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Case Report

Exploring the Rare Association: A Case Report of Dermatomyositis Coexisting with Type 1 Diabetes Mellitus

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ABSTRACT

Type 1 Diabetes Mellitus (T1DM) is an autoimmune disease characterized by pancreatic β-cell destruction, leading to insulin deficiency and hyperglycemia. Coexisting autoimmune conditions are common, with thyroid diseases being prevalent. Dermatomyositis, a rare systemic autoimmune disorder, primarily affecting muscles and skin, has been infrequently reported in association with Type 1 Diabetes Mellitus, indicating a potential shared genetic susceptibility. This article aims to present a rare case of a 32-year-old patient with the simultaneous occurrence of Type 1 Diabetes Mellitus and dermatomyositis, with focus on the unique challenges and considerations in managing these coexisting autoimmune conditions.

1. Introduction

Type 1 Diabetes Mellitus or insulin-dependent diabetes mellitus (IDDM) is an autoimmune disease characterized by the destruction of pancreatic β-cells, leading to insulin deficiency and hyperglycemia. According to available literature, there is a high prevalence of additional autoimmune diseases in patients with Type 1 Diabetes Mellitus, most commonly those affecting the thyroid gland. Dermatomyositis is a rare, systemic autoimmune disease that is defined by inflammation of the muscles and skin. The coexistence of Type 1 Diabetes Mellitus and Dermatomyositis is extremely rare, with few reports published worldwide. Certain genetic factors may contribute to an increased risk of developing autoimmune diseases and shared genetic susceptibility could explain why some individuals may have both conditions. In this article, we present the case of a patient with such rare association.

2. Case Report

A 32-year-old man was admitted to the Dermatology Department complaining of pruritic skin lesions and muscle weakness. On physical examination, he presented with periorbital, confluent, macular, violaceous (heliotrope) erythema/edema and discolorations on the face (Fig. 1a); erythematous papules over the metacarpophalangeal/interphalangeal joints along with periungual erythema and dystrophic cuticles (Gottron sign) (Fig. 1b); symmetrical confluent, macular erythema over the neck and chest, extending over the deltoid areas and posterior shoulders (the shawl sign) (Fig. 1c, d). The patient reported a history of 6 months with these skin lesions, accompanied by proximal muscle weakness in the upper extremities over the last three months. This weakness led to difficulties in routine activities, such as lifting weights or ironing clothes.

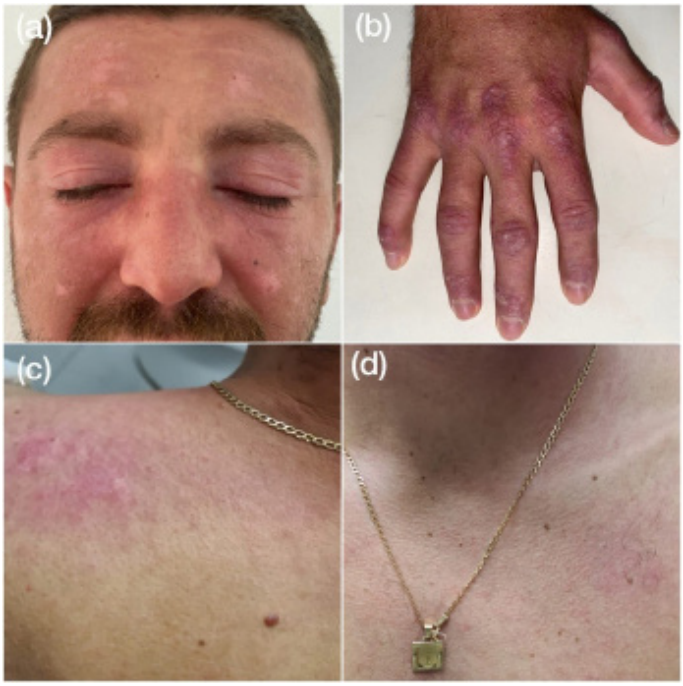

Fig. 1. ***a:*** *Periorbital confluent, macular, violaceous (heliotrope) erythema and discolorations;* ***b:*** *Gottron papules over the distal metacarpophalangeal/interphalangeal joints along with periungual erythema and dystrophic cuticles;* ***c, d:*** *Confluent erythematous papules over the upper back and neck (shawl sign).*

His personal history includes a diagnosis of Type 1 Diabetes Mellitus at the age of 9. He was actually being treated with Insulin Glargine 22 UI/sc and Insulin Aspart 8-12-8 UI/sc. His family history was negative.

Laboratory findings were as follows: glucosuria 1000 mg/dl; fasting plasma glucose level 222 mg/dl, glycosylated haemoglobin (HbA1c) 8.6% (normal <5.7%). Red blood cell count was within normal range. Aspartate aminotransferase (AST) 48 U/L (normal 5-34 U/L), lactate dehydrogenase (LDH) 282 U/L (normal 125-220 U/L), creatine kinase (CK) 433 U/L (normal 30-200), C reactive protein (CRP) 0.21 mg/dL (normal <0.5), Fibrinogen activity 375 mg/dL (normal 200-400 mg/dL). CEA 18.2 ng/ml (normal <5); AFP < 2 ng/ml (normal <8.8 ng/ml), CA 125 negative; CA 19-9 negative. Antinuclear antibodies (ANA) titer 1:320 positive (normal < 1:160); Anti ds DNA 5.8 IU/ml (normal <100); extractable nuclear antigen (ENA) negative.

Dermoscopy of the nailfolds (capillaroscopy) was performed on the patient and it showed the presence of elongated capillaries, dilated capillary loops, cuticular hemorrhage and cuticular overgrowth (Fig. 2).

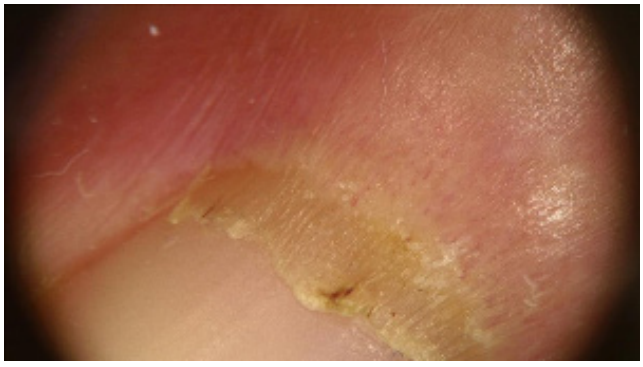

Fig. 2. *Dermoscopy of nailfold (capillaroscopy) shows elongated capillaries, dilated loop capillaries, capillary hemorrhage and cuticular overgrowth.*

Histological examination of a skin specimen revealed hyperkeratosis, inflammatory perivascular plasmo-lymphocytic infiltrates and perivascular/periadnexal myxoid changes; features compatible with dermatomyositis (Fig. 3a, b).

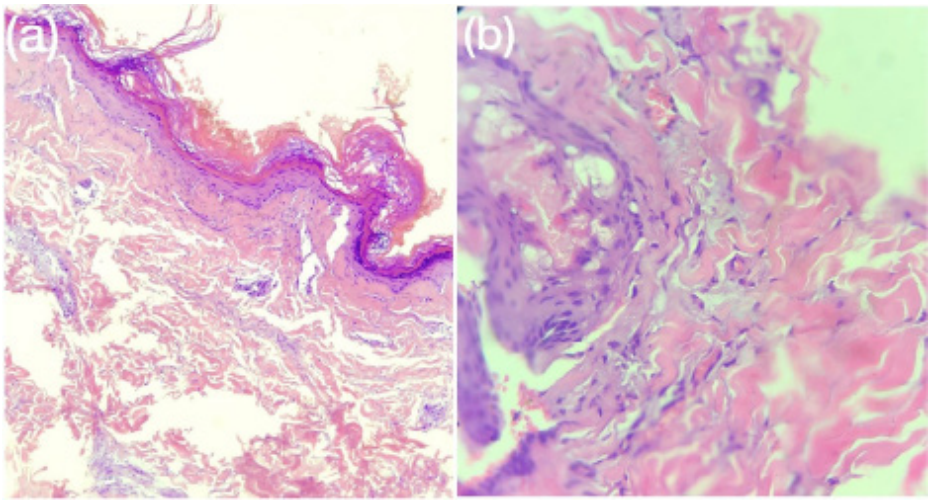

Fig. 3. ***a:*** *Hyperkeratosis, subtle vacuolar interface change, and increased dermal mucin;* ***b:*** *Increased dermal mucin between collagen fibers.*

An electromyogram revealed lesions of degeneration with myopathic pattern of the motor unit action potential along with a myopathic recruitment pattern, compatible with myositis.

Electrocardiogram (ECG) was normal.

The physical examination and laboratories findings, as well as skin and muscle biopsies, confirmed the diagnosis of Dermatomyositis in a patient with a previously diagnosed Type 1 Diabetes Mellitus.

3. Discussion

Dermatomyositis (DM) is a rare, systemic autoimmune disease that is defined by inflammation of the muscles (myositis) and skin (dermatitis). It is presented with a characteristic rash and progressive symmetrical proximal muscle weakness (1). Based on the inflammatory nature of the muscle and skin manifestations, as well as the characteristic humoral autoimmune abnormalities, DM is believed to be caused by a genetically determined, aberrant autoimmune response to environmental factors (2, 3). In the majority of cases, the strongest genetic association is with Human Leucocyte Antigen (HLA) class II alleles. In addition, an array of environmental factors has been linked to DM pathogenesis, including infections (particularly viruses), ultraviolet radiation (DM rates are correlated with proximity to the equator), vitamin D deficiency (common with several autoimmune diseases) and drugs (4).

Dermatomyositis is also associated with a 6-fold increased risk of malignancy compared with the general population (5). Dermatomyositis has been linked to various types of cancer, with breast, ovarian, lung, and hematologic cancers being commonly reported. DM in these cases is considered a paraneoplastic syndrome (5, 6).

Additionally, DM can be associated with non-neoplastic conditions such as vasculitis. Connective tissue disorders like progressive systemic sclerosis, rheumatoid arthritis, mixed connective disease, and systemic lupus erythematosus can also coexist in an overlap group with dermatomyositis. Endocrine diseases like thyroiditis, Cushing syndrome, Crohn disease, and hypophysism have also been linked to DM (6).

Diabetes mellitus is a chronic metabolic disorder, defined by elevated blood sugar over a prolonged period of time. It occurs when the body either doesn't produce enough insulin or cannot effectively utilize it (7). There are two main types of diabetes mellitus: Type 1 Diabetes Mellitus or insulin-dependent diabetes mellitus (IDDM) or juvenile-onset diabetes which is an autoimmune condition where the body's immune system attacks and destroys the insulin producing cells in the pancreas, which results in little to no insulin production; and Type 2 Diabetes Mellitus, non-insulin-dependent diabetes mellitus (NIDDM) or adult-onset diabetes, the most common type, which is characterized by insulin resistance, meaning the body's cells do not respond effectively to insulin (7, 8).

Type 1 Diabetes Mellitus affects around 5-10% of diabetes cases. It is an autoimmune disorder characterized by the destruction of pancreatic β-cells, leading to insulin deficiency and hyperglycemia. The disease's progression varies, with some cases showing rapid β-cell destruction in children and adolescents, resulting in diabetic ketoacidosis. In other cases, the disease progresses slowly, with mild increases in fasting blood glucose levels, only becoming severe under physiological stress conditions (9).

Type 1 Diabetes Mellitus (T1DM) is associated with immune markers, particularly autoantibodies, such as glutamic acid decarboxylase autoantibodies (GADAs), islet cell autoantibodies (ICAs), and insulin autoantibodies (IAAs). Multiple genes, including major histocompatibility complex (MHC) class II alleles and human leukocyte antigen (HLA)-DR3 and DR4, have been found in a large percentage of T1DM patients (8, 9).

According to available evidence, individuals with Type 1 Diabetes Mellitus are more likely to have additional autoimmune diseases due to a common genetic background (10). A total of 80 different autoimmune diseases can be observed in Type 1 Diabetes Mellitus, but celiac disease and hypothyroidism are the most frequently observed, followed by vitiligo, hyperthyroidism, autoimmune adrenalitis, gonadal insufficiency, autoimmune hepatitis, dermatomyositis, and myasthenia gravis (10, 11). Due to the similar pathogenesis and immunological processes of various autoimmune diseases, they sometimes occur in the same family or individual (11).

The co-occurrence of Dermatomyositis and Type 1 Diabetes Mellitus has been reported in medical literature, but it is considered very rare. A study conducted by Petrasovicová, V et al. found a relatively high incidence of Type 1 Diabetes Mellitus occurring in 16% of patients with polymyositis. This suggests a potential coexistence of T1DM and dermatomyositis (12). Certain genetic factors may contribute to an increased risk of developing autoimmune diseases like dermatomyositis and Type 1 Diabetes Mellitus. Shared genetic susceptibility could explain why

some individuals may have both conditions (13).

Sattar, M A et al. describes the coexistence of rheumatoid arthritis, ankylosing spondylitis and dermatomyositis in a patient with diabetes mellitus and the associated linked HLA antigens A2, A9, B8, B27, DR3, and DR4 (14). Charalabopoulos, Konstantinos et al. also reports a rare case of DM in association with Type 1 Diabetes Mellitus in a 28-year-old man (15). The limited literature reports indicate the uncommon occurrence of the association between these two diseases.

It is important to note that the presence of one autoimmune condition may increase the risk of developing other autoimmune conditions. The exact underlying mechanisms and factors contributing to the simultaneous occurrence of these two conditions are not fully understood, but it is thought that it might be influenced by autoimmune predisposition, shared genetic factors, environmental factors and chronic inflammation (16).

A potential shared immunopathogenic mechanism involves molecular mimicry, wherein viral or environmental triggers induce an aberrant immune response leading to autoimmunity. Viral infections, such as enteroviruses, have been implicated in both T1DM and DM, possibly initiating immune-mediated tissue destruction. Chronic inflammation and cytokine dysregulation, particularly involving interferon pathways (e.g., Type I and Type II interferons), have also been implicated in both diseases. This suggests that an innate immune response driven by viral or other environmental exposures could contribute to the simultaneous development of these autoimmune disorders (17).

In the presented case the diagnosis of T1DM is confirmed by the presence of glucosuria 1000 mg/dl; fasting plasma glucose level 222 mg/dl, glycosylated hemoglobin (HbA1c) 8.6%; while the diagnosis of DM is based on the histopathologic examination of skin and muscle specimen and electromyography (EMG), both with features compatible with dermatomyositis.

There is a contradiction in the management of the diseases, since the therapy for DM includes Prednisone, a corticosteroid which is contraindicated in diabetic patients due to the risk of disrupting glucose control and causing acute decompensation.

In such cases, alternative immunosuppressive strategies should be considered. Steroid-sparing agents such as mycophenolate mofetil, azathioprine, methotrexate, and calcineurin inhibitors have demonstrated efficacy in managing dermatomyositis while minimizing the metabolic complications associated with long-term corticosteroid use. Additionally, biologic therapies like rituximab, which targets B cells, have shown promising results in refractory dermatomyositis and may provide a viable treatment option in patients with diabetes (18).

When corticosteroid therapy is necessary despite its risks, careful insulin therapy adjustments are crucial to prevent hyperglycemia and acute decompensation. Patients may require an increase in both basal and prandial insulin doses, with close glucose monitoring to assess and modify the regimen accordingly. Continuous glucose monitoring can aid in identifying fluctuations and optimizing insulin delivery (19).

A multidisciplinary approach involving rheumatologists, endocrinologists, and dermatologists would be necessary to provide comprehensive care and management of both diseases.

4. Conclusions

In summary, in addition to the well-established associations between autoimmune diseases, it is important for clinicians to be aware that a coexistence of Type 1 Diabetes Mellitus with dermatomyositis (DM) may also occur. Regular monitoring and coordination between specialists are crucial to ensure the best possible outcomes for the patient and prevent any potential complications.

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DISCLOSURE

All authors report no conflict of interest.

DATA AVAILABILITY STATEMENT

Data openly available in a public repository that issues datasets with DOIs.

ETHICS STATEMENT

The patients in this manuscript have given written informed consent to publication of their case details.

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