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A Case of Dermatomyositis with Great Improvement after the Resection of Gastric Carcinoma

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Abstract Although the cause and etiological mechanisms of dermatomyositis remain unknown, an association of dermatomyositis with internal malignant tumors has been discussed. A 67-year-old man was seen in our clinic with muscle weakness and erythema and was diagnosed as dermatomyositis. He was then found suffering from gastric carcinoma. Undergone the resection of gastric cancer, the patient showed a considerable improvement in his clinical features of dermatomyositis without steroid administration. We describe the case referring to the recent literature.

Key Words: Dermatomyositis associated with malignancy, Resection of gastric cancer, Treatment without steroid

I. Introduction

It has been well known that dermatomyositis (DM) is often associated with internal malignant tumors, especially when observed in patients over fifty years old¹⁾. We present a case of DM accompanied by gastric carcinoma. The muscular and cutaneous symptoms and other laboratory findings have greatly improved in the patient after the resection of gastric cancer. We describe the case and compared it with other cases previously reported.

II. Case Report

A 67-year-old male, a bus driver, was examined in our clinic, Department of Dermatology in Yamaguchi University Hospital. Three months prior to the examination, he began to notice a little muscular weakness in

his upperarms and scapular regions and difficulties in going up and down the stairs and standing up. Ever since then, he had suffered from a stomach ache after meals as well. One month before the examination, reddish eruptions had appeared on the scalp, face, chest, elbows, knees and dorsa of both hands. Edema was present on the face especially on the eyelids. He complained of a strong itching over the extremities, and general fatigue which increased. He was then hospitalized. It was noted that his father died of gastric carcinoma.

Clinical features at admission:

Strong edema was present on the eyelids which caused his eyes difficulty in opening (Fig. 1). Typical heliotrope bloating was observed on his upper eyelids (Fig. 2). Erythema with scales was seen on the joint regions of the dorsa of both hands (Gottron's sign), and paronychial erythema of fingertips had

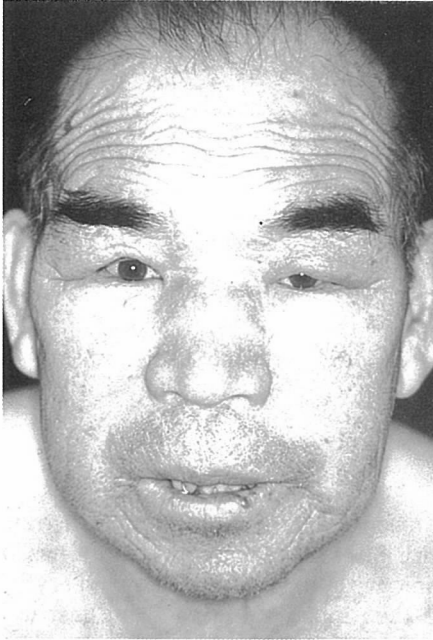


Fig. 1 Remarkable edema present on the face

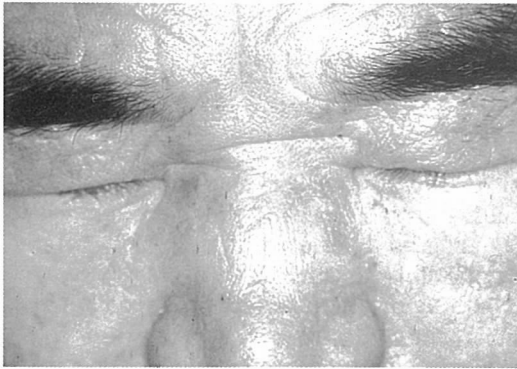


Fig. 2 Typical heliotrope bloating observed on the upper eyelids



Fig. 3 Erythema with scales seen on the joint regions of the dorsa of both hands (Gottron's sign)

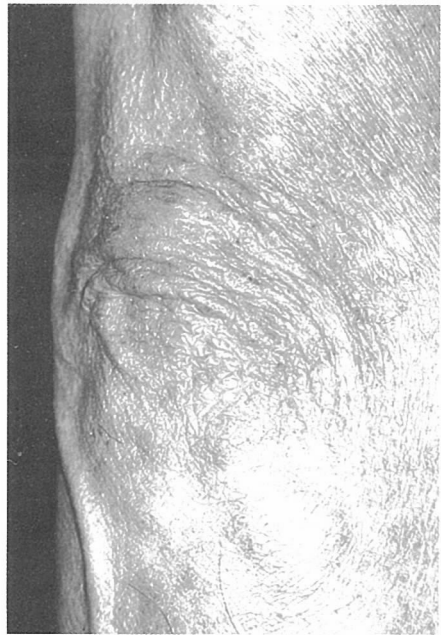


Fig. 4 Erythema on the elbow-joint

developed (Fig. 3). Erythema followed by keratinization and cutaneous thickening appeared on the extensor elbow joints and on both knees (Fig. 4). Diffuse erythema was visible on the V-zone of the chest (Fig. 5), and that on the gluteal regions and on the upper extremities was accompanied with itching.

Histopathological findings :

A biopsy specimen was taken from the skin of erythema with scales on the joint region of the hand (Fig. 6). The epidermis showed hyperkeratosis, diminishing of the rete ridges, and liquefaction degeneration of the basal cell layer. The dermis was edematous and showed a cellular infiltration composed mainly of lymphocytes in perivascular arrangement. The muscle showed bundles composed of fibers of various sizes, vacuolar

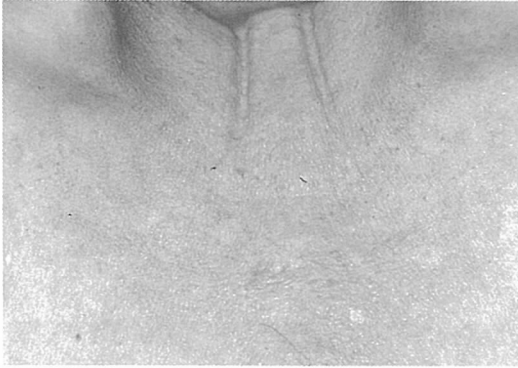


Fig. 5 Diffuse erythema on the V-zone of the chest

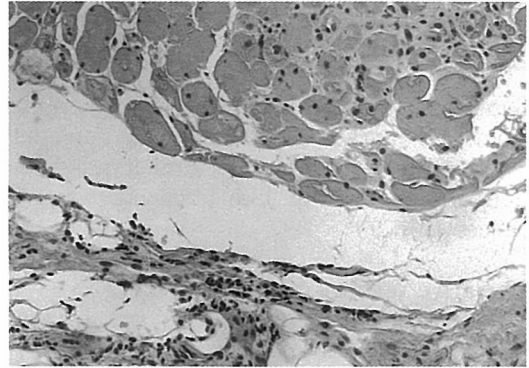


Fig. 7 Biopsy specimen obtained from the left deltoid muscle: The muscle bundles show fibers of various sizes, vacuolar degeneration, and the infiltration of lymphocytes. Lymphocytes infiltrated in the connective tissue. (HE stain, x 300)

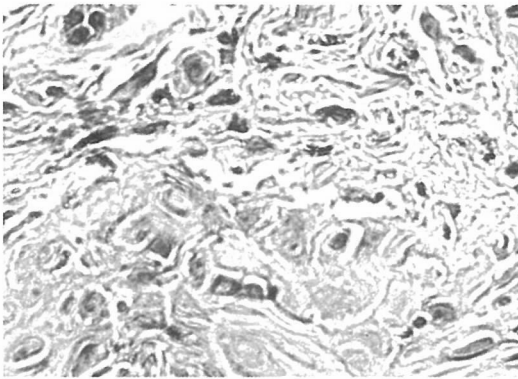


Fig. 6 The specimen obtained from the skin lesion on the wrist joint seen in Fig. 3: The dermis is edematous and shows a cellular infiltrate composed mainly of lymphocytes in perivascular arrangement. (HE stain, x 1,000)

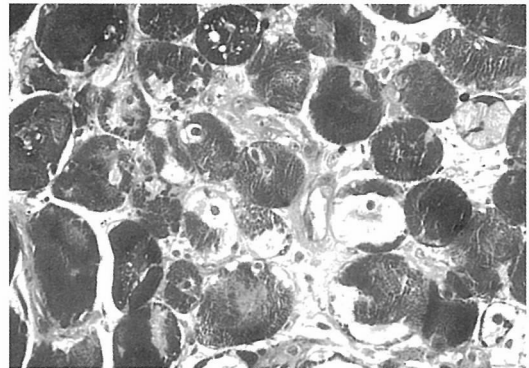


Fig. 8 The specimen obtained from the left deltoid muscle. Some parts of the muscular fibers were not stained with Azan staining. (Azan stain, x 500)

degeneration, and infiltration of lymphocytes. Lymphocytes had infiltrated into the connective tissue (Fig. 7). Some parts of the muscular fibers were not stained with Azan staining (Fig. 8).

Laboratory findings :

Biochemical tests revealed the following: CPK 917U, LDH 423U, GOT 45U, aldolase 15.2U, and serum myoglobin 380ng/ml. All the levels were above the normal limit.

Immunological tests were as follows: anti-nuclear antibody x10 (nucleolar pattern), thyroid test x1600, microsome test x102400.

Tumor markers: CEA 1.5ng/ml, ferritin 231ng/ml, serum β_2 globulin 6445ng/ml, SCC 2.2ng/ml, and CA19-9 85U/ml. Some of the values were higher than the normal limit.

Electromyogram (EMG) studies of arms, thighs and legs showed a pattern of low amplitude and short duration potentials. No abnormal data were obtained in the examinations of ECG, chest x-ray, pulmonary function test, RA, anti Jo-1 antibody, immune complex, I gG, I gM and C 3.

These findings led us to make the diagnosis of dermatomyositis.

To investigate the internal malignant tumor, x-ray examination of the uppergastrointestinal tract was done, which detected ulcers accompanied by tunica mucosa indicative of the II C+III like advanced gastric cancer (Fig. 9).

Operative findings :

A resection of gastric cancer was performed by surgeons of Ymaguchi University Hospital. The tumor was progressive and reached the serous membrane. Metastasis to lymphnodes was observed (Fig.10).

Histopathological findings of the tumor :

A biopsy specimen was taken from the tumor of the stomach. Poorly differentiated adenocarcinoma was found to be mingled with mucinous cells (Fig.11).

Treatment and Prognosis :

Edema on the face began to decrease after the resection, leaving a slight erythema on

patient's face one month after the operation (Fig.12). Erythema with scales on the dorsa of the hands remained unchanged one month after the operation, and began to decrease two month after the operation, leaving a slight pigmentation by the end of three months.



Fig.10 The stomach resected from the patient



Fig. 9 X-ray film detected ulcers accompanied by tunica mucosa indicative of the IIC+III like advanced gastric cancer.

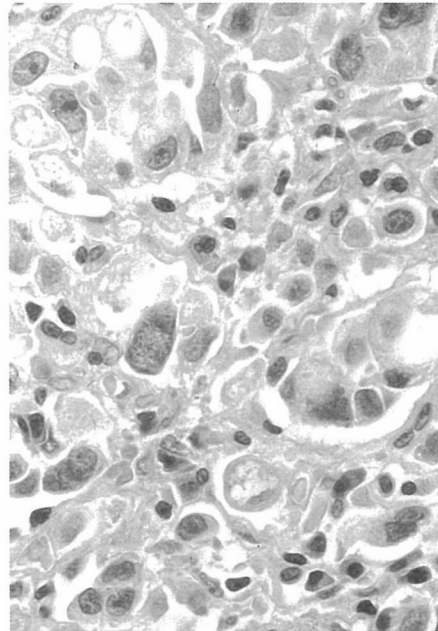


Fig.11 A section from the tumor of the stomach showed poorly differentiated adenocarcinoma which was mingled with mucinous cells. (HE stain, x 1,000)

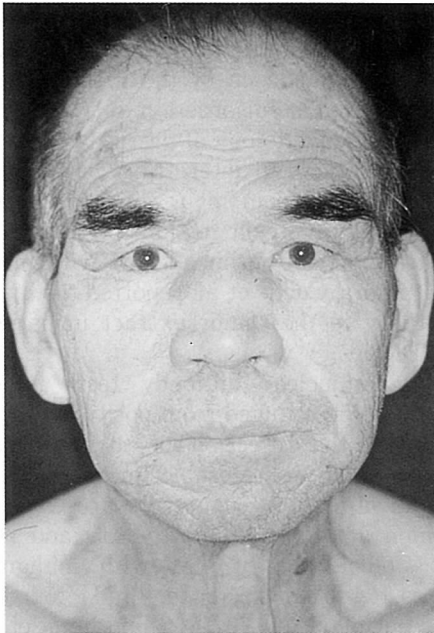


Fig.12 Erythema on patient's face markedly reduced one month after the resection of gastric cancer.

After operation, erythema on the chest became itchy and formed erosions and ulcers due to scratching.

The erosive erythema disappeared after two months, and a slight pigmentation and scars remained after three months. The itchy erosive erythema on the thighs also diminished following a similar course. Table 1 summarizes the laboratory findings before and after the resection. High values of CPK, LDH, GOT, aldolase and serum myoglobin indicating active myositis returned to the normal value after the resection. On the other hand, tumor markers showed high values one month after the resection when the eruption worsened. Three month after the operation, the levels of tumor markers improved.

After one month of the operation, the EMG still showed the weakening of the leg muscles. On the other hand, arm and thigh muscles showed normal patterns. Three months after the operation, the EMG showed a normal pattern for the muscles of legs.

Fig.13 shows the laboratory data and the

Table 1 Changes of Laboratory Findings before and after the Resection of Gastric Cancer

	before resection	after resection (month)					
		1	3	7	9	13	17
Biochemical Examination							
CPK (<260 U)	917	33	25	44	43	46	64
LDH (<240 U)	423	269	200	217	209	198	257
GOT (< 24 U)	45	12	11	11	10	10	13
aldolase (< 4.4U)	15.2	7.3	5.5	5.3	4.7	4.4	n.d.
serum myoglobin (< 70 ng/ml)	380	112	32	n.d.	n.d.	n.d.	n.d.
Immunological Examination							
antinuclear antibody	x 10	-	-	x100	x100	x100	n.d.
thyroid test	x1600	x400	x1600	n.d.	n.d.	n.d.	n.d.
microsome test	x102400	x25600	x102400	n.d.	n.d.	n.d.	n.d.
Tumor Markers							
CEA (< 5.0ng/ml)	1.5	7.6	2.0	2.4	3.5	4.5	17.9
ferritin (< 105 ng/ml)	231	416	241.8	n.d.	290.5	204.4	362.0
serum microglobulin (<2400 ng/ml)	6976.2	6562.5	5350.8	5657.4	4823.7	3660	7200
SCC (< 2.0ng/ml)	2.2	2.9	1.5	n.d.	n.d.	n.d.	n.d.
CA19-9 (< 37 U/ml)	85	280	76	n.d.	n.d.	n.d.	n.d.
EMG	low amplitude short duration normal						

() : normal range - : negative n.d. : not done

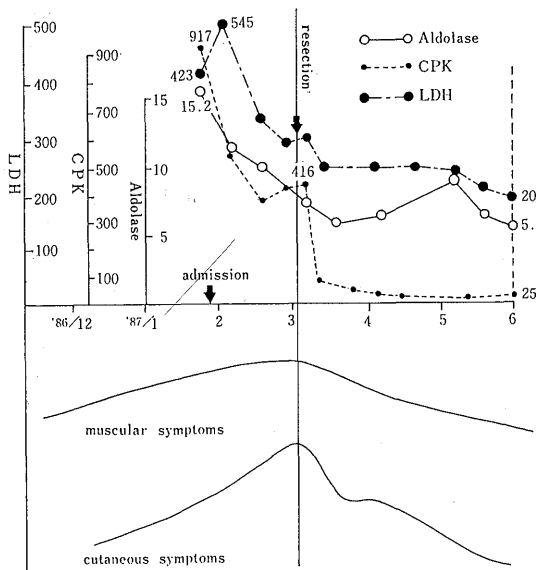


Fig. 13 As the laboratory data became normalized, clinical symptoms of the patient improved.

clinical course. Rehabilitation, which proved effective, started one month after the operation referring CPK values. An antiallergic drug and antihistamins were administered to soothe itching and eruption. Itching diminished three months after the operation.

III. Discussion

We made the diagnosis of DM on the basis of increased levels of muscle enzymes such as CPK, aldolase and LDH, the observation of inflammation in the biopsy specimen, the changes of myographic patterns on EMG and other typical clinical features. DM of the patient was found to be associated with gastric cancer. This type of DM was classified as type III by Bohan & Pearson classification²⁾.

DM is considered to be an autoimmune disorder with unknown etiology. Following reports suggest that DM belongs to autoimmune diseases; Nishikai and Reichlin found the antibody to calf thymus, the anti-Jo-1 antibody in DM patients³⁾, Kakulas reported the destruction of differentiated muscle cultures by sensitized lymphoid cells⁴⁾, and Whitaker and Engel as well as Morita and Morita reported the deposition of I gG, I

gM, C 3, I gA and Ciq on the blood vessels of limb girdle muscles in children with DM⁵⁾⁶⁾.

The high incidence of the association of DM with immune disorders may support the immunological etiology of the disease. Sasaki et al. presented an interesting case of DM associated with pemphigus foliaceus, one of the typical immunological disorders⁷⁾. Concerning the relationship of DM with malignant tumors, Curtis et al. reported the hypersensitivity to the tumor extract in patients with DM⁸⁾.

The antinuclear antibody tests in our patient which changed from positive to negative after the resection of the malignant tumor and the considerable improvement of the clinical features suggest the presence of a common antigen in tumors, cutis and muscles. Coincidence of DM with malignant tumors has been discussed; Ikada found 66 cases out of 190 (34.7%)⁹⁾, Shinoshima et al., 67 out of 177 (37.9%)¹⁰⁾, and Kaneko et al., 170 out of 567 (30%)¹¹⁾. Coincidence ratio of DM with gastric cancer is the highest in Japan. Fifty-seven DM patients were associated with gastric cancer out of 180 DM patients with malignant tumor (32%) according to Kaneko's report. Males in fifth decade are most frequently affected with DM while females are in their forties, and the incident ratio of male to female is one to two. On the other hand, the coincidence of DM with malignant tumor becomes higher after fifty, especially in male (66% by Kaneko) and its ratio of male to female is two to one¹¹⁾.

The frequencies of malignant tumors that occur prior to or concurrent with or subsequent with DM have been reported; cases of antecedence of DM and antecedence of malignant tumor (MT) were 84.9% and 15.3% respectively, according to Shinoshima et al.¹⁰⁾, cases of antecedence of DM, antecedence of MT and concurrence of DM and MT were 59%, 31%, and 10%, respectively, according to Barnes and Mawr¹²⁾, and cases of antecedence of DM and antecedence of MT were 82.4% and 17.6% respectively, according to Kaneko et al.¹¹⁾. Antecedence of DM was found more frequently than antecedence of MT. In any cases, however, the subsequent clinical features of either DM or MT were found within six months. Judging from

the clinical course, DM in our case appeared to be concurrent with MT.

Systemic administration of steroid is generally applied to patients with DM. There are some DM patients with internal cancer who show no response to steroid administration¹³⁾. In such patients, the improvement of DM largely depends on the treatment of internal cancer. Immunosuppressants are given to DM patients with malignant tumors whose symptoms do not respond to steroid or MT treatment. It is true that patients of DM with malignant tumors have poor prognosis. There are a few reports, however, on the cases which showed considerable improvement after the treatment of malignant tumors; five years absence of DM by the treatment of breast carcinoma²⁾, and great improvement of DM after the administration of steroid and the resection of rectum carcinoma¹⁵⁾. However, this is the first report on the case of DM with gastric carcinoma which was greatly improved by the resection of tumor without general steroid administration.

IV. Conclusion

The fact that the symptoms of eruptions, muscular weakness, EMG findings and CPK levels have greatly improved after the resection of gastric cancer without the general steroid administration suggests a close correlation of DM with malignant tumors.

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