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CASE REPORT

Paraneoplastic Dermatomyositis Accompanying Nasopharyngeal Carcinoma

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NASOPHARYNGEAL carcinoma (NPC) is a rare disease in North America and Western Europe, but is common in Asia and North Africa.^{1,2} A small percentage of NPC patients present with paraneoplastic syndrome and less than 0.1% with dermatomyositis.² No prospective case-control double-blinded studies of immunosuppressive therapy in dermatomyositis have been performed, let alone in paraneoplastic dermatomyositis.^{2,3} Treatment of paraneoplastic dermatomyositis largely refers to experience of case reports and remains largely empirical.^{2,3} We present a case of NPC accompanied with paraneoplastic dermatomyositis who was refractory to standard prednisone therapy but was almost completely remitted after treatment with pulse oral dexamethasone therapy subsequently with low dose of prednisone.

CASE DESCRIPTION

A 57-year-old Chinese man presented with periorbital edema and heliotrope macular rash for 4 months. He had been treated with corticosteroids, hydroxychloroquine, tripterygium glycoside, yet the manifestation progressively aggravated. The patient was then admitted to our hospital

in June 2010. His alanine transaminase, aspartate transaminase, creatine kinase, creatine kinase isoenzyme MB and lactic dehydrogenase levels were all within the normal range. Antinuclear antibody, anti-SS-A, anti-SS-B, anti-Sm, anti-RNP, anti-Scl-70 and anti-Jo-1 antibodies were all negative. Result of electromyography showed that the conduction velocity of H wave of the left tibial nerve was decreased, and evidence of nerve damage in right and left quadriceps femoris and right first interosseus. Hematoxylin and eosin staining of the skin biopsy revealed interface dermatitis and perivascularitis throughout the entire dermis. A biopsy *via* endoscopy led to pathological diagnosis of non-keratinizing NPC. Therefore, the patient was diagnosed with paraneoplastic dermatomyositis accompanying NPC.

Treatment began with chemotherapy (oxaliplatin and 5-fluorouracil), but did not relieve symptoms. After chemotherapy, 65 mg oral prednisone (1 mg/kg) daily was administered. External beam radiation therapy was started ten days after chemotherapy. Nonetheless, the patient's manifestations gradually deteriorated. Four weeks after initiation of radiotherapy, the patient could hardly open his eyes. Then, 40 mg oral dexamethasone was substituted for prednisone. Within three days, manifestation was partly remitted, we replaced the 40 mg dexamethasone with an initial dose of 40 mg prednisone, followed by decrement of 5 mg every week. Two weeks later, periorbital edema and

rash were almost complete remitted (Fig. 1). After the prednisone dose reached 10 mg, the weekly decrement was reduced to 2.5 mg until withdrawal. After radiation, one more cycle of chemotherapy was administrated. Currently, the NPC in this case is in remission and the patient is living without any evidence of recurrence of dermatomyositis.

DISCUSSION

In the literature, paraneoplastic syndrome accompanying NPC are mostly described in case reports or review articles. Dermatomyositis and hypertrophic osteoarthopathy are the most common types of paraneoplastic syndrome accompanying NPC.⁴ Paraneoplastic syndrome occurs in 1%-7.4% of cancer patients and less than 0.1% NPC is accompanied with dermatomyositis.² Paraneoplastic dermatomyositis can precede, follow or be concurrent to the diagnosis of a malignancy. Usually paraneoplastic dermatomyositis flare-ups coincide with cancer recurrence, although not found in all cases.⁵ There is about a three-fold increase in risk of malignant diseases after diagnosis of dermatomyositis but the frequencies of specific types of cancer vary greatly in different reports.^{6,7} Therefore, the performance of whole-body FDG-PET/CT for diagnosing occult malignant disease in patients with dermatomyositis is recommended by some researchers.⁷

Treatment of paraneoplastic dermatomyositis remains largely empirical. van de Vlekkert et al⁸ reported that paraneoplastic dermatomyositis could spontaneously recover. Paraneoplastic dermatomyositis healed with anticancer treatment alone was also reported.⁹ Therefore, some oncologists regard the treatment of paraneoplastic dermatomyositis as unnecessary. Some other oncologists believe that the treatment of paraneoplastic dermatomyositis should

be the same as the treatment of dermatomyositis without coexisting cancer, or for a shorter period.¹⁰ Treatment of dermatomyositis has been well described previously.^{3, 11} Oral prednisone at 0.5-2 mg/kg daily and pulse intravenous methylprednisolone at 30 mg/kg for more severe disease are the standard treatment. For patients with refractory dermatomyositis, none corticosteroids therapy (including methotrexate, immunoglobulin, rituximab, azathioprine, cyclosporine, cyclophosphamide, tacrolimus, etc.), combinations of therapies or the addition of newer therapies (including bone marrow or stem cell transplant, anti-tumor necrosis factor therapy, anakinra, alemtuzumab, etc.) are now frequently used.^{3, 11} Paraneoplastic dermatomyositis in this case was refractory to standard prednisone therapy but completely remitted with oral highdose dexamethasone and subsequent low-dose prednisone on a tapering schedule of approximately 10 weeks.

The role of paraneoplastic dermatomyositis in the prognosis of cancer remains to be elucidated. Though paraneoplastic dermatomyositis itself can be fatal (most commonly due to respiratory failure), most researchers believe that paraneoplastic dermatomyositis does not affect the prognosis of NPC.²

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Figure 1. Cutaneous manifestation and periorbital edema before chemotherapy (left) and 2 weeks after dexamethasone treatment (right).

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