Journal of Experimental and Clinical Medicine
https://dergipark.org.tr/omujecm

Case Report

J. Exp. Clin. Med., 2018; 35(4): 111-113
doi: 10.5835/jecm.omu.35.04.004

Pericardial tamponade as the initial sign of systemic lupus erythematosus within Myasthenia Gravis patient

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ARTICLE INFO

Article History
Received 28 / 12 / 2017
Accepted 10 / 01 / 2018
Online Published Date 18 / 11 / 2019

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Keywords:
Myasthenia gravis
Pericardial tamponade
Pleural effusion
Systemic lupus erythematosus

ABSTRACT

Systemic lupus erythematosus (SLE) is an autoimmune connective tissue disorder, mediated by numerous auto antibodies that has many different clinical manifestations and frequently affects the cardiac system. Pericarditis and pericardial effusion are well recognised cardiac complications. SLE is rarely reported with Myasthenia Gravis (MG). Myasthenia gravis (MG) is characterized by the dysfunction of neuromuscular junctions mediated the antibodies against the acetylcholine receptor and presents with weakness, fatigability of skeletal muscle. Thymectomy is a therapeutic option for patients with severe MG. Here we report a case, presented with dyspnea and revealed pericardial tamponade, pleural effusion who was diagnosed with SLE, three years after thymectomy performed for MG. Pericardial tamponade and pleural effusion are the first sign of SLE very rare and thymectomy facilitated the development of SLE.

1. Introduction

Systemic lupus erythematosus (SLE) (non-tissue specific) and Myasthenia gravis (MG) (tissue-specific) are autoimmune diseases that genetic, environmental, and immunological factors have been implicated (Omar et al., 2010; Miskovic et al., 2015). The literature describes cases of SLE after thymectomy for myasthenia gravis, but pathophysiological mechanisms that lead to this phenomenon are not clear (Park et al., 2004). SLE is a connective tissue disorder, mediated by numerous autoantibodies that has many different clinical presentations and often affects the cardiovascular system (Park et al., 2004; Miskovic et al, 2015). Pericarditis and pericardial effusion are well described cardiac complications and pericardial effusions seen in up to 50% of patients with SLE (Omar et al., 2010; Castrejón et al., 2011; Kumar et al., 2012). But, cardiac tamponade has rarely described as an initial presenting feature of SLE (Topaloglu et al., 2006; Kumar et al., 2012). MG is decribed as dysfunction of neuromuscular junctions mediated the antibodies against the acetylcholine receptor. It presents with weakness and fatigability of skeletal muscle. Thymectomy is a therapeutic option for patients with severe MG (Park et al., 2004; Omar et al., 2010; Castrejón et al., 2011).

Here we report a case, presented with dispnea and revealed pericardial tamponade, pleural effusion who

was diagnosed with SLE, three years after thymectomy performed for MG. We found coexisting the pericardial tamponade and pleural effusion as the first clinical sign of SLE.

2. Case

A 35-year-old female patient presented with fatigue, fever, rashes, increased dyspnea symptoms. On physical examination showed a systolic blood pressure of 90 mmHg, a diastolic blood pressure of 60 mmHg, and a heart rate of 115/bpm. Heart sounds were deep and respiratory voices couldn't be obtained on the lung bases. Electrocardiography showed tachycardia, low voltage and electrical alternation (Fig. 1a). Bilateral pleural effusion and large pericardial effusion were observed in thorax tomography (Fig. 1b). Echocardiography revealed pericardial tamponade findings (Fig. 1c). Pericardial effusion was evacuated with pericardiocentesis. Pericardial fluid examination described as transudate and ibuprofen and colchicine therapy started. Patient's diuresis increased and dyspnea improved on follow-up. We learned from patient's history that the patient had a thymectomy operation three years ago with the diagnosis of MG and followed asymptomatic without medication. Laboratory analysis showed high rheumatoid factor, antinuclear antibody (ANA) 1/100 dilution +++ (1/1000), anti-ds-DNA 74.1 IU/ml (>22 IU/ml), anti-cardiolipin IgG 36.5 (> 10 U/ml), sedimentation 84 mm/h, CRP 22 mg/dl, anti-beta 2 glycoprotein-1 screening (IgA,G,M) 18.5 U/ml (>10 U/ml), p-ANCA/c-ANCA (qualitative) 1/10 dilution +++ (1/100) positive. The patient was consulted the rheumatology department. Treatment of methyl prednisolone, azathioprine, hydroxychloroquine were started with diagnosis of SLE. The patient defined hoarseness, so the pyridostigmine therapy was initiated with a diagnosis of myasthenia gravis by neurology. There was no pericardial fluid in control echocardiography. The patient's symptoms regressed and directed to the neurology and rheumatology departments. Pericardial effusion didn't repeat under one year follow-up.

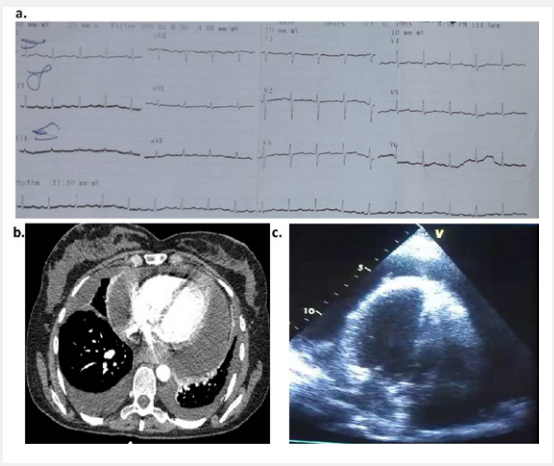

Fig. 1. Pericardial tamponade findings in different imaging modalities Fig. 1a: Electrocardiographic imaging of electrical alternation and voltage drop Fig. 1b: Thoracic tomographic imaging of pericardial tamponade and pleural effusion Fig. 1c: Echocardiographic imaging of pericardial tamponade.

3. Discussion

Pericardial tamponade as the first initial presentation of SLE is very rare and only limited to case reports (Topaloglu et al., 2006; Kumar et al., 2012). We describe a case of coexisting the pericardial tamponade and pleural effusion as the first presentation of SLE. SLE and MG are autoimmune diseases that share certain similarities (Omar et al., 2010). They happen more often in young women, have relapsing remitting clinical progression and usually positive anti-nuclear antibodies (Omar et al., 2010; Castrejón et al., 2011). But they are two different clinical syndromes and one autoimmune disease are increased risk of developing the second one. The entity of second autoimmune disorder is 13–22 % in patients with MG (Omar et al., 2010; Miskovic et al., 2015). Thymectomy is a therapeutic option for patients with severe MG. It can achieve complete clinical remission in 80% of patients (Castrejón et al., 2011; Miskovic et al., 2015). But many SLE cases described in the literature after thymectomy (Omar et al., 2010; Castrejón et al., 2011; Miskovic et al., 2015). So thymectomy operation predisposes patients to SLE and pathophysiological mechanisms are not clear. Thymectomy operation can cause to disproportion of auto-reactive and regulatory T cells and to induction of auto-immune processes due to the loss of central tolerance and over production of auto-antibodies (Omar et al., 2010; Castrejón et al., 2011; Miskovic et al., 2015). So after thymectomy, these patients should follow up intermittent. In the literature, polyarthritis and polyarthralgia are the most common clinical presentations of SLE after thymectomy operation (Park et al., 2004; Miskovic et al., 2015). In our patient, the first sign of SLE was pericardial tamponade and pleural effusion. Although thymectomy is an effective treatment modality in MG patients, clinical cases and literature datas support that this surgical operation may be a precipitating factor for other autoimmune diseases, especially SLE (Park et al., 2004; Omar et al., 2010; Castrejón et al., 2011). Some MG patients who undergo thymectomy, SLE can thrive over the years. The probability of secondary autoimmune pathologies should be highly noted after thymectomy operation (Park et al., 2004; Omar et al., 2010; Castrejón et al., 2011). In our case report, SLE was described as three years after thymectomy operation that was performed for MG and by which long-term improvement of MG was achieved. Also, coexisting of pericardial tamponade and pleural effusion as initial clinical SLE signs is very rare in the literature.

Conclusion

MG patients should be careful about the development of different autoimmune diseases especially SLE after thymectomy operation. So, these patients should follow up intermittent. Coexisting the pericardial tamponade and pleural effusion may rarely be the first sign of SLE, as seen in our case.

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