Paraneoplastic Dermatomyositis in a 34-Year-Old Filipino Female with Stage IV Invasive Breast Cancer: A Case Report*

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Abstract

Dermatomyositis (DM) is a rare autoimmune myopathy characterized by progressive muscle weakness that typically affects the shoulder and hip girdle first. It is a multisystem disorder characterized by symmetric proximal, extensor, inflammatory myopathy, vascular involvement and a constellation of pathognomonic (Gottron papules, Gottron Sign, Heliotrope rash) and characteristic cutaneous eruptions, which frequently presents in the fifth and sixth decades. As adult DM presents as a paraneoplastic syndrome in up to 30% of cases, metastatic workups appropriate for age are warranted.

Keywords: Dermatomyositis, Paraneoplastic Dermatomyositis, Breast Cancer, Paraneoplastic Syndrome

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INTRODUCTION

Dermatomyositis is an autoimmune disease belonging to the spectrum of idiopathic inflammatory myopathies. It is a rare disease with a worldwide occurrence, with an incidence ranging from 2 to 9 per million across various populations and affects women more than men. High index of suspicion must be maintained for malignancies, especially in treatment resistant patients due to its association with dermatomyositis. Successful treatment of an associated neoplasm is often followed by improvement/resolution of Dermatomyositis.

CASE REPORT

This is a case of a 34-year-old female with 2 months history of asymptomatic, erythematous patches on the malar area after prolonged sun exposure. The patient noted progression in number and size of the lesions, eventually involving the scalp, forehead, neck, chest and extremities, with associated pruritus and proximal muscle weakness. She was managed with oral and topical corticosteroids and sun protection showing only moderate response.

Histopathological analysis revealed epidermal atrophy, basal layer vacuolization, basement membrane thickening, superficial perivascular, infiltrates of primarily lymphocytes, and mucin deposition in the dermis. Tissue Direct Immunofluorescence (DIF) was done revealing deposits of +1 C3 and +1 IgM in a linear pattern at the basement membrane zone.

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Patient was also noted with an incidental finding of firm, movable, non-tender mass on the left breast measuring 3x3 cm, and excruciating lower back pain which were extensively worked up, and was diagnosed as a case of Paraneoplastic DM; Invasive Breast Cancer, Stage VI with Bone Metastasis; Compound Fracture C6 secondary to consider Bone Metastasis.

While there is no consensus metastatic workup in patients with dermatomyositis, diagnostics appropriate for age is deemed sufficient. Of note, this patient presented with advanced stage breast cancer in an unusually young age.

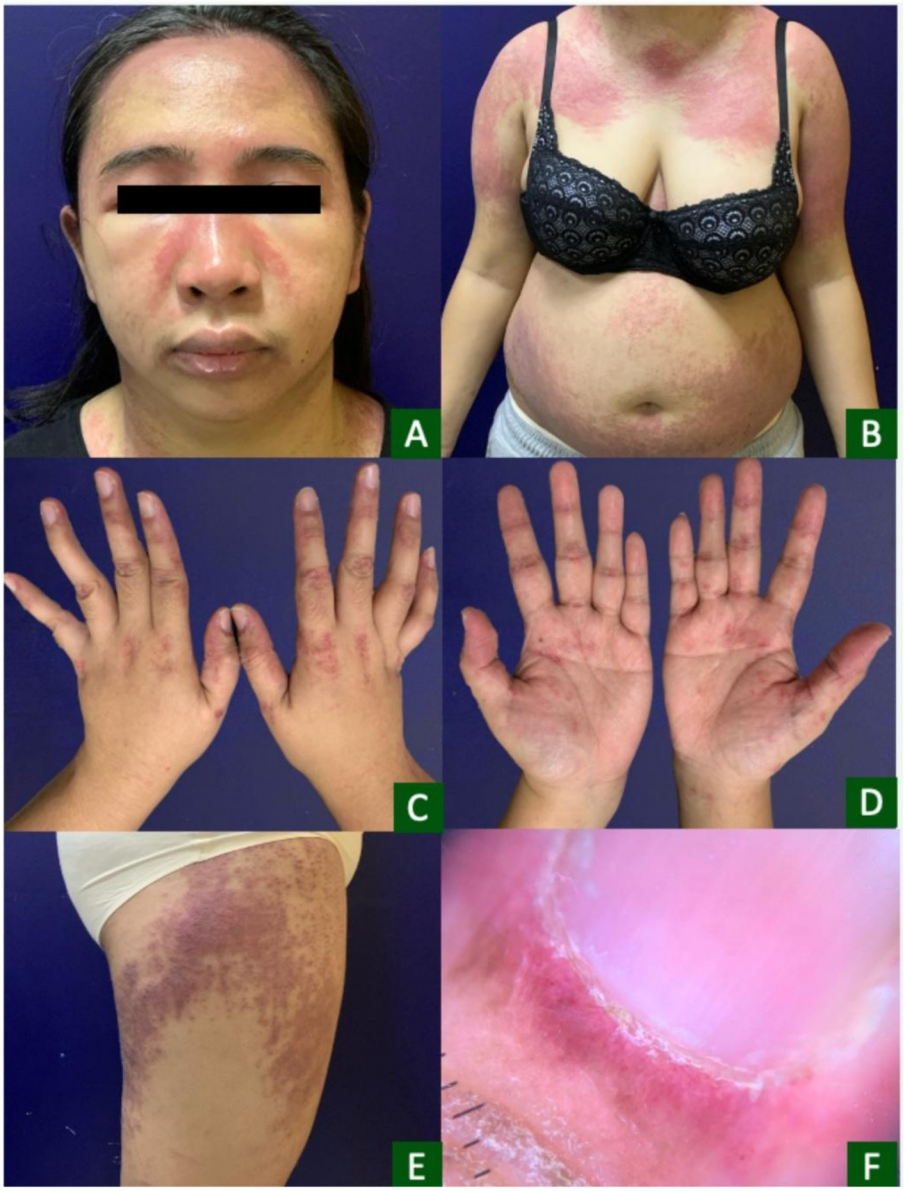

Figure 1. Photographs showing A) Heliotrope rash, Periorbital and facial edema, and scalp involvement, B) Shawl sign ang V Sign C) Gottron sign and Gottron papules, D) Mechanic's hands, E) Holster sign and F) Nailfold changes,

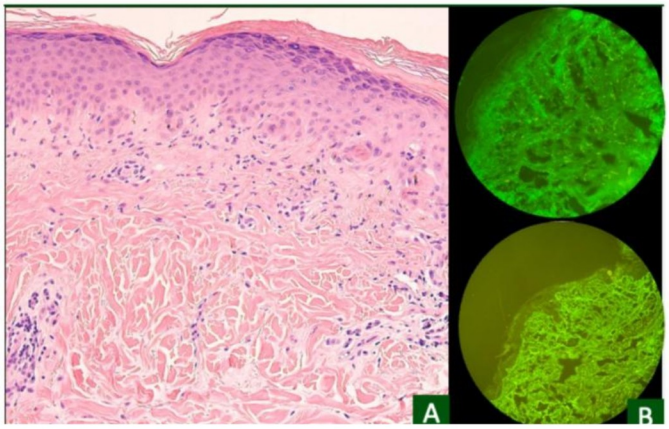

Figure 2. A) Histopathologic report shows; epidermal atrophy, Basal layer vacoulization, Basement membrane thickening, Superficial perivascular infiltrates of primarily lymphocytes, Dermal mucin deposition, while, B) DIF shows deposits of +1 C3 and +1 IgM in a linear pattern at the basement membrane zone.

DISCUSSION

Dermatomyositis (DM) is an autoimmune disease belonging to the spectrum of idiopathic inflammatory myopathies which also includes inclusion body myositis and polymyositis. It is a rare disease with a worldwide occurrence, with an incidence ranging from 2 to 9 per million across various populations, and affects women more than men (2-3:1). DM has a bimodal age distribution, with juvenile DM (JDM) most commonly diagnosed between 4 and 14 years of age and adult DM diagnosed between 40 and 60 years of age.[1]

Local data shows that Dermatomyositis is predominantly seen in 4th to 5th decade of life, and is far more common in females than in males. Our patient is a 34-year-old female and is the second most common demographic for this case.[2]

In our institution, there has only been 6 new cases of Dermatomyositis since 2020. Of note, according to the PDS-HIS data, there are only 3 reported cases of paraneoplastic dermatomyositis.

The pathogenesis of DM is multifactorial, complex, and incompletely understood. Genetic, environmental, and immune mechanisms, are implicated in dermatomyositis development. Multiple environmental factors may trigger chronic immune activation such as ultraviolet radiation, viral infections, medications, and smoking.[2]

A diagnosis of cutaneous DM is suggested by the constellation of characteristic cutaneous findings, muscle weakness, and laboratory findings of myositis. However, in patients who present with ambiguous cutaneous findings or cutaneous findings that are suggestive of DM in the clinical absence of muscle involvement, a skin biopsy should be performed.[2] Traditionally, skin findings have been divided into pathognomonic (Gottron papules, Gottron sign, and heliotrope rash), characteristic, compatible, less common, rare, and nonspecific. Patients may present with one or more DM-related skin changes. Lesions are often pruritic or burning and are usually photosensitive, which can significantly impact patients' quality of life. Cutaneous disease may persist for years. In one systematic review of primarily adult patients with cutaneous DM without myositis, the mean duration of skin disease was 4.5 years[2]

Approximately 80% of patients with DM have myopathy. The clinical course of DM skin manifestations may precede or follow myositis. The classic muscular manifestation is acute or subacute onset of painless, symmetric, proximal muscle weakness. In the patients with cutaneous presents with manifestations suspicious for DM, investigation for concomitant muscle disease must be done. Muscle examination and serum creatinine kinase, which is the most sensitive muscle enzyme, and aldolase levels are recommended every two to three months.[2]

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Dysphagia, dysphonia, and aspiration raises suspicion for involvement of striated muscle of the pharynx and esophagus, and are associated with a poor prognosis. However, DM is not associated with sensory loss, ptosis, involvement of the extraocular muscles, or abnormal reflexes, which can help differentiate it from other neuromuscular disorders. Those with DM-consistent skin findings but without myopathy have what is termed Clinically Amyopathic Dermatomyositis (CADM). CADM may by amyopathic, or hypomyopathic, with evidence of subclinical muscle involvement on laboratory testing, biopsy, or imaging. Arthralgias are seen in 30-40% in DM, involving the small joints of the hands with mild to moderate pain. Both muscular and joint symptoms were observed in our case.

Therapy for cutaneous disease is usually indicated due to the severe pruritus and patient distress over the appearance of skin lesions. The management of cutaneous manifestations of DM can be difficult and are often more resistant to therapy than the myositis.

The initial approach for cutaneous DM typically consists of four approaches: 1) Aggressive photoprotection, 2) Antipruritic agents, 3) Topical corticosteroids or topical calcineurin inhibitors, 4) Systemic medications aimed at attaining sustained control.[1] Most patients require systemic treatment with antimalarial drugs, methotrexate, or other medications.[4]

The long-term survival rate for adults with DM is approximately 65% to 75%, but there are relatively limited data regarding specific organ outcomes such as muscle or skin disease. Major causes of death include malignancy, pulmonary or cardiac disease, and infection. Poor prognosticating factors include older age, male gender, nonwhite race, longer duration of symptoms, and presence of anti-MDA5 antibodies.[6]

DM has variable complications. One of which is the consequences of long-term use of immunosuppressive medications, including opportunistic systemic infections and opportunistic infection-induced lymphoma such (i.e., Epstein–Barr virus infection).[3] Adult-onset classic DM is associated with a higher-than-expected risk for internal malignancy, carcinomas being the most common. Individuals 50 years of age or older at the time of onset of classic DM appear to have a definite 20-30% increased risk of malignancy. In patients with malignancy, the myositis and cutaneous manifestations are less responsive to systemic glucocorticoid therapy. Older age, greater extent and severity of skin disease, and elevated CK levels appear to enhance the risk of malignancy in adults with classic DM.

This case highlights the importance of having a high index of suspicion for cancer in patients with DM, especially when resistant to treatments. Paraneoplastic DM typically improves with the treatment of the implicated cancer.

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