Türk Fizyoterapi ve Rehabilitasyon Dergisi

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CASE REPORT

A POSSIBLE RELATIONSHIP BETWEEN RESPIRATORY MUSCLE WEAKNESS AND FAMILIAL MEDITERRANEAN FEVER: A CASE REPORT

ABSTRACT

To Familial Mediterranean Fever (FMF) is an autoinflammatory disorder with genetic origin. Pleuritis is most common in FMF. Long-term sequelae of respiratory system haven't been described in FMF. We documented pulmonary manifestations in patient with FMF. A 61-year-old woman presented with dyspnea, unilateral chest pain, generalized myalgia and FMF. Physical examination was unremarkable. Radiological data showed left-sided pleuritis, minimal pleural effusion. Pleural effusion resolved spontaneously in one week later but patient had still dyspnea. Pulmonary function tests (PFT) was normal. Further examination detected respiratory muscle weakness and decreased functional capacity. Patient then underwent inspiratory muscle training (IMT) for six weeks. After training, inspiratory muscle strength and functional capacity increased. Perception of dyspnea and fatique decreased. In long-term follow-up, frequency of attacks decreased. To our knowledge, there is no study on respiratory muscle weakness and IMT in FMF. Although this report doesn't provide direct evidence, it may provide recommendation for investigation of respiratory muscle weakness and treatment with IMT in FMF with respiratory impairments. Randomized controlled trials are needed.

Key words: Chronic disease, rehabilitation, physiotherapy, familial mediterranean fever

VAKA RAPORU

SOLUNUM KAS ZAYIFLIĞI VE AİLEVİ AKDENİZ ATEŞİ ARASINDAKİ OLASI İLİŞKİ: VAKA RAPORU

ÖZET

Ailevi Akdeniz Ateşi (AAA) genetik kökenli oto-inflamatuar bir hastalıktır. Plörit AAA'da çok sık görülür. Solunum sistemi üzerinde AAA'nin uzun dönem etkileri tanımlanmamıştır. AAA'li bir hastada akciğerdeki etkilenimleri gösterdik. 61 yaşında AAA'li kadın hasta, nefes darlığı, tek taraflı göğüs ağrısı ve yaygın kas ağrısı şikayetiyle başvurdu. Fizik muayenede belirgin bir bulgu yoktu. Radyolojik bulgular sol taraflı plöriti ve minimal plevral efüzyonu gösteriyordu. Plevral efüzyon, bir hafta içinde kendiliğinden çözüldü ama hastanın nefes darlığı şikayeti devam ediyordu. Solunum fonksiyon testi değerleri (SFT) normaldi. Ayrıntılı değerlendirmede solunum kas zayıflığı ve fonksiyonel kapasitede azalma tespit edildi. Daha sonra hastaya altı hafta boyunca inspiratuar kas eğitimi (IMT) uygulandı. Eğitimden sonra, hastanın inspiratuar kas kuvveti ve fonksiyonel kapasitesi arttı. Nefes darlığı ve yorgunluk algısı azaldı. Uzun süreli takipte, atak sıklığı azaldı. Bizim bilgimize göre literatürde, AAA'de solunum kas zayıflığı ve IMT ile ilgili çalışma yok. Bu vaka raporu, doğrudan kanıt olmamasına rağmen, solunum sıkıntısı olan AAA'li hastalarda, solunum kas zayıflığının araştırılması ve tedavide IMT'nin kullanılması yönünde öneriler sağlayabilir. Randomize kontrollü çalışmalara ihtiyaç vardır.

Anahtar kelimeler: Kronik hastalık, rehabilitasyon, fizyoterapi, ailevi akdeniz ateşi

INTRODUCTION

Familial Mediterranean Fever (FMF) is an auto-inflammatory disasese with genetic origin Turks, Arabs, Armenians, Jews, and rarely Greeks are most often affected (1). The disease starts at a young age and manifests itself with recurrent episodes of abdominal or chest pain, arthralgia, and fever (2). Clinical symptoms often lead to hospitalization and unnecessary surgery. Widespread pain result in decreased quality of life during the attacks. Between episodes, the patients appear completely healthy, but their everyday functioning is affected by the spontaneous nature of the attacks (3).

Pleuritic chest pain and fever as manifestations of the first attack are observed in <10% of patients, but approximately 40% have an episode of febrile pleurisy during the course of the disease. Pleural inflammation is one of the three most common manifestations of the disease. The attacks usually last 1–3 days and resolve without specific treatment. There may be radiological evidence of a small exudate in the costophrenic angle and resolves within 48 hours. In many patients with FMF, chest attacks are misdiagnosed as recurrent pneumonia. This is sometimes due to atelectasis that accompanies the pleural inflammation (4).

The recurrent pleuritis can cause adhesion of pleura and impairment in biomechanics of respiratory system. This decreases functional pulmonary capacity. Long-term sequelae of respiratory system haven't been described in FMF. To our knowledge, there is no study on respiratory muscle weakness and IMT in FMF. In this article, we presented pulmonary manifestations and effectiveness of IMT in patient with FMF.

CASE PRESENTATION

A 61-year-old woman presented to our department complaining of dyspnea, unilateral chest pain, generalized myalgia and FMF. These symptoms initially developed at twenty years of age. Colchicine in a dose of 1 mg/day was administered for 35 years but last 5 years she did not used any medication. There was no history of FMF in first-degree relatives. Physical examination was unremarkable. White blood cell count was increased to 11,400/mm3 with 70.4 % neutrophils. C-reactive protein was elevated to 2.1 mg/dL. Radiological data showed left-sided pleuritis, minimal pleural effusion. Pleural effusion resolved spontaneously in one week later but patient had still dyspnea. Assessment and treatment methods;

Pulmonary function tests: Spirometric measurement was performed using a portable spirometry (Spirobank MIR, Italy) according to the guidelines of the American Thoracic Society (5). Forced expiratory volume in 1 s (FEV1), forced vital capacity (FVC), and peak expiratory flow (PEF) were expressed as the percentages of the predicted values (6). Pulmonary function tests (PFT) was normal.

Respiratory muscle strength: Maximal inspiratory pressure (MIP) and maximal expiratory pressure (MEP) were assessed using an electronic pressure transducer (MicroRPM; Micromedical, Kent, UK). The MIP was measured at residual volume, and MEP was measured from total lung capacity, according to Black and Hyatt (7-9). Before treatment MIP and MEP values were 23 cmH2O and 42 cmH2O respectively. These values were under normal values relationship to age and sex.

Functional capacity: Functional exercise capacity was evaluated using a six minute walk test (6MWT). Subject was instructed to walk as far as possible in six minutes in an enclosed 30 m long hospital corridor. Patient was instructed to walk her own pace but to cover as much meter as possible within 6 min. Each minute standardized encouragement was given to the patient. Patient was allowed to stop and rest during test, but was instructed to go on walking as soon as she was able to do so (10). The maximum distance covered at the end of the test was recorded. Breathlessness and fatigue perception were determined using a 10 point modified Borg Scale during the walking test. Heart rate, respiratory rate, blood pressure and oxygen saturation were measured before and after the test. Maximum heart rate value achieved during the test was recorded (11). The 6MWT was repeated two times. Patient was rested 30 min between the two tests, and the highest distance was recorded. Distance of 6 MWT was 256 m before treatment. Before the test at rest, the results of Perception of Dyspnea (Modified Borg Scale 0-10) and Perception of Fatique (Modified Borg Scale 0-10) value were 2,1

Table 1. Results of measurements

		Before Treatment	After Treatment
PFT	FVC (%)	94	95
	FEV1 (%)	91	91
Functional Tests	6MWT Distance (m)	256	283
	Perception of Dyspnea (Modified Borg Scale 0-10) (cm)	7	2
	Perception of Fatique (Modified Borg Scale 0-10) (cm)	5	2
Dyspnea	Modified Medical Research Council Dyspnea Score (0-4)	3	1
Respiratory Muscle Strength	MIP (cmH2O)	23	47
	MEP (cmH2O)	42	45

Abbreviations: PFT; pulmonary function test, FVC; forced vital capacity, FEV1; forced expiratory volume in one second, 6MWT; six minute walk test, MIP; maximum inspiratory pressure, MEP; maximum expiratory pressur

respectively. After walking, values were 7 and 5 respectively. Results of Perception of Dyspnea (Modified Borg Scale 0-10) and Perception of Fatique (Modified Borg Scale 0-10) value were 7,5 respectively before treatment. The results of test were shown in table 1.

Dyspnea: Modified Medical Research Council (MMRC) dyspnea scale was used to evaluate dyspnea severity during activity. Levels of dyspnea are graded 0 (absence of dyspnea during strenuous exercise), to 4 (dyspnea during daily activities) (12). Before training score of MMRC was 3.

Inspiratory muscle training protocol: Before IMT, patient was taken in one-week familiarization period and instructed to learn breathing adequately. After, training workload was adjusted to lower loads (20-30% of MIP) and patient was instructed to maintain adequate inspiration and expiration while using Threshold IMT device (Respironics, USA). As soon as the patient managed to maintain adequate workload, IMT was started. Patient received IMT at 40% of MIP, and training load was adjusted to maintain 40% of the MIP, weekly. MIP was measured at supervised session each week, and 40% of measured MIP value was the new training workload. Patient trained for 30 min-per day, 7 days per week, for 6 weeks. Once a day at each week, patient's heart rate, blood pressure and breathing frequency were monitorized during the IMT sessions and new workload was adjusted. Six sessions were performed at home and 1session was supervised at the rehabilitation department. In total, 8 sessions were supervised (2 sessions in familiarization period, and 1 session at each week during 6 weeks). Patients were instructed to maintain diaphragmatic breathing, and try to maintain 10-15 breaths and rested 5-10 s between breaths. As soon as the patient managed; they were encouraged to maintain 25-30 breaths at each workload. Patient wore nose-clip during all sessions. IMT diary was checked weekly. Patient was checked by phone calls two times a week, whether she is doing IMT in appropriate manner. Total minute spent during training period was calculated based on her reports written on diary. Patient was told not to exercise or do physical activity over her normal routine during the training period.

After training, inspiratory muscle strength and functional capacity increased. Perception of dyspnea and fatique decreased. In long-term follow-up, frequency of attacks decreased. Results of measurements were shown in Table 1.

Discussion

Among the pulmonary manifestations of FMF, pleuritis is the most common, and occurred in about 45% of the patients described in a clinical study by the Tel-Hashomer group (4,13). Tunca and et al. were found the pleuritis prevalence 31.2% in their study on 2838 patients with FMF (14). Also Livneh and Langevitz described the diagnostic criteria of FMF. According to these criteria, unilateral pleuritis is one of the major criteria of FMF (15). "Chest attacks" are characterized by unilateral pleuritic

chest pain that increases on inspiration, shortness of breath, and rapid, shallow breathing. A typical attack lasts between 1 and 4 days, similar to attacks in other locations. Physical examination and chest radiography are usually unrevealing, although occasionally the costophrenic angle may be blunted on the side of the attack and the recurrent attacks may sometimes result in pleural thickening and adhesions (16). In the present case's history, she reported pleuritic chest pain during all attacks. Also the patient presented here has appropriate ethnic origin because FMF is prevalent among the Turkish population.

According to our knowledge, there is no study about the physiotherapy in FMF. The etiology of recurrent attacks of serositis in familial Mediterranean fever (FMF) is not completely understood. Several uncontrolled clinical series have reported that strenous physical activity; stressful events; feelings of anger, anxiety, excitement, and grief; a high-fat-content diet; and menstrual periods may trigger FMF attacks (17-18)8. However Yenokyan et al. did not find significant differences in the risk of having FMF attacks between the periods of high physical exertion compared with low physical activity (19). So for the modification of the effect of the triggers on the risk of FMF attacks, physiotherapy could be an alternative approach. Treatment with 1–2 mg of colchicine a day has been shown to prevent or alleviate the attacks in the majority of patients (20,21). In the presence of colchicine, low and moderate- intensity exercise, relaxation exercises, breathing exercises for the pulmonary manifestations of FMF and relaxation could be used.

In literature, it has been suggested that an imbalance between muscle energy production and utilization (22). Also, according to our baseline data, our case avoided heavy physical activity because of her disease. The patient presented here has low functional capacity and respiratory muscle weakness. Impaired calcium metabolism, energy production in muscle and inactivity may underlie these results. Our case had normal pulmonary function testing. But in literature, patients with FMF were found to have mild restrictive lung impairment on pulmonary function testing. One explanation for this phenomenon could be the occurrence of occult parenchymal injury during FMF attacks that could not be demonstrated by x-ray studies of the chest (4,23).

The findings of the joint and abdominal pain are most frequent than pleuritis in FMF. Therefore respiratory impairment related with pleuritis is ignored by the clinicians. The measurement of respiratory muscle strength to detect inflammatory changes could add useful information for evaluating thoracic symptoms in FMF patients. Also treatment of respiratory muscle weakness with IMT may cause improvement in functional capacity of patients with FMF. Number of hospital admissions due to FMF attacks was recorded according to hospital records. Before IMT treatment, she had three attacks in one year. But after IMT treatment, she had only one attack during the two-year follow-up, the frequency of attacks decreased. The modification of the effect of the triggers such as stress on the risk of FMF attacks with IMT may underline this result.

This case showed that FMF may cause respiratory muscle weakness and IMT may improve respiratory capacity and function. Therefore, assessment of respiratory impairment and IMT should be in treatment options for recurrent attacks of pleuritis in FMF patients. Although this report doesn't provide direct evidence, it may provide recommendation for investigation of respiratory muscle weakness and treatment with IMT in FMF with recurrent chest attacks. Randomized controlled trials are needed.

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