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A Case of Non Paraneoplastic Anti-TIF1 Gamma Positive Adult Onset Dermatomyositis

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Abstract

Anti-transcriptional intermediary factor 1-gamma(anti-TIF1-gamma) antibody is a myositis-specific antibody seen in patients with dermatomyositis, that has the strongest association with paraneoplastic dermatomyositis. We report a case of anti-TIF1-gamma dermatomyositis patient who didn't develop malignancy in 3 years of close follow up and screening. A 37-year-man applied to rheumatology clinic with proximal muscle weakness, erythematous rash spreading over the whole face, neck and dorsal side of hands, weight loss. Muscle strength was 4/5 in both upper and lower extremities, there were Gottron's papules and "V sign" erythema. Creatinine kinase was normal, but patient had elevated aldolase levels. ANA was 1/320 speckled positive. Myositis specific autoantibodies test demonstrated anti-TIF1 gamma positivity. Once anti-TIF1 gamma positivity was detected patient was vigorously screened for presence of malignancy. PET-CT, gastroscopy and colonoscopy were performed, which did not reveal malignancy. EMG findings revealed diffuse myogenic involvement. Cardiac MRI demonstrated myocardial edema. After 3 days of IV pulse 250 mg methylprednisolone, 1mg/kg/day prednisolone was initiated. Methotrexate and hydroxychloroquine were also initiated. Skin lesions regressed but muscle weakness persisted. For this reason, MTX was stopped and mycophenolate mofetil (MMF) 2*2 was started. Due to partial response to MMF, it was reduced to 2*1 and rituximab was initiated. Although the patient was found to be TIF1-gamma positive at the time of diagnosis, no malignancy was detected during 3 years of follow-up. Therefore, this case emphasizes that not all anti TIF1-gamma positive adult dermatomyositis patients present paraneoplastically but vigorous malignancy screening is still indicated in this patient group.

Keywords: Dermatomyositis (DM)

Introduction

Dermatomyositis (DM) is a slowly progressive chronic disease, which is classified among the idiopathic inflammatory myopathies (IIM), is an autoimmune disease that predominantly involves skeletal muscle and skin. Other organs such as heart, lungs may also be affected. Anti-transcriptional intermediary factor 1-gamma (anti-TIF1-gamma) positive patients have predominant skin manifestations, such as heliotrope rash, Gottron papule, V sign, shawl sign, and Holster sign. Anti TIF-1-gamma positivity also has very significant prognostic implications in adult dermatomyositis patients, since it is the myositis specific antibody that has the strongest association with paraneoplastic dermatomyositis. The tendency to detect malignancy in anti-TIF1-gamma positive patients increases especially in older patients.

Early malignancy screening is important in patients with anti-TIF1-gamma positive dermatomyositis (1). We hereby report an anti-TIF1-gamma positive patient who did not have an underlying malignancy after rigorous malignancy screening and a close follow up period of more than 3 years.

Case Report

A 37-year-man applied to rheumatology clinic with proximal muscle weakness, erythematous rash spreading over the whole face, neck and dorsal side of hands, weight loss(8 kg in the last 3 months). Physical examination was performed, which revealed Gottron's papules on the dorsum of the hand, erythematous rash on the neck, characteristic of "V sign". Muscle strength was 4/5 in proximal parts of both upper and lower extremities. Creatinine kinase was within normal range but patient had elevated aldolase levels. ANA was 1/320 positive, in a speckled pattern. Myositis specific autoantibodies panel demonstrated that patient was positive for anti-TIF1-gamma.Punch biopsy from Gottron's papule was consistent with dermatomyositis.Electromyography findings were consistent in favor of diffuse myogenic involvement.Cardiac MRI showed myocardial edema compatible with dermatomyositis. In order to detect a possible underlying malignancy positron emission tolography-computed tomography(PET-CT), thoracic and abdominal computed tomography with IV contrast, gastroscopy and colonoscopy were performed. All of these imgaing modialites were negative for malignancy. Thoracic compute tomograpphy did not demonstrate interstital lung disease. After 3 days of IV pulse 250 mg methylprednisolone, prednisolone was initiated at a dose of 1mg/kg/day which was subsequently tapered. Subcutaneous methotrexate was initiated at a dose of15 mg/week as a steroid sparing agent. Hydroxycholoroquine was initiated for skin lesions. With the onset of treatment, skin lesions regressed but muscle weakness recovered partially at the third month of treatment. For this reason, methotrexate was stopped and mycophenolate mofetil 500 mg 2*2 was started. Due to partial response to MMF at the third month of treatment, its dose was reduced to 2*1 and rituximab was added at a dose of 2*1000 mg IV every 6 months. Patient's muscle symptoms responded well to this treatment combination. Annual PET-CT scans were performed for 3 years after diagnosis of dermatomyositis which did not reveal any malignancies. Patient is currently stable and continues his treatment regimen.

Figure 1: Erythema on the back of the patient, the "shawl sign" of dermatomyositis.

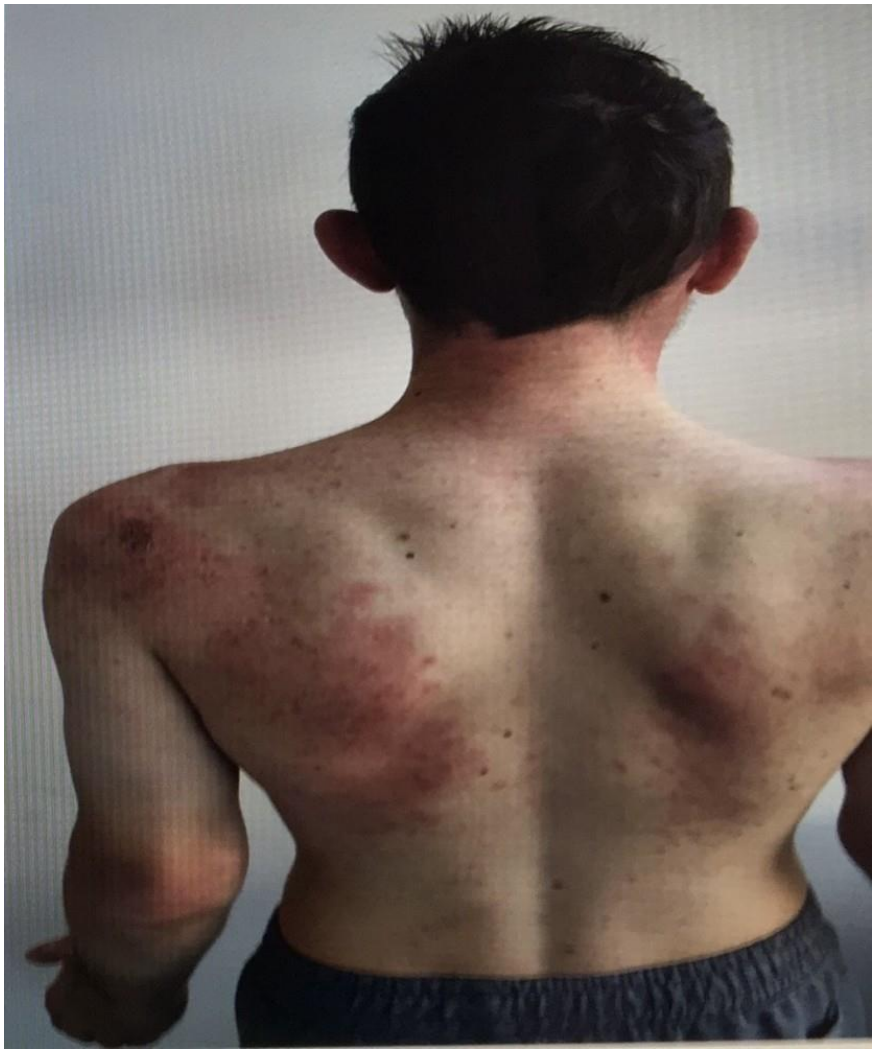

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Figure 2: Periungual erythema and Gottron's papule of the patient

Discussion

Anti-TIF1-gamma positivity is a high risk factor for cancer in patients with dermatomyositis(2). Studies have shown that the incidence of malignancy in anti-TIF1-gamma positive patients is 42.6 % (3). While breast cancer and lymphoma are common in women, cancers affecting almost all parts of the body such as lung, stomach, colon, esophagus, pancreas and thyroid cancer can be seen in men(4). Anti-TIF1-gamma positivity is found in approximately 17% to 35% of JDM patients (5). Juvenile anti-TIF1-gamma positive dermatomyositis can occur without being paraneoplastic. Our patient is also young, in his 30s, so in that sense-due to his young age-the dermatomyositis may have developed without being associated with malignancy**.** Clinicians should recognize that a considerable number of patients with anti-TIF1-gamma positivity do not have and may never develop cancer, especially in younger populations (6). Age related differences in immune response may explain this phenomenon.If juvenile dermatomyositis is compared to adult dermatomyositis where anti-TIF1-gamma positivity is strongly associated with cancer; this difference may be attributed to age-dependent differences in immune system behavior and tumor surveillance mechanisms(7).

According to the "International Guideline for Idiopathic Inflammatory Myopathy-Associated Cancer Screening" of International Myositis Assessment and Clinical Studies Group (IMACS), patients with dermatomyositis who are anti-TIF1-gamma positive, the risk of malignancy is significantly elevated,particularly within the first 3 years following diagnosis (8). Although the patient was found to be anti-TIF1-gamma positive at the time of diagnosis, no malignancy was detected during 3 years of follow-up. We used the annual PET-CT screening protocol of this guideline despite the age of the patient being less than 40 years old, due to the presence of anti-TIF1-gamma antibody and high disease activity despite potent immunsouppression. In these cases of dermatomyositis, it is very important to determine whether paraneoplastic dermatomyositis is present, because this also has a profound effect on treatment decision. In non paraneoplastic cases of dermatomyositis we give immunosuppressive treatments as steroid sparing agents.

In this case report, we explained the clinical features and course of an adult anti-TIF1 gamma dermatomyositis patient with predominant skin, skeletal muscle and cardiac involvement whose malignancy screening remained negative for a follow-up period of 3 years. He is currently in remission under a combination of hydroxychloroquine, mycophenolate mofetil and rituximab regimen.

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