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Letter to Editor: Open Access

Improvement of the Muscle Strength, Gait, and Functional Parameters with Rehabilitation Program in a Patient with Type-IV Spinal Muscular Atrophy

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A 52-year-old man was seen due to weakness and gait disorder. The patient declared that symptoms started eight years ago and worsened over time. The medical and family history was otherwise unremarkable. Physical examination revealed weakness (4/5) bilaterally of both upper and lower extremity muscles (prominent in the proximal group muscles). He was able to walk with a lofstrand crutch on level surfaces, but not on stairs. Laboratory results showed mildly increased creatine kinase levels. Electro diagnostic tests were consistent with lower motor neuron degeneration (fasciculation, impaired recruitment, high-amplitude, and long duration motor unite potentials with normal nerve conduction velocities). Muscle biopsy revealed a typical denervation pattern. Magnetic resonance

Table 1: Rehabilitation program applied to the patient.

- Non resistant bicycle ergometer (10 minutes)
- Stretching exercises
  - Hip flexor/adductor
  - Knee extensor
  - Ankle plantar flexor muscle
- Active resistive exercises with resistance bands (2 set of 10 repetitions)
  - Upper extremity
    - Shoulder flexors-abductors (1 set of 0.5 kg, increasing gradually to 1 kg)
    - Arm flexors-extansors
    - Wrist flexors-extensors
  - Lower extremity
    - Hipflexors-abductors-adductors
    - Knee flexors-extensors
    - Ankle dorsal-plantar flexors
- EMG-bio feedback (20 min/day)
  - Hipflexor
  - Knee extensor
- Posture exercises at an across a mirror
- 5 day/week,for 4 weeks, supervised by a physical therapist at outpatient clinic

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Table 2: Comparison of patients' values before and after the rehabilitation.

	İsokinetic test								RelationFlex/Ext		Time up&go test (s)	6MWT (m)	MSA	
	Kneeextention				Kneeflexion									
	PT (N.kg^{-1})		FI (%)		PT (N.kg^{-1})		FI (%)						Upperlimb	Lowerlimb
	120°.s^{-1}	240°.s^{-1}	120°.s^{-1}	240°.s^{-1}	120°.s^{-1}	240°.s^{-1}	120°.s^{-1}	240°.s^{-1}						
Beforerehab.	24	20	19	5	4	5	22	78	17	50	16.73	50*	4+/4+	4+/4+
Afterrehab.	24	33	10	37	12	4	48	12	27	12	12.6	185	4+/4+	4+/4+

Abbrevations: 6MWD: Distance walked in the 6-minutes; PT: Peak torque; FI: Fatigue index; MSA: Muscle Strength Assessment.

*: Test was stopped lack of balance.

images of the thoracic spine demonstrated atrophy of the spinal cord. Deletion analysis of the spinal muscular atrophy (SMA) genes was not remarkable. Overall, the patient was diagnosed with SMA type-IV. Rehabilitation program was applied to the patient (Table 1). Muscle strength, gait, and isokinetic parameters did improve with a one-month rehabilitation program including posture, strengthening, and walking exercises (Table 2). In addition knee extensor strength, time up & go test and 6MWT were improved, the patient was able to ambulate independently on level surfaces without the use of any assistive devices, and with a straight cane assist on unlevel surfaces and stairs at the end of the rehabilitation. Home-based exercise programmes and a follow-up visit every 3 months were recommended to the patient.

Spinal muscular atrophies are characterized by the degeneration of anterior horn cells in the spinal cord and motor nuclei in the lower brainstem. SMA is classified into 4 clinical courses [1]. Type-I and type-II SMA (intermediate form) are seen in the neonatal or early childhood periods. Type-I is the most common and severe type. Furthermore, SMA type-III (Kugelberg-Welander disease) is the mild and the least severe form. Adult onset of SMA (SMA type-IV) usually presents in the 2nd or 3rd decades of life. Type IV SMA is quite rare and accounts less than 5% of SMA patients [2,3]. It is inherited in either an autosomal recessive or dominant manner. Type-IV SMA clinically presents with slowly progressive proximal limb weakness and fasciculations with absent/depressed deep tendon reflexes. Adult-onset SMA and type-III SMA patients have normal life spans. Symptoms usually include mild muscle weakness, tremor and twitching. Life expectancy is normal and the muscles for swallowing and breathing are rarely affected. Only a small number of patients eventually require wheelchair assistance [3]. Since the clinical course in SMA is heterogeneous and genetic analysis is not always detected, the diagnosis can be challenging [4]. There is not a specific exercise program for patients with SMA Type IV. The main rehabilitation principles applied in neuromuscular diseases is also valid for SMA type-IV.

Regarding the rehabilitation of type-IV SMA patients in the literature, the data is scarce. Accordingly, herein presenting our unusual case we would like to highlight the role of rehabilitation program on improving functional parameters in SMA patients with benign clinical course. In addition, rehabilitation program should be tailored to the patient's clinical course.

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None

Conflicts of interest

None

References

1. Wang CH, Finkel RS, Bertini ES, Schroth M, Simonds A, et al. (2007) Consensus statement for standard of care in spinal muscular atrophy. J Child Neurol 22: 1027-1049.
2. Striano P, Boccella P, Sarappa C, Striano S (2004) Spinal muscular atrophy and progressive myoclonic epilepsy: one case report and characteristics of the epileptic syndrome. Seizure 13: 582-586.
3. Piepers S, van den Berg LH, Brugman F, Scheffer H, Ruiterkamp-Versteeg M, et al. (2008) A natural history study of late onset spinal muscular atrophy types 3b and 4. J Neurol 255: 1400-1404.
4. Nicole S, Diaz CC, Frugier T, Melki J (2002) Spinal muscular atrophy: recent advances and future prospects. Muscle Nerve 26: 4-13.