Dermatomyositis Developing Post Neoadjuvant Chemotherapy and Lumpectomy

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ABSTRACT

Dermatomyositis is an idiopathic inflammatory myopathy known to occur as a paraneoplastic syndrome. The course of dermatomyositis is commonly reported to mirror the course of the malignancy. Here, we report a case of dermatomyositis that developed in a patient after lumpectomy and completed chemotherapeutic treatment.

KEYWORDS: dermatomyositis, breast cancer, chemotherapy, paraneoplastic syndrome

INTRODUCTION

Dermatomyositis is an idiopathic connective tissue disease characterized by inflammatory myositis and characteristic skin lesions. The disease bears a strong association with malignancy: approximately 15–30% of patients with dermatomyositis have an underlying malignancy, and dermatomyositis patients have an 5–7 times increased risk of developing cancer compared to the general population.[1,2] However, numerous previous reports of paraneoplastic dermatomyositis have observed the disease evolving in parallel with cancer.[3–6] Here we present a case of dermatomyositis that developed in a patient with invasive ductal carcinoma of the breast after six cycles of neoadjuvant chemotherapy and right lumpectomy.

CASE PRESENTATION

A 48-year-old woman with a history of grade III breast cancer treated with neoadjuvant chemotherapy and lumpectomy presented with two months of facial and arm swelling, dysphagia, and erythematous rash involving the face, chest, and arms. The symptoms initially developed two weeks after her 6th cycle of taxotere, carboplatin, trastuzumab, and pertuzumab. She first visited her oncologist, who suspected her symptoms to be either a side effect of chemotherapy or an allergic reaction. At the time, she received diphenhydramine, topical steroids, and short courses of oral steroids, which provided only transient improvement in her symptoms. On admission to the hospital, she was afebrile and hemodynamically normal with bilateral arm pitting edema and an erythematous facial rash (**Figure 1**). She had full strength of all extremities. She had a WBC of 12.1 K/μL and unremarkable serum chemistries. Her presentation was thought to be suspicious for superior vena cava syndrome, but chest computed tomography (CT) showed no evidence of venous occlusion. Bilateral arm ultrasound was negative for deep vein thrombosis. Creatinine kinase was 819 IU/L (normal 20–165 IU/L), aldolase 9.1 U/L (normal ≤ 8.1 U/L), and antinuclear antibody was reactive at 1:1280 with a speckled pattern. Further evaluation with an arm MRI revealed edema with enhancement of skin, subcutaneous tissues, and multiple proximal muscles consistent with dermatomyositis (**Figure 2**). A skin biopsy was consistent with dermatomyositis, with vacuolated immune complex deposition at the dermal-epidermal junction. Muscle biopsy showed

Figure 1. Facial rash with prominent periorbital edema.

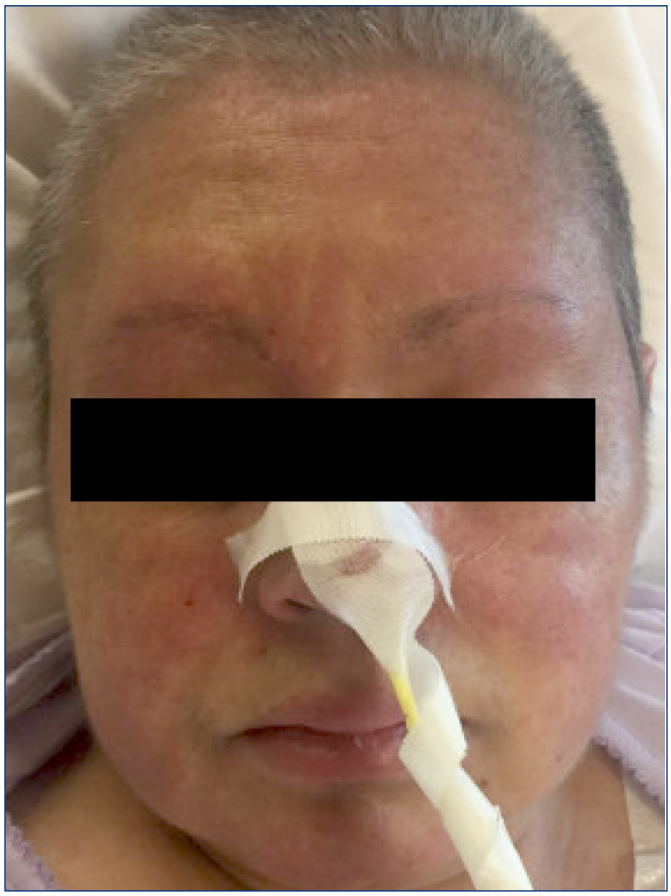
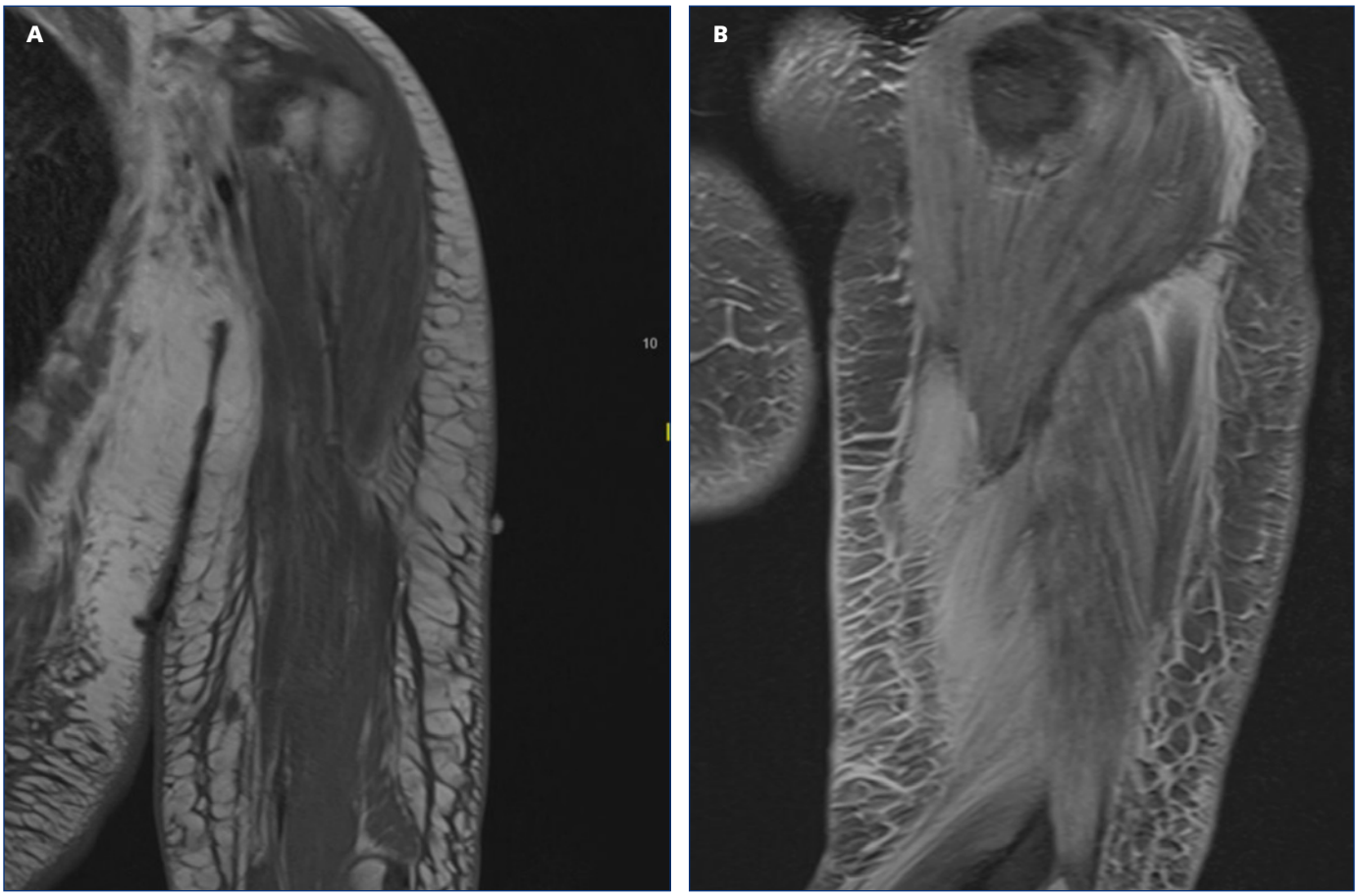


Figure 2. MRI of the humerus precontrast (**A**) and postcontrast (**B**) demonstrating diffuse edema and abnormal soft tissue enhancement.



perivascular and interstitial lymphocytic infiltration, indicating an inflammatory myopathy. The patient was started on azathioprine and a prednisone taper, and her symptoms improved over several months.

DISCUSSION

Dermatomyositis is an idiopathic inflammatory myopathy with classic cutaneous manifestations, including Gottron's papules, heliotrope rash, and the shawl sign.[7] The condition has a strong association with malignancy, with lung, ovarian, and breast being the most common types.[18] Out of the dermatomyositis cases diagnosed in breast cancer patients, 71% present with stage III or IV breast cancer.[9] Recent work shows that the underlying mechanism of dermatomyositis is likely immunological, with over 70% of patients found to have disease-specific autoantibodies.[10] Diagnosis is typically made with muscle biopsy, electromyography, and muscle magnetic resonance imaging. Glucocorticoids are considered the mainstay of management. Steroid-sparing immunosuppressive therapies confer therapeutic benefit in addition to relieving steroid burden, and intravenous immunoglobulin is used in severe cases.[11] If cancer is present, cancer cure is expected to lead to symptomatic improvement. Many have reported that with complete remission of cancer, the physical manifestations of dermatomyositis either diminish or disappear completely.[3,5,6] In a study by Cox et al, dermatomyositis patients who underwent resection of tumors were found to have resolution of their symptoms.[6]

Treatment is monitored clinically for improving muscle strength and resolution of cutaneous symptoms. A fall in creatinine kinase levels is generally seen initially in the recovery course, followed by a more gradual return of muscle power.[12]

This case displays an unusual presentation of dermatomyositis that evaded immediate diagnosis due to a few atypical characteristics. She had no weakness and the serum elevations of muscle enzymes was modest. Most accounts of malignancy-associated dermatomyositis describe the two disease processes occurring simultaneously. Interestingly, this patient presented with symptoms after six cycles of neoadjuvant chemotherapy and tumor excision. The patient had physical exam findings that did not bear similarity to the typical cutaneous findings associated with dermatomyositis. Indeed, limb edema is a rare finding in adult dermatomyositis, and Milisenda et al reported 14 only cases up until 2014.[13] Her symptoms appeared resistant to repeated courses

of steroids and developed two weeks after a cycle of chemotherapy. For these reasons, the diagnosis of dermatomyositis was not immediately entertained. The patient's presentation initially raised the possibility of angioedema, allergic reaction, steroid-induced myopathy, or a previously undiagnosed autoimmune process unmasked by chemotherapy.

CONCLUSION

This case reinforces the importance of maintaining a high suspicion for dermatomyositis in a patient with cancer history presenting with widespread skin rashes, even in the setting of completed treatment. Though paraneoplastic dermatomyositis is known for its temporal association with malignancy, a presentation such as this one demonstrates an atypical time course.

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