Dermatomyositis in a 28-year-old Filipino male with nasopharyngeal carcinoma: A case report

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This is a case of a 28-year-old male newly diagnosed with nasopharyngeal carcinoma (NPC), referred to our service for a 2 month history of poikiloderma on the anterior chest and back, with associated periorbital edema, facial erythema and periungual telangiectasia. Initial diagnostic work-ups revealed a markedly elevated total creatine phosphokinase (CPK-total). The patient subsequently developed symmetric proximal muscle weakness. Electromyography study was positive for myopathy. Based on the physical examination and diagnostic findings, the patient was diagnosed with dermatomyositis (DM), a rare autoimmune connective tissue disease primarily affecting the skin and muscles. Treatment with oral prednisone, topical mometasone, and photoprotection for 1 month yielded improvement of symptoms. The association of dermatomyositis with nasopharyngeal carcinoma (Paraneoplastic Dermatomyositis) is rare. Ultimately, treatment of the underlying malignancy is the cornerstone of management. Early diagnosis led to prompt treatment and amelioration of debilitating symptoms while awaiting definitive management of his concomitant malignancy.

Keywords: dermatomyositis, nasopharyngeal carcinoma, paraneoplastic

INTRODUCTION

Dermatomyositis (DM) is a multisystem inflammatory disorder, primarily affecting skin and muscle. While it has been associated with various forms of malignancies, its association with nasopharyngeal carcinoma (NPC) is rare, with an incidence of less than 1/1000. We present a case of an adult male newly diagnosed with NPC who simultaneously developed cutaneous and systemic manifestations of DM. The distinct cutaneous findings of this disease place the dermatologist at the forefront of recognizing this illness and initiating appropriate management.

CASE REPORT

A 28-year-old Filipino male, known case of undifferentiated non-keratinizing NPC stage IVB, presented with a 2 month history of multiple erythematous and hypopigmented patches on the anterior chest and upper back (Figure 1). Lesions later progressed into crusted erosions, with the concomitant appearance of ill-defined erythematous patches over the face, and periorbital edema. One week later, he developed difficulty of breathing and proximal muscle weakness which manifested as difficulty in raising both hands, getting up from bed, and in climbing stairs.

Physical examination revealed diffuse facial edema and erythema with periorbital swelling (Figure 3), positive V sign with erythematous and hypopigmented patches (poikiloderma) on the anterior chest, positive shawl sign on the upper back, positive Gottron’s papules on hands, periungual telangiectasia on the fingers (Figure 4), and decreased muscle

Figure 1. On initial presentation. A. Multiple poikilodermatous patches (“V sign”) on the anterior chest. B. Multiple erythematous well-defined dusky red patches on the nape & deltoid region (“shawl sign”) with few erythematous patches topped with hemorrhagic crusts on the lower back

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Source of funding: none
Conflict of interest: none
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**Figure 2.** Biopsy of lesion on right anterior chest (H&E x100) A. Epidermis shows basket weave hyperkeratosis, epidermal atrophy & vacuolar interface change (RED arrow). B. Dermis noted to have perivascular lymphohistiocytic infiltrates, dermal edema & increased mucin (ORANGE arrows).

**Figure 3.** At 10 days after starting oral Prednisone. Increase in periorbital edema but decrease in poikiloderma & crusts on anterior chest.

**Figure 4.** At 10 days after starting oral Prednisone. Still with periungual telangiectasia on right hand.

**Figure 5.** At 3 months after starting oral Prednisone & 2 cycles of chemotherapy. Resolution of periorbital edema, poikiloderma & crusts on anterior chest.

**Figure 6.** At 1 month after starting oral Prednisone. Lightening of periungual telangiectasia on right hand.
of Bohan and Pete precluded the need for a muscle biopsy. Patient was initially treated with pulsed IV methylprednisolone for 3 days, then shifted to prednisone 40mg/day PO maintained for 4 weeks. Topical mometasone lotion, saline compress, fucidin cream, antihistamines, emollients and sunscreen were also prescribed. After 1 month of treatment, an 80% improvement of cutaneous lesions (Figures 5 and 6) was observed along with marked improvement of other physical findings, particularly an increase in muscle strength. Repeat CPK-total improved to 2,111 (N<308). As of writing, the patient has completed 2 cycles of chemotherapy with 5-FU + cisplatin and is due for subsequent radiotherapy.

**DISCUSSION**

Dermatomyositis (DM) is fairly uncommon with an overall incidence of 1 per 100,000 people per year and a prevalence of 20 per 100,000 people. Symptoms of patients with DM are highly variable, ranging from localized skin lesions to life-threatening systemic disease. Diagnosis is based on 5 criteria by Bohan and Pete, which includes: 1) progressive symmetrical weakness of proximal limb muscles and anterior neck flexors; 2) dermatologic signs (Heliotrope eruption with periorbital edema and erythema; Gottron’s papules); 3) positive muscle biopsy; 4) elevated muscle enzymes (CPK, LDH and aldolase); and, 5) EMG signs of myopathy. Other characteristic lesions include V sign, shawl sign, facial erythema and edema, photodistributed poikiloderma and perungual telangiectasia. Presence of 3 or 4 criteria plus dermatologic signs is definitive for DM which our patient fulfilled.

Dermatomyositis is known to be associated with various types of cancer with its incidence ranging from 7% to 34% among different studies. Malignancy may precede, occur concomitantly with, or follow the diagnosis of DM with equal frequencies. It should be suspected within the first 2 years after the onset of DM and screening is recommended annually in the first 3-5 years. The association of NPC with DM has been well documented in South-East Asia but is rare with an incidence of less than 1/1000. The underlying mechanism for the association of DM with malignancy, however, remains inconclusive.

The cutaneous and systemic manifestations of DM can be debilitating hence treatment is warranted. For cutaneous manifestations, strict photoprotection, topical corticosteroids, anti-malarials such as oral hydroxychloroquine (200-400mg/day), and emollients are indicated. Cytotoxic drugs (methotrexate, azathioprine, cyclophosphamide) can be given as 2nd line agents. For systemic manifestations, high doses of systemic corticosteroids are recommended with either pulse methylprednisolone therapy for 1 to 3 days, or prednisone in 1-2mg/kg in divided doses. A high dosage of prednisone is maintained for 4-6 weeks followed by a slow taper (5-10mg/week) over the next 10-12 weeks. With this treatment, approximately 90% of patients improve partially, while 50% to 75% of patients achieve complete remission.

Reports have shown that DM associated with cancer is generally more resistant to corticosteroid & cytotoxic therapies. The value of treating the underlying cancer lies in the fact that the activity of DM usually mirrors that of the malignancy. When a patient enters a period of cancer remission, their DM activity can be used to monitor for early relapse.

Patients with DM may spontaneously remit in as many as 20%, while 5% may have a fulminant course with eventual death. Those who have an associated malignancy and who are elderly (>60 y/o) are considered to have a poorer prognosis. Those that survive may develop residual weakness and disability. Therefore, long-term therapy and follow-up is required.

**CONCLUSION**

Patients presenting with characteristic skin findings associated with symmetric proximal muscle weakness are suspicious for DM. Diagnosis is confirmed through clinical and laboratory evaluation, histology, and electromyography. Increasing awareness of the association between DM and NPC can help in appropriate screening especially in patients belonging to susceptible ethnic groups. Dermatologists play an essential role in the early diagnosis, treatment, monitoring and improvement of overall prognosis.

**REFERENCES**