Dermatomyositis-polymyositis and malignancy
Is there a direct relation?

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Summary
An association between dermatomyositis-polymyositis and malignancy has been reported. However, controversy exists regarding a direct causal relationship between the two diseases. We describe a patient who had the concurrent onset and parallel clinical course of dermatomyositis and malignancy, and discuss the possible mechanisms for the association of the two disorders.

Key words: dermatomyositis, polymyositis, malignancy

Introduction
The relationship of dermatomyositis-polymyositis (DM/PM) to internal malignancy has been reported in many studies, although reported incidence figures of malignancy vary widely. However, controversy exists regarding a direct causal relationship, because the temporal relationship of these diseases and cancer as well as their clinical course are quite variable. We describe a patient, in whom malignancy occurred at the same time as the DM, and DM improved after removal of the tumor, and discuss the possible mechanisms for the association of the two disorders.

Case Report
A 26-year-old man was admitted to Nippon Medical School Hospital on March 9, 1992 because of progressive muscle weakness accompanied by skin rashes. The patient had been well until two months earlier, when he began to have repeated episodes of abdominal pain and distension, and noted skin rashes on the upper extremities, which spread to the neck, face and back over the weeks. By one month later, he had difficulty climbing stairs which gradually increased in severity. His past and family histories were unremarkable.

Body temperature was 37.2°C, pulse, 84/min, respiratory rate, 16/min, and blood pressure, 138/70 mmHg.

On physical examination, an erythematous, scaly and macular rash was present in the forehead, upper eyelids, malar area, neck, and extensor surfaces of the extremities (Photo. 1). There was no peripheral edema. A grade 2/6 systolic murmur was heard in the 2nd and 3rd intercostal space. The breath sounds were normal and there were no crackles or wheezes. A firm nontender mass, 4 cm in diameter, was palpated in the right upper abdominal quadrant.
An erythematous and macular rash was present in the forehead, upper eyelids, malar area and neck (A), as well as papular erythema over the nuckles (B).

Microscopic section of the muscle biopsy specimen, showing segmental necrosis within muscle fibers, and interstitial round cell infiltration.

Barium examination of the colon, showing an "apple-core" tumor of the proximal portion of the transverse colon (arrows).

Neurologically, the muscle strength of the triceps brachii, deltoid, supraspinatus, infraspinatus, rectus abdominis, iliopsoas, quadriceps femoris, hamstrings muscle groups was graded as 3 to 4/5. The rest of the neurological examination revealed no abnormalities.

Urinalysis was negative. Stool specimens were positive for occult blood. The hematocrit was 35.8%, the white-cell count, 5,400/mm³, the platelet count, 355,000/mm³, and the erythrocyte sedimentation rate, 8 mm/hr. The serum aspartate aminotransferase was 96 IU/l, the lactate dehydrogenase, 781 IU/l (normal, 195 to 365), the creatine kinase, 731 U/l (normal, 35 to 200), and the aldolase, 21.5 IU/l (normal, 1.7 to 5.7). An electrocardiogram as well as an X-ray film of the chest were within normal limits. The electromyogram revealed a myogenic pattern. Microscopic
examination of a muscle biopsy specimen obtained from the rectus muscle showed histological changes compatible with polymyositis (Photo. 2). A barium-enema examination disclosed an "apple-core" tumor of the proximal portion of the transverse colon, just distal to the hepatic flexure. (Photo. 3). Fiberoptic colonoscopy confirmed the findings revealed by the barium examination of the colon. Histological examination of the biopsy specimens taken from the colonic tumor showed a moderately differentiated adenocarcinoma (Photo. 4). A diagnosis of dermatomyositis associated with adenocarcinoma of the colon was made, and a transverse colectomy was performed. The postoperative course was uneventful. Following successful surgical treatment of the cancer, there was a partial improvement in muscle strength and the creatine kinase level was normalized (Fig. 1). After prednisolone was begun in a dose of 40 mg a day, muscle strength improved further, the aldolase level was normalized, and skin rash ameliorated. The shaded areas are normal range.

Discussion

Whether there is a causal relationship of DM/PM to internal malignancy remains controversial, because the temporal relationship between the onset of DM/PM and the discovery of malignancy as well as their clinical courses vary widely2-3). It has been shown that one third of the patients with both DM/PM and malignancy had their malignancy preceding the onset of DM/PM, another one third had a concurrently diagnosed malignancy, and the remaining one third had a malignancy after DM/PM was diagnosed2,5-7). There have been several reports of improvement in myositis and/or skin rash following successful treatment of malignancy, and relapse of DM/PM with recurrence of the tumor2-8), although this beneficial effect of tumor therapy is not consistent9). Callen mentioned that the occurrence of DM/PM and malignant disease may be coincidental when DM/PM either precede or follow the cancer by many years, or DM/PM neither

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Photo. 4 Microscopic section of the tumor of the colon, showing a moderately differentiated adenocarcinoma.

Fig. 1 Clinical course. Following successful surgical treatment of the cancer, there was a partial improvement in the muscle strength and the creatine kinase level was normalized. After prednisolone was begun, the muscle strength improved further, the aldolase level was normalized, and the skin rash ameliorated. The shaded areas are normal range.
improve after removal of the tumor nor relapse with reappearance of the tumor. On the other hand, the concurrent onset and parallel clinical course of DM/PM and malignancy may share a causal relationship. In our patient, adenocarcinoma of the colon was detected when the diagnosis of DM was established. In addition, improvement in symptoms of myositis and objective muscle findings followed successful surgical treatment of the malignancy. Thus, our case strongly suggested a causal relationship between the two conditions.

Although the precise mechanisms for the association of DM/PM and malignancy are not clear, there are several possible explanations. First, an underlying malignant disease may trigger the expression of DM/PM by an immunological reaction to the tumor. There may exist a genetic predisposition in the patient to exhibit characteristic cutaneous changes in the presence of various kinds of tumors. Second, DM/PM and associated malignancy may share a common pathogenesis resulting from abnormal immune function. The cause of DM/PM is not well understood, but alterations of both cellular and humoral immune mechanisms have been suggested to play a major role in their pathogenesis. Callen and associates speculated that the two disorders could stem from the common denominator of an altered immune system, resulting in a hampered ability to suppress and eliminate the emergence and proliferation of neoplastic cells.

In summary, we have described a patient who had the concurrent onset and parallel clinical course of dermatomyositis and carcinoma of the colon, and discussed the possible mechanisms for the association of the two disorders.

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References


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