



# Dermatomyositis Associated With Breast Cancer: A Case Report and Review of the Literature

## Meme Kanseri ile ilişkili Dermatomiyozit, Bir Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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### Summary

Dermatomyositis (DM) is a connective tissue disease characterized by specific inflammatory lesions in muscle biopsy. An increased risk of underlying malignancy has been found in patients with DM. The common cancers associated with DM include ovarian cancer, lung cancer, pancreatic cancer, melanoma, colon cancer, non-Hodgkin's lymphoma, stomach cancer and breast cancer. The association between DM and breast cancer has not been fully evaluated. We present the case of a 58-year-old female patient with DM diagnosed to be suffering from breast cancer. A literature review revealed that the association of dermatomyositis with breast cancer is rare and consistent with poor prognosis. *Turk J Phys Med Rehab 2012;58:246-8.*

**Key Words:** Dermatomyositis; breast cancer

### Özet

Dermatomiyozit (DM) kas biopsisinde spesifik inflamatuvar lezyonlarla karakterize bir kollajen doku hastalığıdır. Dermatomiyozitli hastalarda altta yatan artmış bir malignensi riski bulunmaktadır. DM ile ilişkili yaygın kanserler over, akciğer, pankreas maligniteleri, melanoma, kolon kanseri, non-Hodgkin lenfoma, mide ve meme kanserleridir. Meme kanseri ve DM arasındaki ilişki yeterince değerlendirilmemiştir. Biz 58 yaşında DM ve meme kanseri olan bir olguyu sunmayı amaçladık. Literatürün gözden geçirilmesi DM ve meme kanseri ilişkisinin nadir olduğunu ve kötü prognoz sergilediğini göstermektedir. *Türk Fiz Tıp Rehab Derg 2012;58:246-8.*

**Anahtar Kelimeler:** Dermatomiyozit; meme kanseri

### Introduction

The idiopathic inflammatory myopathies are defined by the combination of proximal muscle weakness, increased serum concentrations of enzymes derived from skeletal muscle, myopathic changes on electromyography and inflammation of the muscles (1).

Polimiyozitis (PM) is the prototype of idiopathic inflammatory myopathies. Dermatomyositis (DM) is differentiated clinically from PM by the presence of a rash. It is also characterized by perivascular infiltration of helper T cell and B cell lymphocytes (1).

The idiopathic inflammatory myopathies are relatively rare diseases, and the incidence of dermatomyositis is estimated at 1-10 cases per million in adults (2). The incidence appears to be

increasing, although this may simply reflect increased awareness and more accurate diagnosis. The autoimmune mechanism of the disease is not fully recognized. Several lines of evidence showed the link between DM and neoplastic disease (2). Compared with the general population, patients with DM are at a 10-fold increased risk for cancers (3). There is a well-recognized association between DM and several cancers such as ovarian, lung, pancreatic, melanoma, colon cancer, non-Hodgkin's lymphoma, stomach cancer, and breast cancer (4,5).

The association of DM with malignancy was first reported by Stertz in 1916. He described a patient with proximal muscle weakness, eyelid changes muscle biopsy evidence of myositis as well as coexisting gastric carcinoma. In the same year, Kankeleit described a patient with DM and breast cancer (2,6).

In the present study, we report the case of a 58-year-old female patient with DM and breast cancer, and review the relevant literature.

## Case Report

A 58-year-old female was admitted to our hospital on July 20, 2009. She was hospitalized with fever, fatigue, morning stiffness, weight loss, anorexia, and arthralgia. Her complaints started at the age of 56 years. She complained of symmetric proximal muscle weakness in the shoulder and pelvic girdle and, she also had neck flexor muscle weakness. The patient presented with arthralgia, mild muscle pain and tenderness in the lower extremities. The clinical examination of this patient showed heliotrope rash of the eyelids and erythema on the extensor surface of the finger joints.

Laboratory investigations revealed white cell count  $4.2 \times 10^3$ , hemoglobin 13 g/dl, hematocrit 41.5%, platelets  $350 \times 10^3$ , Erythrocyte Sedimentation Rate (ESR) 45 mm/h, C-Reactive Protein (CRP) 20 mg/dl. Renal (BUN, creatinine) and liver function (AST, ALT) tests were normal. Serological examinations showed normal Rheumatoid Factor (RF) level, negative results for Antinuclear Antibody (ANA), anti-ds DNA, anti-CCP, anti-Jo-1, anti-mi-2, c-ANCA antibody. The patient's urinalysis showed no microscopic hematuria and red blood cell casts in the urinary sediment. A muscle biopsy was performed and the results of histological examination were consistent with inflammatory myositis. Serum Creatine Kinase (CK) and other muscle enzyme concentrations were normal. Electromyography (EMG) demonstrated myopathic changes. The results of Magnetic Resonance Imaging (MRI) were consistent with muscle inflammation.

We diagnosed our patient as having DM, based on the clinical symptoms, pathological changes on EMG, and histological findings of the muscle, and the results of MRI. In addition, breast cancer was detected with a control mammography. The histological type of breast cancer was adenocarcinoma.

## Discussion

The idiopathic inflammatory myopathies are a heterogeneous group of conditions characterized by symmetric proximal muscle weakness and non-suppurative inflammation of the

skeletal muscles. These patients have increased serum concentrations of enzymes derived from the skeletal muscles, myopathic changes demonstrated by EMG and inflammatory changes in muscle identified by MRI. Some patients have circulating myositis-specific autoantibodies (1). Patients with DM fulfill the criteria for PM and also have cutaneous involvement (7). Rash may be the presenting complaint, antedating the onset of weakness by more than a year. Gottron's papules are considered pathognomonic (1,7).

Laboratory investigations reveal increased serum enzyme concentrations of the skeletal muscles. These include CK, aldolase, serum glutamic-oxaloacetic transaminase, serum glutamic-pyruvate transaminase, and lactate dehydrogenase. Increased concentrations of these enzymes are found in inflammatory muscle diseases, but are not specific for the diagnoses. The concentrations of CK and other muscle enzymes are increased at some time during the course of the disease, and in most patients, this is a helpful indication of the disease. Normal concentrations of CK may be found very early in the disease course (1).

EMG demonstrates myopathic changes that are consistent with inflammation. However, these findings are not specific for DM. EMG changes may be limited or localized, and may be completely normal in 10-15% of patients (8). The muscle histopathology of adult DM is characteristic. Skin histopathology varies according to the stage of the disease and the type of lesion. Inflammation may also be apparent on fat-suppressed T2-weighted MRI short time inversion recovery sequences (1).

Our patient presented with fever, fatigue, morning stiffness, weight loss, anorexia, and arthralgia. She had complaints of symmetric proximal muscle weakness in the shoulder and pelvic girdle. Besides, she had neck flexor muscle weakness as well as mild muscle pain and tenderness. The clinical examination showed heliotrope rash of the eyelids and erythema on the extensor surface of the finger joints.

Systemic inflammatory signs, such as serum CRP concentrations and ESR, were increased. Serum CK and other muscle enzyme concentrations were within normal ranges. EMG demonstrated myopathic changes (polyphasic motor unit potentials with spontaneous fibrillation potentials). Muscle biopsy revealed pathologic findings consistent with inflammatory myositis. The results of MRI were consistent with muscle inflammation.

In our patient, DM was diagnosed according to the "revised criteria for the diagnosis of idiopathic inflammatory myositis", proposed by Bohan and Peters (1). They suggested a set of criteria to aid in the diagnosis and classification of DM and PM. Four of the 5 criteria are related to the muscle disease as follows: progressive proximal symmetrical weakness, elevated levels of muscle enzymes, an abnormal finding on electromyography, and an abnormal finding on muscle biopsy. The fifth criterion was compatible with cutaneous disease. In addition to DM, the diagnosis of breast cancer was obtained with a control mammography. The histological type of breast cancer was adenocarcinoma, as Andras et al. (9) obtained similar consequences in a 21-year retrospective study.

The association between DM and various types of malignancies has been reported in several studies, with an estimated frequency of about 20-25%. Cancer may appear before the onset of DM, concurrently with DM or after its onset (10). The incidence of cancer is higher in older DM patients and myositis associated with malignancy is more common in patients over 50 years of age. Women are affected more frequently than men, by a 2:1 ratio. Female predominance is especially great between ages 15 and 45 years (1). Despite the small series, the incidence of cancer in patients with DM (25%) is similar to that observed in larger series (15%-30%) (11).

Although breast cancer is the most common malignancy, coexistence of DM with breast cancer is a rare phenomenon. The onset of DM may precede, coincide with, or follow the diagnosis of breast cancer (12). In a recent case series, it has been indicated that malignant breast tumor screening is indicated in women with DM, especially in those aged older than 50 years. (13). Also in another relatively large case series including patients with DM and breast cancer, it has been shown that sudden occurrence of DM later in life, rapid course, unusual and more severe clinical presentation could suggest a paraneoplastic course of DM (14). As the association between DM and breast cancer has not been fully evaluated, in a population-based case-control study, Gadalla et al. (15) suggested that systemic inflammation may affect breast epithelial neoplasia. In addition to the weakness and fatigue that occur in patients with PM or DM and a coexisting malignancy, the symptoms can develop in the progress of a neoplastic disease as the result of the systemic effects of cytokines released by the tumor cells. The proximal myopathy that develops in patients with carcinoid syndrome is probable to the result of compounds produced by the cancer cells. Neuromuscular changes can develop as features of paraneoplastic syndrome (1). The course of DM in such patients usually correlates closely with the activity of the underlying malignancy (16).

In conclusion, we present a case presenting with DM and diagnosed as suffering from breast cancer. A literature review revealed that the association of DM with breast cancer is rare and consistent with a poor prognosis.

#### Conflict of Interest:

Authors reported no conflicts of interest.

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