocution. The weakness first noticed at thenar and hypothenar eminence of right hand which has been intensified in last 8 months and more marked in distal parts. He felt difficulty in performing his job due to compromised hand grip function. Initially there were no fasciculation but later on after 4 months he noticed fasciculation on his triceps, deltoid and extensor muscles of forearm. On general examination patient is anxious, ill looking, cooperative, Pulse:78 beats/min, BP:110/80mmHg and other vital parameters are within normal limit. On left upper limb examination, Wasting of thenar, hypothenar, dorsal guttering present. Hyperextension of MCP joints, flexion of interphalangeal joint giving an appearance of complete claw hand. A contracture over palmer surface of left 4th MCP due to healed burn injury from electrocution.

Temperature normal. radial ,brachial and axillary pulsation present, sensory intact. Weakness of all small muscle of right hand present. Pinch grip , hook grip , power grip of right hand is not possible. Ceiling test: positive, Card test: positive. froment test cannot perform due to weakness of both ulnar and median nerve, Grip power: 40 on right hand. Other system examination reveals normal findings. Laboratory and radiological Investigations showed normal complete blood counts. ESR, CRP in normal range. SGPT, S. Creatinine normal. NCS and EMG showed Anterior horn cell disease involving left upper limb(Monomelicamyotrophy). Positive Findings were: Increased insertional activity on 1st dorsal interossei, abductor digitize minimi, abductor pollicisbrevis, Extensor indices and over paraspinal muscles at cervical region. (Table II)

Table II: NCS and EMG findings

Nerve	Amplitude(mV)	Latency(msec)	NCV(m/sec)
Median motor at wrist	10.92mVm (>4mV)	4.02ms (<4.2msec)	
Median motor at elbow	11.53mV (>4mV)	8.6ms (<4.2msec)	50.2 (>45m/s)
Ulnar motor at wrist	3.5(>3.5mV)	3.5(<4msec)	
Ulnar Motor at Elbow	1.4	8.18msec	51.3(>45m/s)

Discussion

After electrocution neurological complication may affect CNS and PNS or both. Develop immediately or they can develop later on after the electric injury. Seventeen percent of those who sustained high voltage electric injury experienced delayed neuropathy. Latency period varies from several days to even decades. Mechanism of peripheral injury: Thermal damage, Sympathetic stimulation, Vascular damage, Histological changes, Electrophysiological changes, Direct mechanical trauma. Mechanism of cerebral Ischaemia: Rarely reported, Mechanism poorly understood, Possible mechanism:

vascular endothelial damage,thrombus formation, Other risk factors should be considered: hypertension, smoking, dislipidemea. Multisystem presentation after electrocution: Skin, Cardiac, Respiratory, Vascular, Neurologic, MSK, Renal and other. Among them transient paralysis, peripheral neuropathy, loss of consciousness. Low voltage (<1000 volt), High voltage (>1000 volt), Lightning (10 crore volt). Sources of low and high voltage electricity which may cause injuries can be found in appliances and cables found in the home, office, shops or workplace. However, these are often insulated by non-conducting materials such as plastic or rubber to prevent injuries from occurring. When a current above 10 mA travels through flexor muscles, it causes a sustained contraction. The victim got stuck with the current and increase the severity of injury due to electrocution. When a current above 10 mA travels through extensor muscles, it causes a violent spasm. If the muscles affected are the hip extensors that lengthen the limbs away from the body, the victim may be propelled, sometimes few meter distant from the site of electrocution. Fatalities of Lightening: 0.9 per 10lakh per year, High mortality and significant long term morbidity. Furthermore, there have been observations of changes in the peripheral nerves such as a breaking down of the outermost layer of the nerve bers in the peripheral nervous system as a result of electric injuries and accelerated demyelination and neuronal death in brain tissue of experimental animals exposed to electricshocks, All these have raised the question of whether electricshocks cause neurological diseases, and studieshave linked electric injuries to amyotrophic lateralsclerosis (ALS).[10-12] High quality diagnostic neurophysiology provides essential support to the diagnosis. It is important to appreciate that electrophysiological tests are confirmatory, not diagnostic, and must be interpreted in the context of the clinical syndrome. In typical MND motor nerve conduction velocity is normal (slow conduction suggests a neuropathy), sensory studies are normal, and electromyography reveals diffuse fibrillation and fasciculation.

Two cases were diagnosed according to EI Escorial Criteria.[7] Management includes mobility, postural support, and prevention of contractures; a speech and language therapist to assess swallowing and provide communication aids; an occupational therapist to provide aids to maintain function (wheelchair, mobile arm supports, etc); a dietitian to advise on maintaining weight and percutaneous endoscopic gastrostomy feeding. The only drug licensed for the treatment of MND is riluzole, which was designed as a specific glutamate antagonist. In two large randomised trials of patients with the ALS form of MND, the drug was shown to prolong tracheostomy free survival by 3-6 months.In neuromuscular disease weakness and muscle fiber degeneration may be accelerated by overwork and heavy exercise.[8,9] There also deleterious effect of exercise reporting benefits of both active and resistive exercise in patient without evidence of overwork weakness. In general, however, results of human studies in neuromuscular disease have been mixed. Strength increases have been demonstrated even with high resistance exercise, but benefit was generally greatest in stronger muscles and in patients with slowly progressive disease. A more recent study demonstrated some deterioration in eccentric upper extremity strength with a high resistance protocol. 8° Chart and Sinaki 81 recommend a strengthening program of six maximal isometric contractions, each lasting 6 seconds, to improve and maintain strength, and/or theraband isotonic exercises at a submaximal level. It appears that strengthening exercise programs, at least at a submaximal level of intensity, are probably appropriate and helpful in patients with a slowly progressive course and only in those muscles without marked weakness. Exercise includes: Intrinsic muscle strengthening exercise with putty, glidding exercise. Indoor rehabilitation followed by outdoor follow up and upgrading exercise with prescription of necessary orthosis is better process to rehabilitate completely. Treatment update includes (riluzole, edaravone) and autologous stem cell therapy and antisense oligonucleotide gene therapies are in phase 3 clinical trial.[13] ALS patient follow up done by patient global satisfaction, MRC grading of individual affected muscle by making muscle chart, Functional level by Functional Independent Measure (FIM) score and diagnostic tool NCS and EMG screening specially CMAP. Reduction of CMAP amplitude signify poor prognosis.

As these two patient has recently diagnosed only one follow up done by global score, MRC and FIM. Follow up of case I after 6 months stated that global score was reduced, MRC grading of intrinsic muscles of hand: Palmar Interossei of index and middle finger slightly improved, FIM was initially 98 then it improved to 104, pinch grip and hook grip improved after exercise but patient develop depression as he was madrassa student and study interrupt due to his grip weakness. Follow up of case II after 3 months showed global score improved, MRC grading of APB and 1st lumbricals improved and FIM was 104 then improved 108. None of patient was agreed to do the diagnostic follow up. As indoor rehabilitation is not available that's why it was not possible and we have only physiotherapist. Shortage of occupational therapist and speech therapist also notify. It would be better approach if team work would possible. Low socioeconomic status, improper team work due to lack of therapist, social worker, nutritionist and lack of awareness, lack of indoor rehabilitation facility and lack of neuropalliative care causes hindrance in proper treatment. As long duration disease process affect psychology of both patient and care-giver.

Figure 1

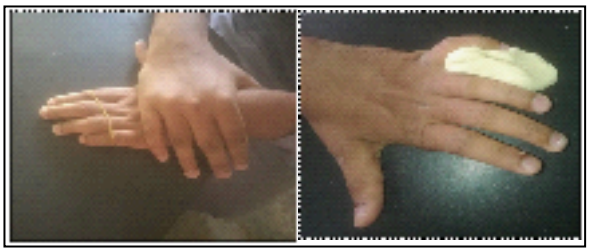

Figure 2

Conclusion

As motor neuron disease are less common at younger age but we found two cases within one year at different institute, atr emendous amount has been learned about the diversity of clinical manifestations of MND and advances in clinical care havesignificantly improved the average life span of individuals suffering from this disease. Though common presentation at older age group young age group also been in consideration. Awareness about safe work place should confirmed. Multidisciplinary team approach is needed for the management of this disease. Early starting of rehab measures can be helpful in improvement of quality of life. Continuation of rehabilitation with the help of caregiver is most important in these cases at later stage. As a longer disease process psychological issue also should keep in mind in case of both patient and caregiver.

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