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Dermatomyositis Relapse Complicated with Gastric Carcinoma and Lupus Nephritis Five Years after the Initial Diagnosis of Dermatomyositis

Key words: overlap syndrome, collagen disease, malignant disease, glomerulonephritis

Dermatomyositis (DM) is frequently associated with malignant disease. In Japan, about 30 percent of all cases with DM are co-inflicted with a malignancy (1). DM patients with associated malignancies generally do not suffer complications with other collagen diseases (2). Some papers (2–4) reported that DM with malignancy or other collagen diseases responds poorly to corticosterone and has a poor prognosis. We report a relapsed case of DM complicated with both gastric carcinoma and lupus nephritis that responded well to corticosterone five years after the initial diagnosis of DM.

A 53-year-old man was admitted to our hospital on April 6, 1995, because of proximal muscle pain and weakness. At physical examination we found truncal and symmetrical proximal muscle weakness; and irregular erythema on his face, back, thorax, shoulders, and upper arms. Gottron's sign and heliotrope rashes were not found, but biopsy of erythematous tissue from his back showed liquid degeneration and perivascular infiltration of inflammatory cells.

Electromyography showed the presence of positive sharp waves, fibrillations, and polyphasic high-frequency discharge. We diagnosed the condition as DM based on the patient's muscle weakness, electromyography, skin biopsy, and elevated serum enzyme levels (5): creatine kinase (CK) 8,747 IU/l (normal 52–239 IU/l), lactate dehydrogenase (LDH) 2,323 IU/l (250–452 IU/l), myoglobin 6,880 ng/ml (<50.0 ng/ml). Although anti-Jo-1 antibody was positive, muscle biopsy of the quadriceps femoris did not show specific findings. Antinuclear antibody (ANA) and anti-DNA antibody were negative.

His symptoms improved after steroid pulse therapy and oral prednisolone. He was discharged on July 8, 1995, and oral prednisolone was discontinued in November 1996 after remission was clearly confirmed. Two years later he decided to stop visiting the hospital for his regular examinations.

In December 1999, he began to experience muscle pain and weakness with macrohematuria. A few weeks later, on January 11, 2000, he visited our hospital and was readmitted. Hematological study showed elevations of CK 1,291 IU/l, LDH 1,992 IU/l, and Cr 5.5 mg/dl (0.6–1.2 mg/dl). Urinary test showed mild proteinuria. Urinary sediment revealed blood cells (55–99/high-power field), granular cast, and cellular cast. We initially ascribed his elevation of serum Cr level to acute tubular injury, but serum levels of C3, C4, and CH50 were decreased and immune complex was elevated. In addition, ANA, anti-ds DNA antibody, and anti-Jo-1 antibody were positive. We noted an irregular, nonspecific erythema on the patient's face similar to that seen at his first admission, but we could not determine whether the erythema was induced by systemic lupus erythematosus (SLE) or DM.

Gastrointestinal endoscopy revealed a IIc lesion of more than 3cm in the upper body of the stomach, and biopsy showed a poorly differentiated adenocarcinoma. Computed tomography and ultrasonography did not detect any metastasis.

Serum levels of CK, LDH, and Cr became normalized after three repeated sessions of steroid pulse therapy and oral prednisolone (40 mg daily). We added oral cyclophosphamide (75 mg daily) and tapered the prednisolone to 20 mg daily for the operation. A proximal gastrectomy and open renal biopsy were performed on March 27. Light microscopy revealed a mild global mesangial hypercellularity in all glomeruli, and an immunofluorescence study revealed deposits of IgG and C3 in glomerular mesangium (Fig. 1). An electron microscopic study also showed mesangial deposits. As the patient totally did not meet the criteria for SLE, the above findings prompted us to diagnose his condition as mesangial glomerulonephritis (Lupus nephritis, type IIb of WHO classification). We concluded that his elevated Cr was induced by a combination of lupus nephritis and acute tubular injury due to myoglobinemia. Cyclophosphamide was stopped due to liver injury after the operation, but normal serum levels of CK, LDH, and Cr remained. The patient is now in remission with oral prednisolone (20 mg daily). DM associated with neoplasia is classified as type III. The risk of cancer in patients with DM is very high for the first five years after the diagnosis of DM (6). Our patient remained in favorable condition and felt comfortable staying away from

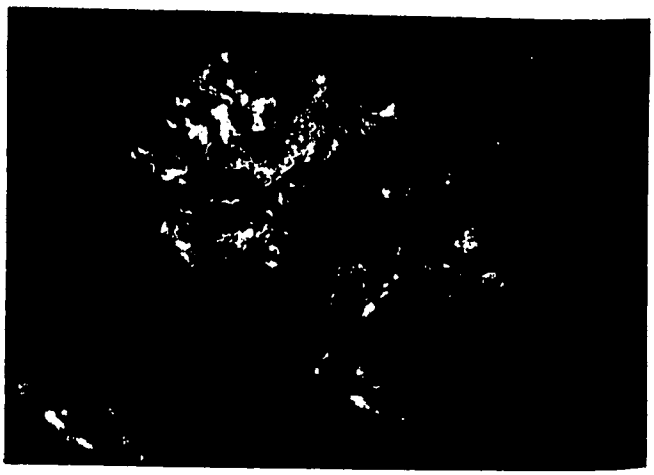

Figure 1. Immunofluorescence photomicrograph (×200) showing deposits of C3 in the glomerular mesangium.

the hospital during that time. By good fortune, no metastasis was detected and his gastric carcinoma could be resected curably. DM with malignancy is reported to have a satisfactory prognosis after curative operations. For reasons we cannot explain, the present case has also remained in good condition postoperatively.

Overlap syndrome is more often seen in younger female patients (1), but our patient was an elderly male accompanied with gastric carcinoma. SLE also has associations with malignant diseases such as hemopoietic malignancies and cancers of the cervix (7). However to our knowledge, DM with gastric carcinoma and lupus nephritis has never been previously reported. While we cannot deny that the co-appearance of gastric carcinoma and overlap syndrome was a coincidence, it is significant that the relapse of DM with lupus nephritis occurred just when the gastric carcinoma appeared.

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