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Role of physical therapy in patient with myotonic dystrophy type-2

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Abstract
Myotonic dystrophy type 2 (DM2) is a rare and mild variant of myotonic dystrophy characterised by progressive muscle weakness in predominantly proximal distribution and painful myotonia in limb muscles. Present case report includes the description and physical therapy management plan of a 39 year old male patient with DM2.

Keywords: Myotonic dystrophy, physical therapy, DM2

Introduction
Myotonic dystrophy is an autosomal dominant disorder classified into two types: Type - 1 (DM1) also known as 'Steinert's disease' and type - 2 (DM2) also known as proximal myotonic dystrophy. DM2 is a rare and mild form of myotonic dystrophy with late onset of symptoms. DM2 is characterised by myotonia, proximal muscle weakness and multisystem disorder which includes cardiac conduction defects, cataract, diabetes mellitus type 2, endocrine changes and problems with cognition. Grip myotonia prevents quick release of the grip after a handshake. Myotonia of thenar muscles can be observed by percussing with the hammer. Muscle weakness and wasting predominantly affects proximal muscles which includes neck elbow extension and hip flexors in comparison with early DM1 which initially tends to affect distal upper limb muscles. Muscle pain, stiffness and fatigue are also commonly found in patients with DM2 than in DM1. Typical facial features include long thin face with hollow temples and cheek due to wasting of temporalis and masseter muscles along with drooping eyelids.
As DM2 is extremely rare condition, there is lack of substantial literature and evidence supporting the role of physical therapy in this patients.

Case description
A 39 year old male presented to the physiotherapy OPD with complains of pain in bilateral UE and LE, difficulty in walking and breathlessness. Past history revealed onset of weakness since two years with progressive worsening. Patient also reported admission in ICU 1.5 months ago due to fever and severe breathlessness due to pleural effusion which resulted into unconsciousness. Patient had to be artificially ventilated via tracheostomy tube during his ICU stay for 15 days. ECG on discharge from the hospital was suggestive of nonspecific intraventricular conduction delay and nonspecific T wave abnormality.
Patient belong to a middle class family who owns a grocery store in the city. He lives on the ground floor and has very supportive family members that includes his parents, wife and three children. Patient was found to be cooperative, alert, oriented, attentive and was able to recall past events. No abnormality was detected in cognitive functions.
Present physical examination revealed frontal balding, long thin face with atrophy of temporalis and masseter muscle atrophy and drooping eyelids (fig:1 and fig:2). On motor examination, severe weakness was noted in muscles of neck and face (<3 on MMT). The muscles of shoulder girdle and trunk were found to be moderately weak (3 on MMT) and only mild weakness was evident in other limb muscles. No abnormality was detected in sensory system examination. Patient reported pain score of 4 in bilateral LE which increased to 6 after walking along with feeling of extreme fatigue. Spasm was evident in bilateral hamstring and calf group of muscles. Length testing of the muscles revealed tightness in bilateral hamstring, calf and iliopsoas.

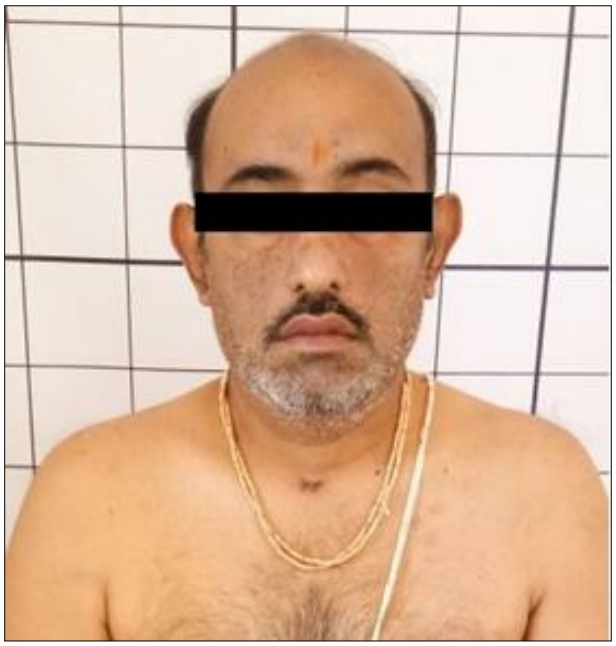

Fig 1: Frontal baiding, atrophy of temporalis and masseter muscles

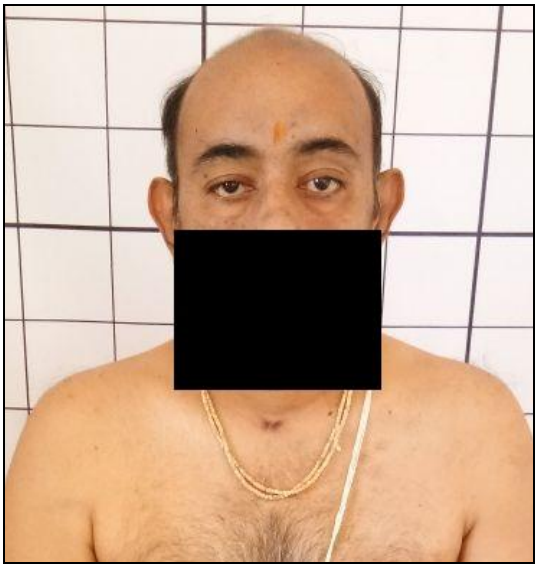

Fig 2: Drooping of eyelids

Patient reported somewhat severe dyspnoea at rest (Modified Borg's scale score 4) and had to take frequent pauses while speaking. His voice was reduced in volume and there was evident nasal twang.

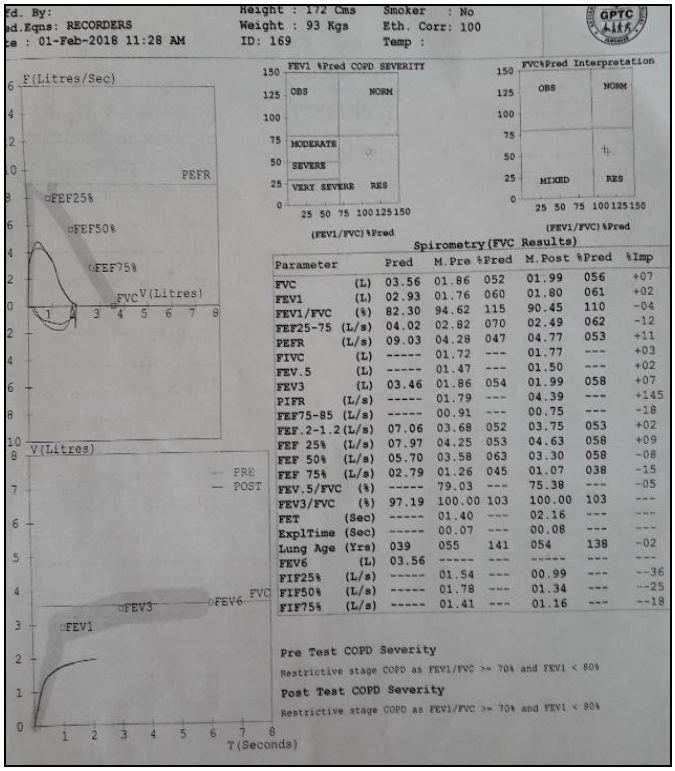

Fig 3: PFT flow loops

Auscultation of lungs revealed reduced air entry in bilateral lower lobes with absence of any adventitious sounds. Patient could not complete 6 min walk test and had to stop after 3 mins due to lower limb pain. Pulmonary function test was performed which revealed moderate restriction (FEV1/FVC >=70%, FVC<80%). MMT of diaphragm revealed muscle strength of grade 4.

Physical therapy management plan

1. To increase the vital capacity-
   a. Inspiratory and expiratory muscle training using incentive spirometer with emphasis on holding at the end. (4-5 repetitions thrice in a day)
   b. Chest mobility exercise in sitting included forward bending, side bending and pectoralis major stretch position with inspiration during flexion and expiration during extension. (5 repetition of each exercise twice a day)
   c. Cycling on a stationary bicycle. (monitor dyspnea, SPO2, heart rate)
2. To relieve pain and spasm in LE
   a. Application of superficial heat. (home advice)
   b. Conventional TENS (20 mins for each LE)
3. To maintain muscle length
   a. Low intensity long duration stretching exercise to hamstring, calf and iliopsoas. (3 repetitions once a day)
4. To prevent muscle atrophy and maintain muscles strength
   a. Active neck and scapular muscles exercise. (three set of 5 repetitions)
   b. Strengthening exercise for shoulder, elbow, wrist muscles using manual resistance. (two sets of 5 repetition)
   c. Partial squats and spot jogging for LE strengthening.

Outcome

Frequent rest periods were required during all the above mentioned exercises due onset of fatigue. BP, HR and SPO2 were measured before and after exercise.

Patient reported increased endurance and reduced episodes of fatigue after 10 days of physical therapy. He also had significant relief of pain in LE. However, no improvement was noted in muscle strength.

Conclusion

As DM2 is a progressive disorder, the aims of physiotherapy management are to prevent complications secondary to immobility, improve and manintain cardiorespiratory capacity, manage pain and fatigue and maintain functional independent status of the patient for as long as possible. It is important to monitor the effect of exercise on cardiorespitory system and fatigue as sudden death due to ventricular tachycardia has been reported in patients with DM2

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