#### LETTER TO THE EDITOR

# Dermatomyositis associated with thyroid cancer: a paraneoplastic syndrome?

Takao Nagashima · Sachiko Onishi · Yasuyuki Kamata · Seiji Minota

Received: 12 November 2008 / Accepted: 5 January 2009 / Published online: 14 January 2009 © Springer-Verlag 2009

Dear Editor,

We have read the recent article by Fujita et al. [1] "Dermatomyositis associated with thyroid cancer" with great interest. They reported the removal of the coexisting thyroid cancer in a patient with corticosteroid resistant dermatomyositis (DM) resulted in improvement of DM. We have experienced two cases of DM associated with thyroid cancer, both of them responded to conventional immunosuppressive treatment without removal of thyroid cancer.

## Case 1

A 59-year-old Japanese woman was referred to our hospital due to muscle weakness and elevation of serum creatine kinase (CK) level. On examination, she had Gottron's signs and periungual erythema on her hands. Heliotrope rash was not observed. Fine crackles were heard on chest auscultations. Blood tests showed serum CK of 4,220 IU/l (normal 32–180), serum anti-nuclear antibody (ANA) was high at 1:2,560 with a speckled pattern. Her rheumatoid factor (RF) was 1:1,280, anti-RNP, SSA, Scl-70, Jo-1, double-stranded DNA antibody were all negative. Magnetic resonance imaging (MRI) of the shoulder demonstrated high-signal intensities in trapezius, subscapular, infra and supraspinatus muscles on gadolinium-enhanced images and fat-suppressed images, but no abnormal signal was

detected in biceps and triceps muscles. Electromyography demonstrated normal findings for biceps and triceps muscles. Therefore, muscle biopsy was not performed. Chest computed tomography (CT) revealed interstitial pneumonia (IP) in the bilateral lung base, and incidentally, low-density area in left thyroid lobe was detected. The size was  $22 \times 18 \times 16$  mm under ultrasonography (US). US-guided needle aspiration biopsy demonstrated class V cytology, thyroid papillary carcinoma. Lymph nodes metastasis was not detected. She was diagnosed as DM with IP and thyroid cancer. The resection was considered before corticosteroid therapy; however, the thyroid surgeons recommended the treatment of DM first, not the resection of the cancer. Therefore, she was treated with intravenous methylprednisolone pulse therapy and prednisolone (PSL) 50 mg/day (1 mg/kg). She showed a good response to corticosteroid therapy in both muscle weakness and IP. During tapering the prednisolone at an outpatient basis, cyclosporine 100 mg/day (2 mg/kg) was added due to the slight increase of the CK levels. After 1 year, she is on PSL (11 mg/day) and cyclosporine (100 mg/day) with no evidence of relapse. The size of the thyroid cancer was not changed.

## Case 2

A 48-year-old Japanese woman was referred to our hospital due to fever, dyspnea, muscle weakness, and arthralgia. She was diagnosed as rheumatoid arthritis (RA) and IP a year ago. On examination, she had arthralgia in bilateral shoulders, wrists, metacarpophalangeal and proximal interphalangeal joints. Gottron's papule and periungual erythema were observed on her hands, but heliotrope rash was not seen. Laboratory findings showed CK of 2,298 IU/l, ANA was negative, RF, anti-Jo-1 antibody, and anti-cyclic

T. Nagashima (☒) · S. Onishi · Y. Kamata · S. Minota Division of Rheumatology and Clinical Immunology, Jichi Medical University, Yakushiji 3311-1, Shimotsuke, Tochigi 329-0498, Japan e-mail: naga4ma@jichi.ac.jp



citrullinated peptide antibody were positive. MRI of the thigh showed high-signal intensities in bilateral gluteus, obturator, and deeper quadriceps muscles. Electromyography of the thigh showed myogenic abnormalities; however, quadriceps muscle biopsy revealed normal findings. Chest CT showed IP in the middle and lower lung field, and incidentally, irregular low-density area in the thyroid was detected. The size was 9 mm diameter under US, and needle aspiration biopsy demonstrated class V cytology, papillary carcinoma. She was diagnosed with coexistent RA, DM with IP, and thyroid cancer. We again consulted to the surgeons, but they recommended treatment of DM. Oral PSL 75 mg/day (1 mg/kg) and cyclosporine 100 mg/day were started, and she showed a good response to the therapy. After 11 months, she is on PSL (8 mg/day) and cyclosporine (100 mg/day). The size of the thyroid cancer was unchanged.

So far, very few cases are reported about an association of DM and papillary thyroid carcinoma [1–5]. In Japanese autopsied DM cases, the frequency of thyroid cancer is reported at 9.5% [2]. By contrast, the frequency of thyroid carcinoma in several autopsied case series has been reported at 5.6–35.6% [6]. Incidence of thyroid cancer is increasing in general population. It is due to increased detection of subclinical diseases by increased diagnostic scrutiny, not an increase in the true occurrence [7]. Fujita et al. [1] suggest that DM associated with thyroid cancer may be more frequent than generally expected. It may be true, but the frequency of thyroid cancer in DM may not differ from that in general population.

They also insisted that removal of the thyroid cancer resulted in improvement of the DM [1]. We do not agree with their assertion. The patient was diagnosed as thyroid cancer 10 days after the start of the therapy, and the patient was considered "corticosteroid resistant" because of insufficient improvement. We think it is too early to conclude "corticosteroid resistant" in such a short period. The initial corticosteroid treatment often needs more than a month to normalize CK levels [8]. In another case, resection of the

thyroid cancer did not normalize CK levels and needed additional immunosuppressive treatments [3].

In both of our cases, the tumor size was unchanged 1 year later. They might have already existed long before the diagnosis of DM was made. Patients who had interstitial lung disease have a significantly lower frequency of malignancies [4, 9]. Our cases contradict the findings. We believe that thyroid cancer is not associated with DM. The rarity of the evident thyroid cancer with DM supports the notion [10].

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