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CONCURRENT DERMATOMYOSITIS AND METASTATIC BREAST CARCINOMA:

Case Report

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Simultaneous occurrence of neoplasia and dermatomyositis was first reported by Stertz in 1916[1]. He described the case of a 55-year-old male with dermatomyositis, who subsequently was found to have carcinoma of the stomach. In 1935, the relationship again was mentioned by Bezecny[2], who reported three cases of dermatomyositis associated with tumor.

Various, and occasionally conflicting, statements have been made regarding the incidence of coexisting dermatomyositis and malignant tumors. In a review of 590 cases of dermatomyositis reported in world literature, Williams[3] found 92 patients (over 15%) with concurrent malignant tumors.

Dermatomyositis occurs most frequently in the younger population, particularly in the second and third decades.[4] When dermatomyositis exists in middle-aged and elderly people, the incidence of concurrent neoplasia increases with each decade. Thus, Shy[5] found cancer and polymyopathy associated in 5% of patients under 50 and 39% of those over 50. The youngest patient reported was an 11-year-old girl with a chromophobe adenoma[6]. In patients over 50, the frequency of polymyositis with malignancy was 71% in males as compared to 24% in females. Pearson[7] later pointed out that dermatomyositis, unassociated with malignancy, affected females twice as often as males; but the syndrome of dual disorders was observed nearly three times more often in men than in women.

Occurrence of dermatomyositis with malignant tumors has been described with various primary neoplasms, the most common being breast, stomach, and lung malignancies. Carcinomas of most sites of the body have been reported in the combined occurrence, with the exception of thyroid and testicular neoplasms[4].

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Case Report

A 44-year-old-Caucasian female underwent a right radical mastectomy and bilateral salpingo-oophorectomy in October 1960, because of mass in her right breast, which proved to be adenocarcinoma. Right axillary node recurrence three years later was treated with radiation therapy.

In October 1965 she developed nausea, vomiting, and generalized weakness, for which no definite cause was found. Radiological survey of the skeleton did not demonstrate metastatic diseases. Peripheral blood smear contained 18% segmented neutrophils, 15% lymphocytes, 10% monocytes, and 27% eosinophils.

In December 1965 the patient developed an erythematous rash on her malar eminences, hands, neck, thighs, as well as marked proximal thigh weakness.

On January 18, 1966, physical findings in this acutely ill patient were these: a temperature of 101°F; a scaling, erythematous eruption across the malar eminences; erythematous, bullous lesions on the chest and lateral aspect of thighs; a heliotrope appearance of the para-ungual skin of hands; a 2 x 2 cm. mass in the medial superior portion of the left breast, and a 1 x 2 cm. lymph node in the left axilla. Weakness of pelvic girdle and quadriceps muscle was present, and severe.

Laboratory findings were as follows: hemoglobin 8.3 g.%; WBC 3,100 cells cm.^{3}, with a normal differential count; normal urinalysis sedimentation rate of 51 mm./hr.; SGOT 43 units; latex fixation test was weakly reactive; normal alkaline phosphatase; BSP negative; urinary creatinine-creatine ratio was 2:1; C reactive protein was positive to a dilution of 1:8. At this time, skeletal roentgenograms revealed destructive lesions in the skull, ribs, and pelvis.

The patient's symptom complex was felt to represent adenocarcinoma of the breast with associated dermatomyositis. A skin and muscle biopsy of the left thigh revealed pathological changes in both skin and muscle consistent with the diagnosis of dermatomyositis.

A left simple mastectomy was performed, and confirmed the diagnosis of adenocarcinoma; but it could not be determined whether this lesion was primary or metastatic.

The day following the procedure, the patient showed a marked increase in the inflammatory appearance of her cutaneous lesions, with new bullous formation. The skin lesions and muscle weakness responded rapidly and favorably to high doses of corticosteroids.

In an attempt to palliate the extensive metastatic breast cancer, a hypophysectomy was performed two weeks later. No effect was noted on the rapid progress of the patient's metastatic disease. Moderately severe diabetes insipidus developed necessitating large doses of intramuscular pitressin for control. Corticosteroids continued to be required because of the dermatomyositis.

Six weeks later, the patient developed the clinical picture of obstructive jaundice, and at laparotomy a metastatic tumor mass was found in the porta hepatis. A cholecystostomy was unsuccessful in decreasing the severe jaundice, and she expired on the fifteenth postoperative day. Autopsy confirmed the presence of extensive metastatic carcinoma.

Discussion

Muscular weakness is a common symptom of dermatomyositis, occurring in 95% of patients. Even though the skin manifestations may precede myositis, diagnosis cannot be established with certainty, without both skin and muscle findings. As shown in this case, usually the first symptoms are weakness in the proximal muscle groups. Although muscular atrophy may be present, the degree of weakness is disproportionately greater than the atrophy.

The most helpful laboratory tests are those which reflect tissue destruction — such as serum aldolase, creatine phosphatase, transaminases, and lactic dehydrogenase.[7] The urinary creatinine-creatine ratio usually is less than 10:1. Eosinophilia, as described in this patient, has been reported by Dostrovsky.[8]

Skin changes in the disease vary, ranging from a local or diffuse erythema to a scaling eczematoid or exfoliative dermatitis. When no skin lesions are present, the disorder is called polymyositis.

Frequently, polymyositis has been confused with the muscular dystrophies.[9] Dowben *et al.*[10] maintain that the clinical features most helpful in the differential diagnosis are the presence of cutaneous lesions, muscular pain or tenderness, and dysphagia. Diagnostically, subjective decrease in muscle weakness and improvement in appearance of skin lesions, with corticosteroid therapy, are felt to be important in dermatomyositis. Although these features do not always occur in dermatomyositis associated with malignancy, they may be present, as in the case being reported. Dermatomyositis associated with malignancy is exceedingly rare in young patients.

The etiology of dermatomyositis remains obscure. Diverse factors, such as infection, endocrine disorders, allergy, and aberrations of Vitamin E metabolism have been suggested as possibly etiologic in the pathogenesis of the disorder. Many investigators now feel that the syndrome represents an autoimmune process.

Because of the high incidence of coexisting malignancy and dermatomyositis, it is apparent the relationship is more than coincidental. In 1935, Bezecny[2] reported improvement in dermatomyositis, following primary therapy of the malignancy. Sheard and Knoepfler[11] reported a patient with metastatic breast cancer and dermatomyositis. After hypophysectomy the patient experienced a regression of the breast cancer, with subsequent improvement in dermatomyositis. Unfortunately, such was the the case in our patient. Curtis[12] in 1952 suggested that tumors might be antigenic. Williams

in 1959[3] in a literature review further affirmed this hypothesis. He found evidence of improvement of the dermatomyositis in nine patients, after primary surgical or radiological therapy of the associated tumor.

In 1959 Grace and Dao[13] intradermally injected extracts of removed tumor and normal tissue into a patient with concurrent dermatomyositis and adenocarcinoma of the breast. A marked reaction occurred at the injection site of the tumor tissue extract while only a minimal reaction occurred at the site of the normal tissue. They also demonstrated, by passive transfer studies, that the reaction was specific for the tumor tissue extract.

The tumor antigen itself is thought to be a polysaccharide[14]. Thus, it is possible that muscle and skin may be the "shock organs" in an autoimmune process.

In the present case, postoperative flare of the patient's skin lesions perhaps can best be explained by the sudden release of large quantities of antigenic material, caused by manipulation of the tumor during surgery.

Summary

The case of a 44-year-old woman with concurrent dermatomyositis and metastatic breast carcinoma is presented. Incidence, factors to be considered in diagnosis, and a possible explanation of the relationship of dermatomyositis and malignancy are discussed.

Because of the high incidence of malignancy in older patients with dermatomyositis, all patients over 50 with dermatomyositis or polymyopathy should be suspected of having an associated malignancy. The theoretical explanation for the increased incidence of dermatomyositis with malignancy is an autoimmune process, with the antigens derived from the neoplastic tissue.

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