The Comorbidity of Morphea, Raynaud's Phenomenon and Sarcoidosis: Case Report

Morfea, Raynaud Fenomeni ve Sarkoidoz Birlikteliği

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ABSTRACT Sarcoidosis is an autoimmune disorder of unknown etiology which is characterized by non caseating granulomas and can be seen with other autoimmune diseases. Morphea is an autoimmune connective tissue disorder which occurs because of over production of the dermal collagen. Raynaud's phenomenon is an episodic self-limited vasomotor disturbance manifested as colour change in the fingers. In this case we reported a patient who had chest pain, dyspnea, weight loss and colour change in her fingers when she was exposed to cold. She was diagnosed as sarcoidosis. She had also two morphea plaques on her abdominal skin. We especially preferred to report this patient on account of the fact that sarcoidosis cases with morphea and Raynoud's phenomenon have never been reported.

Key Words: Sarcoidosis; scleroderma, localized; Raynaud disease; autoimmune diseases

ÖZET Sarkoidoz, non-kazeifiye granülomlar ile karakterize, etyolojisi bilinmeyen multisistemik otoimmun bir hastalıktır ve diğer otoimmun hastalıklarla birlikte görülebilir. Morfea, dermal kollajen üretiminin artmasından kaynaklanan otoimmun bir konnektif doku hastalığıdır. Raynaud fenomeni parmaklarda renk değişikliği ile ortaya çıkan ve kendi kendini sınırlayan epizodik bir vazomotor bozukluktur. Biz burada göğüs ağrısı, nefes darlığı, kilo kaybı ve soğuğa maruz kalınca parmaklarında renk değişikliği şikayetleri ile başvuran ve sarkoidoz tanısı koyduğumuz bir hastayı sunduk. Hastanın aynı zamanda karın bölgesinde iki adet morfea plağı vardı. Bu vaka, literatürde sarkoidoz, morfea ve Raynaud fenomeninin birlikte olduğu hiçbir vaka bildirilmemiş olması nedeniyle sunulmuştur.

Anahtar Kelimeler: Sarkoidoz; skleroderma, lokalize; Raynaud hastalığı; otoimmün hastalıklar

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Sarcoidosis is a multi-systemic disease which is characterized by non-caseating granulomas and usually affected lungs and intrathoracic lymph nodes.[1] Sarcoidosis may present with three different clinical manifestations; asymptomatic sarcoidosis, sarcoidosis with non-specific symptoms and sarcoidosis with symptoms associated with specific organ involvement. Approximately 30-50% of patients are asymptomatic at the time of diagnosis and usually detected with routine chest radiography.[2] Sarcoidosis, which is not clear whether it is an autoimmune disease or not, can be accompanied with autoimmune diseases.

Morphea is a connective tissue disorder which is characterised by fibrosis of skin and subcutaneous tissues. The most common type is plaque type morphea which can occur with other autoimmune diseases concomitantly.[3,4] Raynaud's phenomenon (RP), which is a disorder characterized by colour changes of finger skin, frequently associated with autoimmune connective tissue disorder.[5,6]

In this case presentation, we report a patient diagnosed with sarcoidosis, morphea and RP because a case with these three diseases is not available in the literature.

CASE REPORT

A 54 year old woman, presented to our outpatient clinic with fatigue, low back pain, chest pain and dyspnea. Although patient had been having these symptoms for four years, her symptoms progressed in last six months. The patient stated having purple colour change of digits when she was exposed to cold. The patient lost 23 kg in last two months. She also complained about night sweats accompanied by fever.

The patient's past medical and surgical history were insignificant except hypertension and hysterectomy due to uterine myoma. She stated that she has been taking an anti-hypertensive medication. She denied smoking and consuming alcoholic drinks.

In physical examination, the patient had RP with cold water test. There were two morphea skin lesions in size of 3x4 cm on abdomen (Figure 1). The examination of pulmonary, cardiovascular and the other organ systems were unremarkable. There was no evidence of infection or inflammation in the laboratory results. The renal and liver function tests, serum calcium and phosphorus levels, and viral markers were normal. Serum angiotensin converting enzyme (ACE) level, anti-nuclear antibody level, epithelial neutrophil activating protein level were within normal range. Urine calcium level within 24 hours was also normal. The patient's blood count was normal except for slightly increased platelet count.

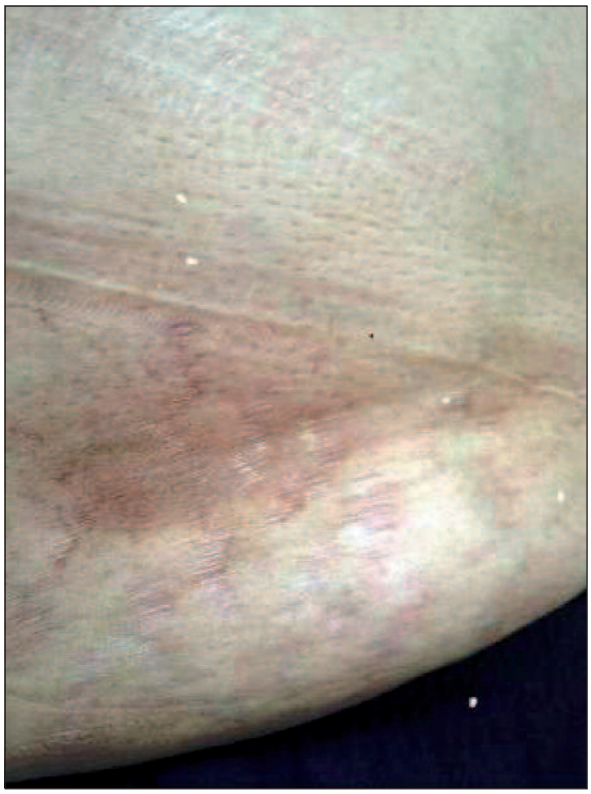

FIGURE 1: Morphea plaque.
(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

The chest radiography demonstrated bilateral mediastinal involvement. High-resolution computerized tomography (HRCT) of the chest revealed bilateral mediastinal and hilar lymphadenopathy, pleuroparenchimal abnormalities and non-calcified pulmonary nodules (Figure 2).

The patient underwent lymph node and lung biopsy. Pathologic findings of the biopsies were correlated with non-caseating granulomas. Tissue PCR studies were negative for tuberculosis. Also patient's skin lesions' biopsy was resulted as morphea (Figure 3).

Abdominal ultrasonography demonstrated hepatosteatosis and cholelithiasis. Patient's electrocardiogram and transthoracic echocardiography were found to be normal.

According to pathologic and clinical findings, patient was diagnosed with sarcoidosis. The patient was started on prednisone 10 mg/daily. Her symptoms and skin lesions improved with treatment.

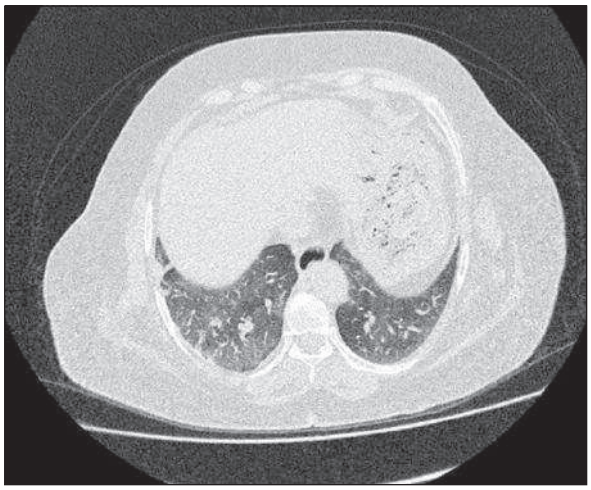

FIGURE 2: Pulmonary High-Resolution CT scan shows ground-glass attenuation in the lung parenchyma.

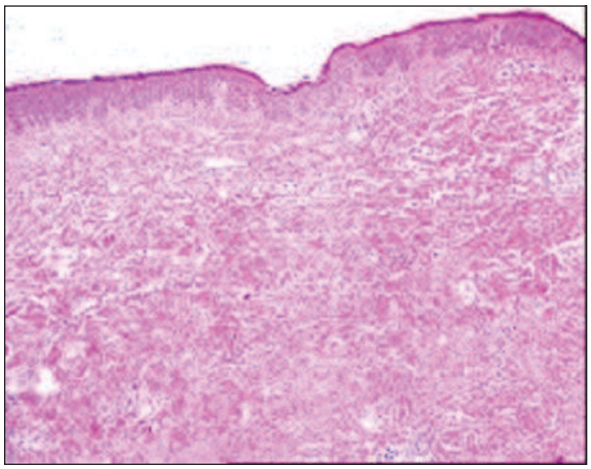

FIGURE 3: Morphea pathology image (HEx100) : Coarsening and flattening in the structure of collagen and infiltration of hyperchromatic nucleus containing tumor cells in the dermis.
(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

Oral and written consent was obtained from the patient.

DISCUSSION

Sarcoidosis thought to occur as a result of genetic predisposition and environmental factors is a disease of unknown cause. Sarcoidosis is defined as an exaggerated immune response against some unidentified antigens.[7] Despite there are some factors indicates that sarcoidosis has common etiology with autoimmune diseases, it is still unknown whether sarcoidosis is an autoimmune disease or not.[8]

In the literature, some cases have been reported that coexistence of sarcoidosis with various autoimmune diseases. In the majority of these cases, sarcoidosis was associated with generalized scleroderma, but only two case of localized scleroderma.[9,10]

Our patient had two morphea plaques in lower abdomen. The pathologic findings of these plaques weren't related with sarcoidosis. Thus, the patient was evaluated as a sarcoidosis case accompanied by morphea.

RP can be seen with other diseases such as autoimmune collagen tissue diseases. Although, RP is most commonly seen with scleroderma, it is not known whether it is accompanied by morphea. In addition there is only one case report of RP accompanied by sarcoidosis in the literature.[11]

Sarcoidosis and morphea are two diseases in the etiology of which immune mechanisms play an important role. RP also is a condition which is often accompanied by autoimmune disorders. We reported this patient because she has these three diseases. The fact that these three diseases are concurrently seen makes us to think that the same common mechanisms may be responsible for their etiology.

Laboratory findings often seen in sarcoidosis are lymphocytopenia, mild eosinophilia, increased sedimentation rate, hyperglobulinemia, hypercalcemia and hypercalciuria. More than half of the patients have high levels of ACE and it is found to be correlated with the disease activity. Hyperglobulinemia and increased sedimentation rate might be seen in acute illness.[12] However, in our patient, laboratory results were unremarkable except a slight increase in platelet count.

Sarcoidosis can be asymptomatic or presented with many different symptoms.[2] Therefore, the diagnosis can be delayed. Our patient had chest pain, dyspnea and severe back pain for four year. The patient's chest radiography showed bilateral hilar lymphadenopathy, and HRCT of the chest demonstrated bilateral mediastinal lymphadenopathy and pulmonary parenchymal lesions.

Sarcoidosis should be considered in patients presenting with non-specific symptoms and it should also be kept in mind that it may be accompanied by other autoimmune diseases.

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