A Case of Pancreatic Cancer Presenting as Dermatomyositis

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Dermatomyositis is an idiopathic, inflammatory myopathy characterized by proximal muscle weakness and cutaneous lesions. The association of malignancy with dermatomyositis is well established, especially with lung, breast, ovary, stomach, and colorectal cancers. The incidence of cancer appears to be increased especially in elderly persons, and the prognosis is very poor. Malignancy may occur before the onset of dermatomyositis, concurrently, or afterward. Therefore extensive screening tests for occult malignancy should be conducted in patients with dermatomyositis. We report a 76-year-old man presented with dermatomyositis and a search for possible occult malignancy found an otherwise asymptomatic pancreatic adenocarcinoma with massive lymph node metastasis.

Key Words: Dermatomyositis, Pancreas, Malignancy

INTRODUCTION

Dermatomyositis is an idiopathic inflammatory myopathy involving striated skeletal muscle with characteristic skin manifestations. Dermatomyositis demonstrates cutaneous features including erythema on the dorsal surfaces of the phalangeal joints (Gottron's sign), violaceous rash involving periarticular area (heliotrope rash), and poikiloderma (the combination of atrophy, dyspigmentation, and telangiectasia). Increased incidences of cancers in patients with dermatomyositis have been reported in a number of studies. Dermatomyositis often heralds paraneoplastic manifestations in which the clinical features can either accompany the malignancy or appear after and even before detection of the tumor. Therefore the evaluation for the presence of occult malignancy is important. Lung, breast, and gastrointestinal tract cancers are commonly detected malignancies. However, the association between dermatomyositis and pancreatic cancer is not common. According to a recent population based study of patients with biopsy proven inflammatory myopathy, of whom 104 were found to have malignancy, only one case of pancreatic cancer was identified. We described a patient with pancreatic cancer who presented with features of dermatomyositis.

CASE REPORT

A 76-year-old man was admitted to the hospital because of progressive muscle weakness and pruritic skin rash. He has been well until two months earlier, when he had developed purple-colored eruption on the trunk. Few weeks later, erythematous rash on the forehead and eyelids were developed. Despite treatment with oral antihistamine and corticosteroid ointment, skin lesions got worse and more extending to the extremities. Two weeks prior to admission, progressive muscle weakness particularly on both arms developed. Past medical history was unremarkable except for renal cyst detected 1 year ago. On physical examination, body temperature was 37°C, heart rate 75/min, and blood pressure 120/80 mmHg. Finally scalp plaques were present in a shawl pattern over the upper back and chest (Fig. 1). Faint violaceous macules around eyelids consistent with a heliotrope rash and Gottron's papules on the extensor surfaces of fingers were also found. The neuromuscular exam revealed moderate weakness of the shoulder...
Laboratory tests revealed hemoglobin 11.3 g/dL, white blood cell 5,400/mm$^3$, with a differential of 60% neutrophil, 21% lymphocyte, and platelet 163,000/mm$^3$. Blood chemistry showed protein 5.9 g/dL, albumin 2.7 g/dL, bilirubin 0.8 mg/dL, AST 134 IU/L, ALT 70 IU/L, alkaline phosphatase 81 IU/L, creatinine 1.2 mg/dL, CPK 823 IU/L (normal range 55-227 IU/L), and LDH 964 IU/L (normal range 218-472 IU/L). Thyroid function test was normal. Serum antinuclear antibody was positive at a titer of 1:320 with a homogenous pattern. The results of the serologic profiles including anti-Jo-1, anti-dsDNA, anti-Smith, anti-histone, anti-RNP, anti-SSA/Ro, anti-SSB/La, and anti-Scl 70 were all negatives. Complement component of C3, C4 and CH50 hemolytic activity were normal range. EMG disclosed decreased amplitude and duration of motor unit action potential and increased insertional activity and fibrillation, consistent with myopathy. The histology of the rectus femoris muscle showed mild atrophy with degenerated necrotic muscles and lymphocytic endomysial infiltration (Fig. 2). These findings confirmed a diagnosis of dermatomyositis. Therapy with 60 mg/day of prednisolone was started. After that skin rash and muscle weakness are improved moderately and the CPK levels are decreased to 67 IU/L. Search for occult malignancy was carried out. Chest X-ray and gastroscopy were unremarkable. Colonoscopy revealed multiple colonic polyps, of which were biopsied and diagnosed as tubular adenoma. The levels of tumor markers were CA19-9 331 U/ml (normal range 0–37 U/ml), CEA 10.6 ng/ml (normal range 0–5 ng/ml), AFP 21.5 ng/ml (normal range 0–8 ng/ml), PSA 0.7 ng/ml (normal range 0–4 ng/ml). Abdominal CT scan showed mass on the pancreatic head portion with enlarged lymph nodes in porta hepatis, portocaval and paraaortic space and multiple renal cysts also found (Fig. 3). Aspiration histologic exam was performed, and diagnosed poorly differentiated adenocarcinoma of pancreas with lymph node metastasis. The patient underwent gemcitabine-based systemic chemotherapy.
The association of cancer with inflammatory myopathy has been investigated in a number of studies. The frequencies of cancers in patients with inflammatory myopathy have ranged from 6 to 60%\(^2\)-8). The increased incidence of malignancy was found in both polymyositis and dermatomyositis, although the relative risk was higher in dermatomyositis\(^3,5\). Hill et al. reported 13% of dermatomyositis patients developing cancers, the majority of cases occur within 2 years of diagnosis\(^4\). The relative risk of cancer is 2.4 in men and 3.4 in women patients with dermatomyositis in comparison with general population\(^6\). Cancer may occur before the onset, concurrently, or after the development of dermatomyositis. The malignancies frequently associated with dermatomyositis are ovary, lung, stomach, colon, prostate, breast, bladder, nasopharynx and lymphoma\(^5,8\). However, the association between dermatomyositis and pancreatic malignancy is not common.

The search for the presence of occult cancer in patients with dermatomyositis is important. All suggestive symptoms or signs should be evaluated thoroughly. Investigations should include CBC, blood chemistry, urinalysis, stool exam, chest X-ray, abdominal and pelvic ultrasonography, gastroscopy, mammography in women to detect the commonly associated malignancies. The differences in clinical features of cancer-associated dermatomyositis have been reported. These patients are more likely to have normal CPK level and digital vasculitis, and less likely to have myositis-specific autoantibodies than those without cancer\(^9\). Cutaneous necrosis and an increased ESR have also been proposed as predictive signs of malignancies\(^8,10\). But the severity of the myositis is not associated with the occurrence of malignancy\(^11\). In a retrospective study of 79 patients with polymyositis or dermatomyositis, Marie et al. reported that the frequency of malignancy was higher in patients older than 65 years compared with younger patients. About half of malignancies were colon cancers in elderly patients. Therefore, lower gastrointestinal tract investigations are needed in elderly patients. And elderly patients had more frequent hypoproteinemia, hypoalbuminemia and anemia\(^12\).

The exact pathogenesis of dermatomyositis is still unknown. It has to be considered an autoimmune disorder in which genetic and environmental factors seem to play a role. Various hypotheses have been suggested; a direct toxic effect on the muscle fibers of a substance released from the tumor cells, a hypersensitivity response caused by an error in the immune mechanism, a single immune response affecting both the tumor and muscle fibers, and the malignancy and dermatomyositis being independent effects of a single causative agent\(^13\). There is some evidence to suggest that both humoral and cell-mediated immune mechanisms are involved\(^13,14\). Early studies showed that some patients have serum antibodies cross-reactive with muscle components; abnormal lymphocyte transformation tests to tumor and muscle cultures\(^15\).

Treatment regimens include corticosteroid, azathioprine, cyclophosphamide, plasmapheresis, and intravenous \(\alpha\)-globulin. Improvement of dermatomyositis with successful anticancer therapy has been noted, with flare of muscle weakness associated with relapse or progression of malignancy, further suggesting a paraneoplastic origin\(^1\). However, dermatomyositis may not respond to appropriate cancer therapy, even worsening may occur.

We report the case of pancreatic cancer presenting as dermatomyositis. We suggest that pancreatic cancer should be borne in mind in patients with dermatomyositis.

**REFERENCES**


