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Amyopathic Dermatomyositis Revealing Triple-Negative Breast Cancer: A Case Report and Literature Review

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Abstract | **Case Report**

Amyopathic dermatomyositis (ADM) is a rare form of dermatomyositis characterized by specific cutaneous lesions without muscle involvement. It can reveal an underlying malignancy, particularly breast carcinoma. We report the case of a 48-year-old woman presenting with a skin eruption suggestive of ADM, preceding the diagnosis of triple-negative breast cancer. Paraneoplastic etiology was confirmed by the presence of anti-TIF1γ antibodies. Management included corticosteroids, intravenous immunoglobulins, and neoadjuvant chemotherapy followed by mastectomy, resulting in rapid improvement in cutaneous manifestations. One-year post-treatment, no recurrence has been observed. This case highlights the importance of systematic oncologic screening in ADM and the value of multidisciplinary care.

Keywords: Amyopathic dermatomyositis, Triple-negative breast cancer, Diagnosis, Personalized treatment, Cancer screening.

INTRODUCTION

Dermatomyositis (DM) is a rare inflammatory myopathy characterized by specific cutaneous manifestations, proximal muscle weakness, and, in some cases, systemic involvement. Its amyopathic form accounts for approximately 10–20% of cases [1]. DM may reveal an underlying neoplastic process in 15–30% of cases, particularly breast cancer [2], which represents the second most common association in Asia [3]. To date, no standardized protocol exists for the management of breast cancer associated with DM, requiring a personalized therapeutic approach. We report the case of a patient diagnosed with triple-negative breast cancer (TNBC) and amyopathic dermatomyositis, who showed significant remission following treatment.

CASE REPORT

A 48-year-old woman with no significant past medical history (G7P5), using oral contraception for 12 years, presented with a progressive cutaneous eruption evolving over three months, along with the clinical discovery of a left breast nodule.

Breast examination revealed a 6×4 cm hard, fixed left-sided mass without local inflammatory signs. Breast ultrasound and mammography identified a suspicious lesion in the upper outer quadrant (UOQ), classified as BIRADS 5, associated with an ipsilateral axillary lymphadenopathy. Biopsy confirmed an invasive ductal carcinoma, triple-negative (ER-, PR-, HER2-). Thoraco-abdomino-pelvic CT scan showed no metastases.

Dermatological examination showed a facial and cervicothoracic erythematous rash consistent with the V-neck sign (Fig 1), an alopecic plaque of the scalp (Fig 2), erythematous, scaly, and keratotic lesions over the metacarpophalangeal and interphalangeal joints, consistent with Gottron's papules (Fig 3). The absence of muscle weakness and normal levels of muscle enzymes (CK, AST, aldolase) suggested an amyopathic form of dermatomyositis. Immunological workup revealed positive anti-TIF1-γ antibodies, confirming the paraneoplastic origin.

The patient completed neoadjuvant chemotherapy with sequential EC-P regimen (epirubicin/cyclophosphamide followed by weekly paclitaxel), proceeding to definitive surgical management via total mastectomy with axillary lymph node dissection. Concurrently, corticosteroid therapy

and intravenous immunoglobulins were administered. Rapid improvement of the skin lesions was observed after initiation of chemotherapy. Postoperatively, corticosteroids were gradually tapered without recurrence of cutaneous symptoms or emergence of muscle weakness. Surveillance at one year confirmed sustained remission following adjuvant radiotherapy.

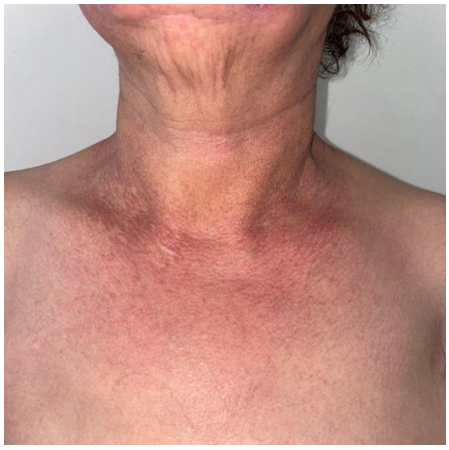

Fig 1: *V-neck sign* – Ill-defined, V-shaped telangiectatic erythematous plaque on the neck

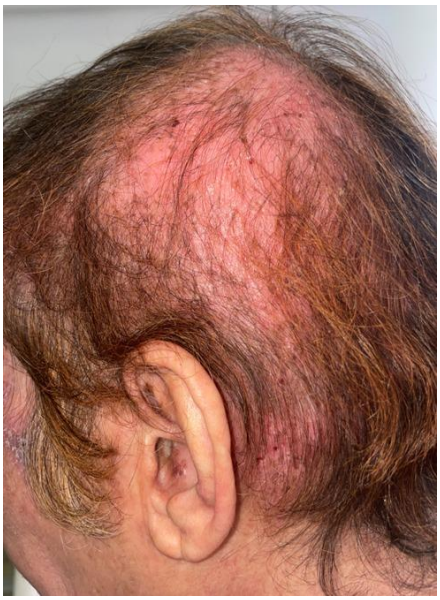

Fig 2: Alopecic plaque of the scalp with desquamation

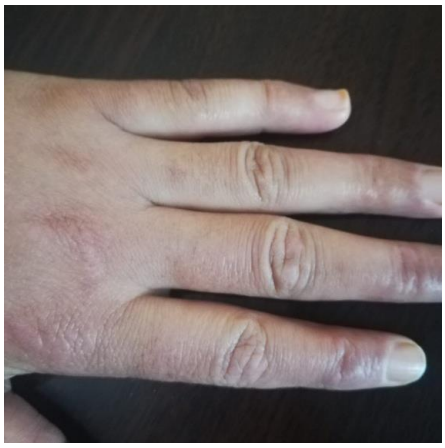

Fig 3: Gottron's papules – Erythematous, scaly, keratotic lesions over the metacarpophalangeal and interphalangeal joints

DISCUSSION

Amyopathic dermatomyositis (ADM) is a rare subtype of dermatomyositis. This idiopathic connective tissue disease is characterized by typical cutaneous findings in the absence of clinical or biological evidence of myopathy. Unlike the classic form, ADM shows no proximal muscle weakness, electromyographic abnormalities, elevated muscle enzymes, or inflammatory infiltration on muscle biopsy [4]. Classic cutaneous manifestations include Gottron's papules, heliotrope rash with periorbital edema, periungual telangiectasia, and poikilodermic eruptions [5].

Despite the absence of muscle involvement, ADM shares epidemiologic and dermatologic features with classic DM and carries a similar increased risk of malignancy, estimated at 14–28% (6,7). Patients with DM are approximately six times more likely to develop malignancies, although the mechanism remains unclear [8], with the highest risk occurring during the first year after diagnosis [9].

ADM is frequently associated with breast cancer. One review identified 88 ADM cases associated with malignancy, of which 29% involved breast cancer [10]. Unlike the classic form, in which lung cancer predominates, breast cancer is more commonly associated with ADM [7]. Invasive ductal carcinoma is the most frequent histologic type, without significant correlation to hormone receptor status or HER2 expression. Patients with ADM are often diagnosed at an advanced cancer stage [11]. ADM symptoms may precede, coincide with, or follow cancer diagnosis, and symptom recurrence may signal cancer relapse [12].

In our case, the skin lesions preceded the cancer diagnosis, confirming the paraneoplastic nature of the dermatosis.

Diagnosis relies on punch skin biopsy, which typically shows dermoepidermal junction alterations (basal keratinocyte vacuolization, interface lymphocytic dermatitis, mucin deposition, and occasionally epidermal atrophy) [13].

Detection of specific myositis-associated antibodies (MSAs), including anti-MDA5, anti-TIF1γ, anti-Mi2, anti-NXP2, and anti-synthetase antibodies, assists with diagnosis and stratifies malignancy risk [14]. Anti-TIF1 antibodies, in particular, are strongly associated with malignancy [15]. Studies suggest that the antigenic profiles of certain adenocarcinomas, especially breast cancer, resemble those of muscle cells in myositis patients, supporting the paraneoplastic immune response hypothesis [16]. Anti-TIF1γ antibodies are especially noteworthy, being linked to cutaneous-dominant and frequently amyopathic disease forms [17]. These antibodies are present in 58% of cancer-associated cases [18], and their presence is a predictive marker of breast cancer [19]. Additionally, patients over 39 years old with DM and anti-TIF1 antibodies have a significantly higher risk of breast cancer [20].

Management of DM requires a personalized approach based on disease severity, extent of skin involvement, visceral involvement, autoantibody profile, and presence of associated malignancy [21]. In amyopathic forms, first-line treatment includes synthetic antimalarials (hydroxychloroquine or dapsone). Methotrexate remains the reference second-line immunosuppressant after failure of initial therapy [23]. For refractory cases, mycophenolate mofetil or intravenous immunoglobulins may be used [22]. Systemic corticosteroids are essential in muscle involvement but are less effective on cutaneous symptoms, warranting their use at low doses in resistant forms [23]. Although current treatments are largely immunosuppressive, there is growing interest in targeted therapies such as anti-IFNβ monoclonal antibodies (e.g., Dazukibart), which have shown clinical improvement in cutaneous symptoms [24]. A phase 3 trial is underway [25]. Emerging therapies under investigation include JAK inhibitors and CAR-T cell therapies [26,27].

In TNBC, neoadjuvant chemotherapy increases pathologic complete response rates and reduces surgical burden. It is often associated with concurrent improvement in paraneoplastic cutaneous manifestations of dermatomyositis [12,28].

Currently, no official guidelines exist for managing breast cancer associated with ADM, with available data mainly derived from case reports [2]. These do not provide clear recommendations regarding immediate versus delayed surgery after neoadjuvant treatment. The role of neoadjuvant chemotherapy or hormone therapy also remains poorly defined. Surgery is usually preferred due to infection risks associated with immunosuppressive therapies. In cases of severe muscle involvement, corticosteroids should be initiated before definitive cancer surgery [29]. When symptoms are limited to skin involvement, they can often be controlled with neoadjuvant chemotherapy alone, without specific immunosuppressive therapy, optimizing preoperative conditions and reducing infectious risks. Total mastectomy is generally preferred in paraneoplastic ADM, but conservative surgery remains an option if the tumor is suitable and the patient wishes to preserve the breast [30].

In our case, the patient received corticosteroids and intravenous immunoglobulin infusions, with rapid improvement in cutaneous lesions following chemotherapy initiation. Postoperatively, corticosteroids were tapered without recurrence of cutaneous signs or emergence of muscle weakness. The parallel improvement of DM symptoms following cancer treatment supports the paraneoplastic nature of the disease.

Finally, every patient newly diagnosed with ADM should undergo specific oncologic screening, including enhanced gynecological surveillance (annual mammography, CA-125 assay, and pelvic ultrasound), particularly in those with anti-TIF1γ antibodies due to the increased risk of breast and ovarian cancer. Close oncologic follow-up is recommended for at least three years [31].

CONCLUSION

Amyopathic dermatomyositis may be an initial manifestation of breast cancer, particularly in its triple-negative form. The detection of anti-TIF1γ antibodies is a valuable diagnostic and prognostic marker. Improvement of cutaneous symptoms after cancer treatment confirms the paraneoplastic nature of the disease. An individualized, multidisciplinary approach is essential to optimize outcomes.

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