Breast Cancer Associated Dermatomyositis: Parallel Occurrence, Remission, and Relapse of the Two Diseases in a Patient: A Case Report

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ABSTRACT

Dermatomyositis (DM) is an uncommon inflammatory disease with characteristic cutaneous findings associated with muscle weakness. Although the majority of cases of DM is idiopathic, an increased incidence of underlying malignancy in patients with dermatomyositis has been reported. An underlying breast cancer is found in nearly 10% of cases of malignancy-associated DM. We report a case of a 50-year-old nulliparous woman who was diagnosed with dermatomyositis according to Bohan & Peter criteria. After performing a cancer screening which was negative, the patient was treated with topical and oral corticosteroids; then she was put under immunosuppressors with minimal improvement. One year later, a right breast mass was detected at systemic clinical examination prompting diagnostic workup of breast cancer. After mammogram which showed a suspicious mass, the patient underwent stereotactic breast biopsy which revealed an invasive ductal carcinoma. Computed Tomography (CT) scan of the chest, abdomen, and pelvis and bone scan revealed no distant metastases. The patient underwent mastectomy and axillary dissection followed by adjuvant chemotherapy, radiotherapy, and adjuvant tamoxifen. Simultaneously, she continued the treatment for her Dermatomyositis. Her cutaneous and musculoskeletal symptoms improved dramatically following the treatment of her breast cancer. After four years of uneventful follow-up, the patient reported the recurrence of symptoms suggestive of dermatomyositis. A thoracic-abdominal-pelvic CT scan has been performed and revealed distant metastases. Therefore, our case demonstrates the parallel evolution between dermatomyositis and breast cancer after surgery, local radiotherapy and systemic therapy.

Keywords: Dermatomyositis, Breast cancer, Relapse, Metastases.

1. INTRODUCTION

Dermatomyositis (DM) is a rare disease which includes an inflammatory myopathy and cutaneous manifestations. Although the etiology of dermatomyositis remains unknown and the disease is considered idiopathic in the majority of cases, an association with histocompatibility antigens, environmental agents and autoimmunity have been reported¹. Moreover, an increased risk of malignancy has been demonstrated in adult patients with dermatomyositis (¹,²,³). An underlying breast cancer was found in nearly 10% of cases of malignancy-associated DM and may precede the occurrence of the DM by months, may be discovered concurrently with the DM, or may occur several months to years after the DM is diagnosed⁴.
2. CASE REPORT

A 50-year-old nulliparous perimenopausal woman with no known history of neoplasia and no contributory family history presented to dermatology department with complaints of skin rash associated with progressive fatigue, upper extremity muscle weakness, and arthralgia. On physical examination, the patient found to have a purplish rash on the face, neck, shoulders and chest. There was no detected breast mass during this first clinical examination. Laboratory investigations showed an elevated muscle enzyme levels: creatine kinase (CK) of 1270UI/l, aspartate aminotransferase (AST) of 100UI/l and lactic dehydrogenase (LDH) of 570UI/l. However, connective tissue serologies including antinuclear antibodies and Jo-1 antibodies were substantially normal. Electromyogram (EMG) revealed proximal inflammatory myopathy which was strongly suggestive of DM. Cutaneous and muscular biopsies were then performed, and histopathological examination confirmed the findings of inflammatory myopathy. According to Bohan & Peter criteria, the diagnosis of DM was, therefore, definite. In order to detect an underlying malignancy, a cancer screening including complete blood count and tumor markers such as cancer antigens (CA 125 and CA 15-3) and carcinoembryonic antigen (CEA) Chest X-ray, abdominal and pelvic ultrasound and mammogram, was performed but was negative. The patient was therefore treated with topical and oral corticosteroids; then she was put under immunosuppressors with minimal improvement. One year later, a right breast mass was detected at systemic clinical examination prompting diagnostic workup of breast cancer. After mammogram which showed suspicious mass classified BI-RADS 4 associated with an ipsilateral axillary lymphadenopathy, the patient underwent stereotactic breast biopsy which revealed an invasive ductal carcinoma grade I. Breast cancer staging workup including Computed Tomography (CT) scan of the chest, abdomen, and pelvis, and bone scan showed no distant metastases. The patient underwent mastectomy with axillary dissection. Histologic examination of mastectomy specimen confirmed the findings of an invasive ductal carcinoma grade I, measuring 3cm in the long axis diameter associated to three invaded lymph nodes among thirteen lymph nodes dissected. Therefore the tumor was classified pT2N1. On immunochemistry analysis, the tumor was positive for hormone receptors and negative for a human epidermal growth factor receptor 2 (HER2), and the Ki67 level was at 30%. Such results warranted adjuvant therapy. Thus, surgery was followed by six cycles of adjuvant chemotherapy, hypofractionated radiotherapy to the chest wall and supraclavicular lymph nodes with a total dose of 42 Gy at 2.8 Gy per fraction in 15 sessions and adjuvant tamoxifen. Simultaneously, the patient continued the treatment for her Dermatomyositis. The evolution was marked by the dramatic improvement of cutaneous and musculoskeletal symptoms following the treatment of breast cancer. The patient was put then on a close follow-up schedule in both of dermatology department and radiotherapy department. After four years of uneventful follow up with complete remission, the patient reported the recurrence of skin rash (Figure 1) associated with muscle weakness.

Fig. 1: Skin rash during dermatomyositis relapse

Moreover, laboratory examination revealed high muscle enzyme levels. Such symptoms and biochemical results were suggestive of dermatomyositis relapse. Metastatic workup for breast cancer (including brain CT scan, thoracic-abdominal-pelvic CT scan, and bone scan) was therefore performed and showed pulmonary metastases (Figure 2).
As the patient was put on an aromatase inhibitor hormone therapy, the evolution was marked by stability in the size of lung nodules with the improvement of cutaneous and muscular symptoms.

3. DISCUSSION

Dermatomyositis (DM) is an uncommon inflammatory myopathy with specific cutaneous findings, such as heliotrope rash and Gottron’s papules\(^{(1,4)}\). Bohan & Peter\(^{(5)}\) defined DM according to the presence or absence of five major criteria for diagnosis consisting of typical dermatologic features, symmetrical proximal myopathy, elevated muscle enzyme levels, abnormal electromyogram and inflammatory myositis in muscle biopsy. Actually, according to Bohan & Peter, “definite DM” corresponds to the presence of three or four criteria plus the rash, “probable DM” corresponds to the presence of two criteria plus the rash, whereas “possible DM” consists of the presence of one criterion plus the rash\(^{(5)}\). Dermatomyositis affects adults and children alike. In adults, the average age at...
Diagnosis is 40, and almost twice as many women are affected as men\(^1\). It has been reported an increased incidence of malignancy in patients with DM. Thus, the prevalence of cancer in patients with DM ranges from 9.4% to 30%\(^6\). Furthermore, breast cancer is the third most frequent malignancy in women with DM, and it is found in nearly 10% of cases of malignancy-associated DM\(^2,3,4,7\). In the majority of reports in the literature, most cancer cases were detected within the first year after DM diagnosis\(^3\). Thus, early diagnosis of dermatomyositis and performing the extension study probably contributed to the earlier diagnosis of the tumors\(^8\). However, malignancy preceding myopathy by two years, or occurring even after five years of disease have also been described\(^6\). Moreover, dermatomyositis can also present as a sign of recurrent malignancy or progression by patients with previous cancer in their medical history\(^9\). Independent risk factors for an underlying tumor have been reported, including an abrupt and rapid onset of the skin lesions and muscle weakness, necrotic lesions and periangual erythema, persistently high erythrocyte sedimentation rate (ESR), and the presence of myositis-specific autoantibodies (anti-p155 or anti-p155/p140 antibodies)\(^6,8\). The identification of predictive factors for malignancy in adult dermatomyositis will make it possible to more accurately select those patients in whom an exhaustive search for a tumor is of the highest priority\(^8\). With regards to age, only a few reports have focused on the age effect of cancer risk among DM patients. This age effect remains still controversial\(^5\). While Hill et al.\(^2\) indicated increased risks of cancer among DM patients younger than 44 years and older than 45 years, Stockton et al.\(^7\) indicated that cancer risk decreases with age in DM patients. In contrary, Chen et al.\(^3\) demonstrated that the relative risk of cancer in DM remains high in every age group, the fact which highlighted the true link between DM and malignancies despite the age effect. In a recent retrospective study, Souza et al.\(^6\) reported that age at the time of disease diagnosis was a predictive factor of the development of malignancy in DM patients and they suggest that DM initiating at advanced age should be considered when a more extensive assessment for cancer is recommended. Regarding pathogenesis of dermatomyositis related to underlying malignancies including breast cancer, it remains unclear. Several hypotheses have been proposed, including immune complex, complement system, cell-mediated immunity abnormality, infectious diseases, and HLA type\(^4\). Regarding treatment, no standard treatment regimen has been established yet regarding dermatomyositis associated with malignancy\(^9\). While systemic corticosteroids continue to be the treatment of choice in patients with dermatomyositis, other immunosuppressive therapy should be considered if no improvement in objective muscle strength occurs after three months of corticosteroid therapy\(^1,8\). Furthermore, treatment of breast cancer-associated DM involves standard breast cancer therapies including surgery, chemotherapy, radiotherapy and hormone therapy according to the staging and molecular classification of the breast cancer. Actually, it has been noted that DM improved after treatment of cancer\(^9\). In our case, complete remission of DM has been observed after breast cancer therapy, but DM symptoms have flared up simultaneously with the occurrence of breast cancer metastasis. Such observation suggests the paraneoplastic character of DM described in our case.

**4. CONCLUSION**

Dermatomyositis is an inflammatory disease which could reveal primitive malignancy including breast cancer or metastatic relapse. Parallel evolution observed between DM and cancer suggests the paraneoplastic character of DM and the necessity of performing a cancer screening and metastatic work up respectively whenever DM occurs or flares up after malignancy removal.
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