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Treating Fatigue in Patients with Myotonic Dystrophy Using Modafinil: a Case series

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Introduction

Myotonic dystrophy type 1 (DM1) is progressive disease caused by an expansion of unstable CTG trinucleotide repeat in the 3' untranslated region of the DM protein kinase (DMPK) gene, located on chromosome 19q13.3. Daytime sleepiness and fatigue are one of the most frequent non-muscular symptoms among DM1 patients.

We report cases of myotonic dystrophy patients who has been treated fatigue and somnolence with modafinil.

Case 1

A 50-year-old male was referred to our clinic. He had been diagnosed as DM1 by 400 CTG trinucleotide repeats identified in DMPK gene test. The patient complained difficulty in walking, dyspnea, severe fatigue and somnolence. Since his arterial blood gas analysis showed hypercapnia, we applied non-invasive ventilation (NIV), but he couldn't adapt well due to severe obesity (body mass index 40) and sleep disordered breathing. In addition, because of fatigue and somnolence, which especially become worse at early afternoon, he couldn't do his daily life properly. We prescribe 200mg of modafinil at morning, but since the patient reported dizziness, we reduced the dose to 100mg and maintained. The patient was asked to fill out questionnaires named the Schedule of Fatigue and Anergy/General Physician (SOFA/GP) and the Fatigue Severity Scale (FSS) serially, to assess change in degree of perceptive fatigue. The scores SOFA/GP and FSS showed marked reduce by 26 and 56 points before taking the drugs and 13 and 17 points two weeks after taking them, respectively, and the scores remained at a similar level after 6 weeks (Table 1 and 2). The patient reported dramatic improvement of somnolence and fatigue. His compliance on NIV use also improved.

Case 2

A 52-year-old female was visited our clinic. Her DMPK gene test had revealed 430 CTG trinucleotide repeats which implies DM1. She could walk independently despite the limb weakness due to disease progress, but she felt hard to climb the stair up. Fatigue and somnolence, as well as muscle weaknesses, adversely affected her daily life. Since her symptoms became worse in the afternoon, we prescribed 100mg of modafinil at morning.

We asked her to fill out the SOFA/GP and FSS serially. Her initial scores were 27 and 53 points, respectively. Two weeks after treatment, the SOFA/GP and the FSS scores elevated unexpectedly to 29 and 57 points, so we increased dose to 200mg. Along with her symptoms, her SOFA/GP and FSS scores have also slowly dropped to 24 and 37 points, respectively, 6 weeks after taking drug (Table 1 and 2).

Discussion

Modafinil is a newly invented central nervous system stimulant that has been proved to be effective treatment of narcolepsy and hypersomnia, and is usually used for treating fatigue in patients with multiple sclerosis. Many studies have reported that modafinil is also effective in patients with DM1, but only little study had been conducted in Korea. Further studies are needed to confirm the efficacy of modafinil in neuromuscular patients.

Table 1. Changes in the Schedule of Fatigue and Anergy/General Physician (SOFA/GP) scores before and after taking modafinil in each case. The SOFA/GP consists of 10 items which are Likert scale ranging from 1 (none to little) to 4 (most of the time).

Item	Case 1			Case 2		
	Before treatment	After 2 weeks of treatment	After 6 weeks of treatment	Before treatment	After 2 weeks of treatment	After 6 weeks of treatment
1. 신체 활동을 하고 나면 오랫동안 피곤하다	3	1	2	3	4	4
2. 집중력이 부족하다.	3	2	1	4	2	1
3. 신체 활동을 하고 나면 근육에 피로를 느낀다.	3	1	1	4	4	4
4. 두통이 생긴다.	2	1	1	2	2	1
5. 오랜 시간 동안 잠을 자야 한다.	3	1	1	3	4	3
6. 신체활동을 하고 나면 근육에 통증을 느낀다.	3	1	1	3	4	4
7. 깊은 잠을 못잔다.	2	1	1	3	4	3
8. 말하는데 어려움이 있다.	3	2	1	2	1	1
9. 기억력이 좋지 않다.	2	2	1	2	2	1
10. 쉴 때 조차도 근육에 통증을 느낀다.	2	1	1	1	2	2
Total score	26	13	11	27	29	24

Table 2. Changes in the Fatigue Severity Scale (FSS) scores before and after taking modafinil in each case. The FSS consists of 9 items which are Likert scale ranging from 1 (strongly disagree) to 7 (strongly agree).

Item	Case 1			Case 2		
	Before treatment	After 2 weeks of treatment	After 6 weeks of treatment	Before treatment	After 2 weeks of treatment	After 6 weeks of treatment
1. 피로하면 의욕이 없어진다.	6	2	2	7	7	5
2. 운동을 하면 쉽게 피로해진다.	6	2	2	7	7	2
3. 쉽게 피곤해진다.	6	2	2	5	7	2
4. 피로 때문에 신체 활동에 감소된다.	6	2	2	5	7	3
5. 피로로 인해 종종 문제가 생긴다.	6	2	1	5	4	5
6. 피로 때문에 지속적인 신체활동이 어렵다.	7	2	2	5	6	5
7. 피로 때문에 업무에 책임을 다 하지 못한다.	6	2	1	6	6	5
8. 내가 겪고 있는 가장 힘든 문제를 세 가지 뽑는다면 그 중에 피로가 포함된다.	6	2	2	7	7	6
9. 피로 때문에 직장, 가정, 사회 활동에 지장을 받는다.	7	1	1	6	6	4
Total score	56	17	15	53	57	37