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Case Report

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A Case of a 50-Year-Old Female with Dermatomyositis Secondary to a Paraneoplasm (Infiltrating Ductal Carcinoma, Grade 2)

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ABSTRACT

Introduction: Dermatomyositis is an autoimmune disorder with an incidence of 5 to 10 per 1 million per year. 10 to 20% of patients with adult-onset Dermatomyositis have an underlying malignancy. There are currently no specific cutaneous markers for Paraneoplastic Dermatomyositis. This case highlights a patient with uncommon features of paraneoplasm after breast malignancy.

Case Report: A 50-year-old female with Infiltrating Ductal Carcinoma, presented with patches on the face, neck, chest, upper back, outer arms, dorsal hands, thighs, hair loss and weakness. Histopathology revealed widened basal layer vacuolization with widened spaces between collagen bundles filled with mucin deposition, and pigment incontinence in the papillary dermis. Direct immunofluorescence showed granular deposits of IgG, C3, IgM and IgA in the basement membrane zone, which are consistent with Dermatomyositis. The patient was prescribed with Clobetasol ointment mixed equally with petrolatum on the body and Mometasone cream on the face. She was subsequently started on Prednisone at 0.5 mkd with eventual tapering, Omeprazole, and Calcium + Vitamin D tablet daily. Hydroxychloroquine 200 mg/tab daily was added. Referral to Rheumatology for co-management and Surgery for management of the breast mass was also done.

Conclusion: This case emphasizes the rare occurrence of a Paraneoplastic Dermatomyositis following a diagnosis of breast malignancy. A careful review upon diagnosis may detect occult malignancies especially in patients who present primarily with cutaneous Dermatomyositis. Clinicopathologic correlation and a multi-disciplinary approach, with Surgery, Oncology, Rheumatology and Dermatopathology are needed to identify the underlying cause and facilitate successful treatment of a neoplasm followed by improvement of Dermatomyositis.

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Introduction

Dermatomyositis is a genetically determined autoimmune disorder that predominately targets the skeletal musculature and/or skin and results in symptomatic skeletal muscle weakness and/or cutaneous inflammatory disease. It usually presents in the fifth and sixth decade, affecting more females than males, and is known to be associated with malignancy especially in patients who present with Dermatomyositis during adulthood.

Case Report

This is a case of a 50-year-old Filipino female who initially presented at our institution with patches on the face, neck, chest, back, extensor aspect of the arms and dorsal aspect of the hands. History of present illness started when the patient noted erythematous patches on the forehead, associated with occasional pruritus graded 5/10. Subsequently, the patient noted persistence of the patches on the forehead, spreading to the cheeks, nose, chin, anterior neck and chest, nape, shoulders, both extensor upper arms, dorsal aspect of the hand, and the back, aggravated by sun exposure and associated with intake of seafood. Interval history showed appearance of erythematous papules on the dorsal aspect of the hands, followed by diffuse hair loss. The patient also observed weakness of both of her arms manifested as difficulty reaching her back when closing her bra strap. Persistence led to consult at our institution. The patient was diagnosed with Infiltrating Ductal Carcinoma in July 2018, status post excision biopsy of the right breast mass in the same year. The patient had no swelling, no limited range of motion, but with Manual Muscle Testing of 5/5 on all extremities except the right upper arm at 4/5 with negative Gower sign. Cutaneous examination showed multiple, ill to well-defined, irregularly shaped erythematous to hyperpigmented, lichenified patches on the forehead, cheeks sparing the nasolabial folds (malar rash), anterior neck and chest (V sign), back, shoulders (Shawl sign) and both upper outer arms, as well as multiple, well-defined erythematous patches (Gottron sign), and papules on the dorsal aspect of the proximal interphalangeal joints and metacarpophalangeal joints (Gottron papules), periungual telangiectasia on both hands, and diffuse thinning of hair (Figure 1). Complete Blood Count, Urinalysis, ALT, BUN, Creatinine, ANA, Lupus Panel, CK MB, CK MM, were done and revealed normal results, however, AST and LDH were elevated. Modified Radical Mastectomy was being planned for the patient care of the service of General Surgery. The patient

was initially prescribed by Dermatology service with Clobetasol ointment + Vaselina Blanca (1:1) twice a day on affected areas of the body and Mometasone Furoate cream on affected areas of the face. The patient was advised sun protection and use of sunscreen with SPF 30 or more. The patient was also referred to the service of Rheumatology for co-management. Our service (Dermatology) performed a skin punch biopsy and revealed widened basal layer vacuolization with widened spaces between collagen bundles filled with mucin deposition, as well as pigment incontinence in the papillary dermis, which is consistent with a connective tissue disease such as Dermatomyositis. (Figure 2). Direct immunofluorescence showed granular deposits of IgG, C3, IgM and IgA in the basement membrane zone compatible with a diagnosis of connective tissue disease including Dermatomyositis (Figure 3). The patient was started on Prednisone at 0.5 mkd, Omeprazole 20 mg/tab 1 tab once daily and Calcium + Vitamin D tab daily. The patient was advised to have the following repeat laboratories to check response to treatment: CBC was normal, but the AST, LDH and ESR were elevated. At this time, a muscle biopsy and EMG were advised, however, was not done by the patient. Improvement of symptoms were noted with intake and application of medications, but the patient had new erythematous patches on both lateral aspects of the thighs (Holster sign), thus, both Rheumatology and Dermatology services agreed to increase Prednisone to 0.7 mkd for 2 weeks, and to start Hydroxychloroquine 200 mg/tab daily and continue Omeprazole and Calcium + Vitamin D. The patient was still for EMG and muscle biopsy, and was for repeat AST, LDH, CBC, and ESR after 2 weeks. Sun protection was emphasized as well as consult with Rheumatology, Oncology and Surgery for multidisciplinary management. Since the patient is diagnosed with Infiltrative Ductal Carcinoma stage 2, a Modified Radical Mastectomy was recommended by the service of Surgery. The patient was slowly tapered off Prednisone but was lost to follow-up afterwards.

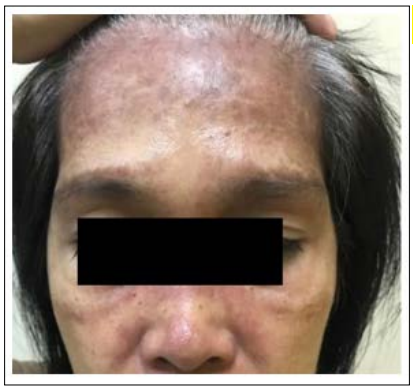

Figure 1A: Hyperpigmented, Lichenified Patches on The Forehead, Cheeks Sparing the Nasolabial Folds (Malar Rash)

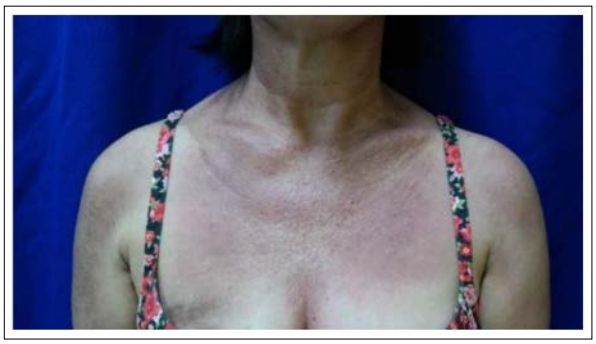

Figure 1B: Erythematous Patches on the Anterior Neck and Chest (V sign)

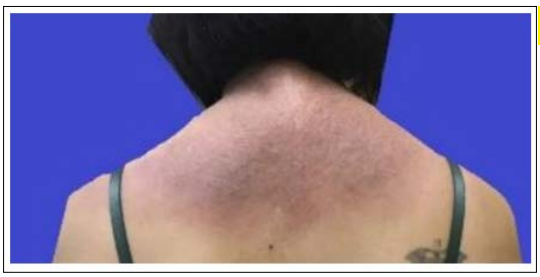

Figure 1C: Erythematous to Hyperpigmented Patches on the Back, Shoulders (Shawl sign)

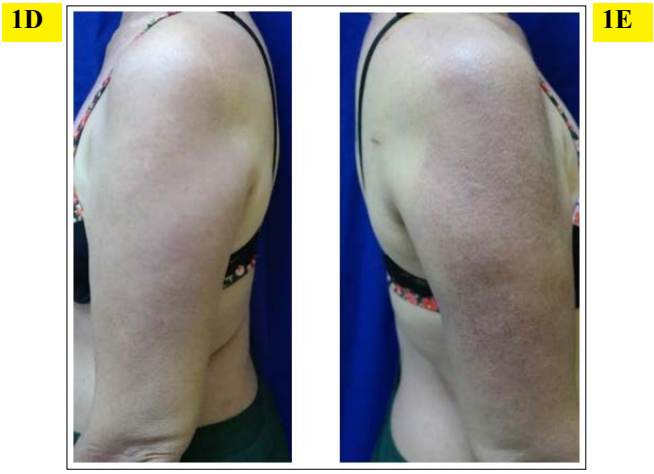

Figure 1D, 1E: Erythematous to Hyperpigmented Patches on the Upper Outer Arms

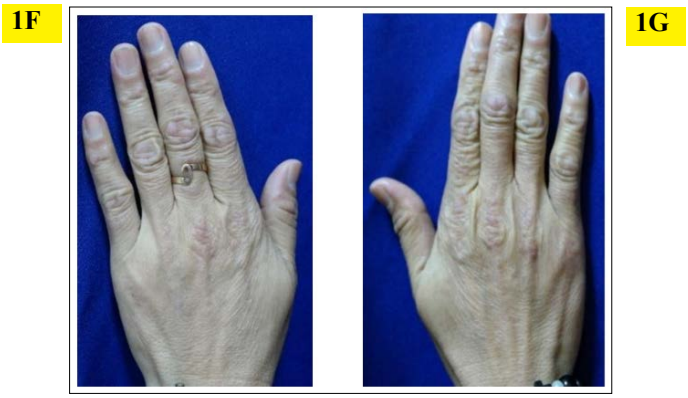

Figure 1F, 1G: Multiple, Well-Defined Erythematous Patches (Gottron Sign), And Papules on the Dorsal Aspect of the Proximal Interphalangeal Joints and Metacarpophalangeal Joints (Gottron Papules)

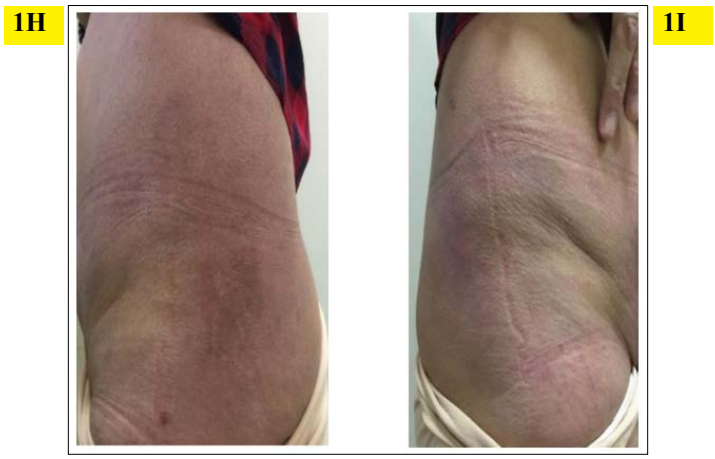

Figure 1H, 1I: Erythematous Patches on Both Lateral Aspects of the Thighs (Holster sign)

Figure 2: Skin Punch Biopsy from the Back

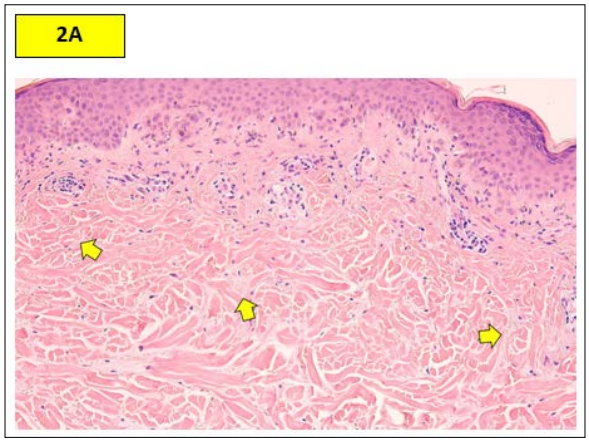

Figure 2A: Low Power Magnification Shows Widened Basal Layer Vacuolization with Widened Spaces Between Collagen Bundles Filled with Mucin Deposition (yellow arrow)

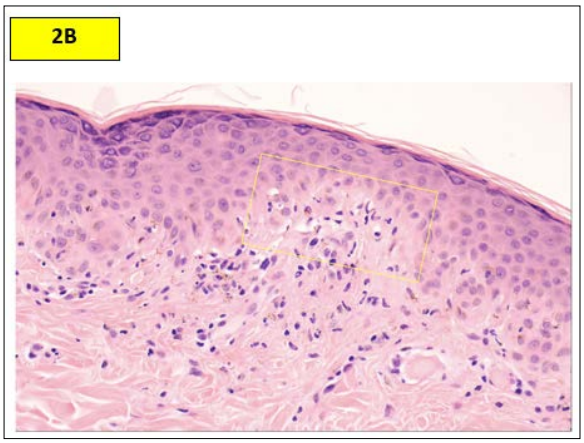

Figure 2B: Higher Magnification Focusing on the Basal Layer Vacuolization as well as Pigment Incontinence in the Papillary Dermis (Yellow Box)

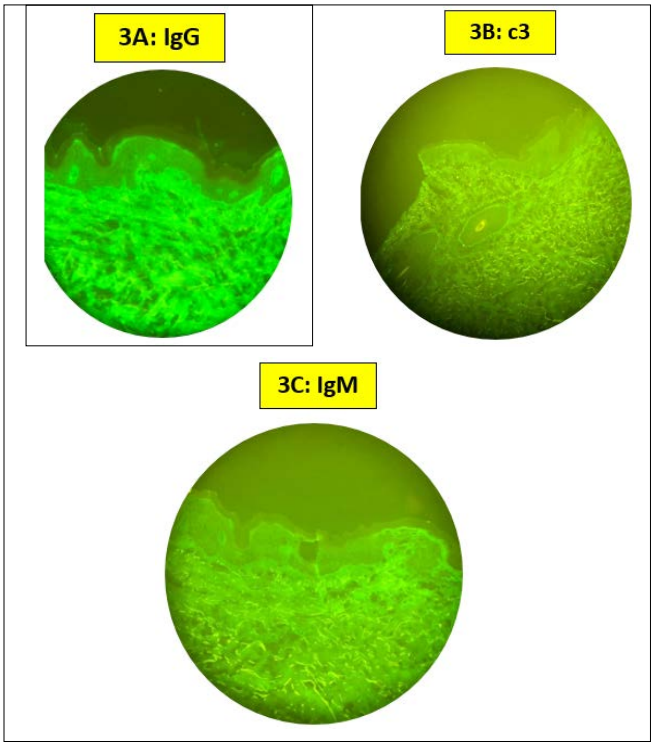

Figure 3: Direct Immunofluorescence from the Back

LPO shows granular deposits of **(3A)**: IgG (+1), **(3B)**: C3 (+1), **(3C)**: IgM (intermittent, +/-), IgA (intermittent, +/-), and no abnormal deposits of fibrinogen in the basement membrane zone.

Discussion

Dermatomyositis is an autoimmune disorder that predominately targets the skeletal musculature and skin. It most frequently presents in the fifth and sixth decades, affecting more females than males. Skin change is described as symmetric, confluent, macular violaceous erythema variably affecting the skin overlying the extensor aspect of the fingers, hands, and forearms (Gottron sign); the arms, deltoid areas, posterior shoulders, and neck (the shawl sign); the V area of the anterior neck and upper chest (V sign); and the central aspect of the face, periorbital areas (Heliotrope), forehead, and scalp, as well as flat-topped, violaceous papules (Gottron papules) with atrophy on the nape of the neck and shoulders and over the knuckles and interphalangeal joints. The lateral aspects of the hips and thighs (holster sign) are also frequently involved. If there is muscle involvement, it shows as symmetric weakness of the proximal muscles of the extremities. With lower-extremity weakness, the initial clinical finding includes difficulty in performing routine activities of daily living, such as rising from a chair or bathtub and climbing stairs. Weakness of the upper extremities soon follows, often manifested by difficulty in raising the arms above the head to perform routine activities such as combing the hair. Pain or tenderness in the affected muscle groups commonly occur. Some patients experience a fulminant disease course that results in disabling weakness. Disease triggers include environmental factors, and infection with coxsackievirus, parvovirus B19, Epstein–Barr virus, human immunodeficiency virus, and human T-cell leukemia virus type 1 [1-4]. Based on the Philippine Dermatological Society Health Information System data, which were collected from the years 2011 to 2018, there were a total of 162 reported new cases of Dermatomyositis. 56 were new male patients and 106 were new female patients, out of which, only 2 were reported diagnosed

with Dermatomyositis associated with malignancy. Paraneoplastic syndromes show in up to 25% of individuals with adult-onset dermatomyositis [5]. In individuals older than 50 years, there is a 20 to 30% higher risk of malignancy including breast cancer [6]. In Asia, Dermatomyositis is most associated with cancers of the nasopharynx, lungs and hematopoietic malignancies [7]. In Paraneoplastic Dermatomyositis, the malignancy can precede a diagnosis of Dermatomyositis, can appear during the disease course, or even follow the diagnosis of the condition [8]. The Bohan and Peter criteria have been widely used in the diagnosis of Dermatomyositis. The criteria include symmetric proximal muscle weakness, elevation of serum skeletal muscle enzyme, electromyographic changes, muscle biopsy abnormalities and typical skin rash of Dermatomyositis [9]. A skin punch biopsy and direct immunofluorescence help to rule in Dermatomyositis, but specific cutaneous findings and muscle weakness are characteristic and can distinguish Dermatomyositis when ruling out autoimmune diseases such as Systemic Lupus Erythematosus [1]. Paraneoplastic disease can occur not only in association with the primary tumor, but also in case of recurrence or progression of the tumor disease. Thus, tumor screening may be beneficial to individuals with a history of malignancy and who present with cutaneous manifestations and myopathy compatible with dermatomyositis [10]. Mortality rates vary from 25% to 80%. Individuals who receive early systemic treatment for their disease have a better prognosis [1]. Since we are treating this as a case of Dermatomyositis secondary to a paraneoplasm, it is important to emphasize diligent follow up with other services like Surgery and Oncology, to monitor for response and recurrence of cancer [11]. There can be rapid regression of Dermatomyositis symptoms following treatment of breast cancer with surgery [12]. Systemic glucocorticoids remain the first-line therapy for Dermatomyositis, with early administration being associated with a better overall prognosis. Hydroxychloroquine sulfate, Quinacrine and Chloroquine are also therapeutic options. In individuals who cannot tolerate the side effects of long-term high dose glucocorticoid treatment, medications like Azathioprine, Cyclophosphamide, methotrexate, chlorambucil, cyclosporine and mycophenolate may be alternatives [1].

Conclusion

This case emphasizes the rare occurrence of a Paraneoplastic Dermatomyositis following a diagnosis of malignancy, particularly breast cancer. In the Philippines, only 2 of the 162 new cases of Dermatomyositis reported to the Philippine Dermatological Society, from 2011 to 2018, had Dermatomyositis associated with malignancy. A careful review upon diagnosis may detect occult malignancies especially in patients who present primarily with cutaneous Dermatomyositis. Clinicopathologic correlation and a multi-disciplinary approach, with Surgery, Oncology, Rheumatology and Dermatopathology are needed to identify the underlying cause and facilitate successful treatment of an associated neoplasm followed by improvement of Dermatomyositis.

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