

Bullous Dermatomyositis Associated with Nasopharyngeal Carcinoma

-A Case Report

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Abstract

Cutaneous manifestations of dermatomyositis commonly include Gottron's papules, heliotrope rash, photosensitivity, poikiloderma and nailfold telangiectasia. Vesicles and bulla are rare. We report a patient with dermatomyositis who presented with blisters and oral ulcers. It is important to recognise this bullous variant in order to avoid a delay in diagnosis. Bullous dermatomyositis may also portend a poorer prognosis. Our patient was subsequently diagnosed to have undifferentiated nasopharyngeal carcinoma.

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Key words: Bullous, Dermatomyositis, Malignancy, Nasopharyngeal carcinoma

Introduction

Bullous dermatomyositis is a rare variant of dermatomyositis and has been reported in the literature. It is believed to be strongly associated with the presence of a malignancy. However, oral ulcers have not been reported in association with dermatomyositis. We present the first such case of a patient with dermatomyositis presenting with blisters and oral ulcers, who was later diagnosed with nasopharyngeal carcinoma (NPC).

Case Report

A 48-year-old Chinese man presented with mouth ulcers and a blistering eruption over the scalp, arms, thighs and upper trunk for 2 months. The blisters were pruritic and were aggravated by exposure to sunlight. The mouth ulcers were painful. He also complained of difficulty breathing, arthralgia, loss of appetite and weight loss over the same time. He had mild diabetes mellitus on dietary control. There was no significant drug history.

Clinically, there were multiple erosions on erythematous patches and a few bullae over the arms, dorsum of hands, thighs, upper back, chest and scalp (Fig. 1). Several erosions were seen in the buccal mucosa. The face and other mucosae were spared. He had fine crepitations in the lung bases. The rest of the systemic review was normal and there was no muscle weakness at initial presentation.

The differential diagnoses considered at presentation included pemphigus vulgaris, bullous pemphigoid,

bullous drug eruption, porphyria cutanea tarda, bullous lupus erythematosus, migrating necrolytic erythema and bullous dermatomyositis.

Bullae and oral ulcers were clearly documented by the attending dermatologists at first visit to the clinic. However, these resolved spontaneously and quickly before a skin biopsy could be arranged. The patient did not have subsequent new blister formation or oral ulceration. The biopsy was taken from an erythematous patch over the thigh and showed dermal oedema with a superficial perivascular lymphocytic infiltrate. The direct immunofluorescence (DIF) showed linear deposits of C3 and C1q along the dermoepidermal junction. Indirect immunofluorescence was negative. There was a positive ANA (titre 1:40, homogeneous pattern). Anti-Jo-1, Ro and La



Fig. 1. Healed erosions over thighs.

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were negative. The arterial blood gas showed hypoxaemia and chest radiograph showed basal reticulonodular shadowing. A lung function test and high resolution computed tomogram of the thorax suggested the diagnosis of interstitial lung disease. Muscle enzymes and electromyogram were normal. Tumour markers (carcinoembryonic antigen, prostate specific antigen, alpha-fetoprotein, CA 19-9, faecal occult blood) were negative but a biopsy of a normal appearing post nasal space yielded a diagnosis of undifferentiated nasopharyngeal carcinoma. Over the following month, he developed progressive proximal muscle weakness over both upper and lower limbs. Together with this, the more typical cutaneous features of dermatomyositis such as Gottron's papules and poikiloderma also appeared. A diagnosis of bullous dermatomyositis associated with nasopharyngeal carcinoma was made and oral steroids were commenced. He is now co-managed by a rheumatologist and a respiratory physician, and is undergoing radiotherapy for the nasopharyngeal carcinoma.

Discussion

Dermatomyositis is an autoimmune disease affecting the skin and musculature, producing a typical cutaneous eruption as well as proximal muscle weakness. The most characteristic and possibly pathognomonic cutaneous features are the heliotrope rash and Gottron's papules/Gottron's sign. The former consists of a lilac or violaceous dusky erythema involving periorbital skin and the eyelids, which may occur with periorbital oedema. The heliotrope eruption follows disease activity. Gottron's papules (raised papules or plaques with erythema and scaling) or Gottron's sign (only erythema) occurs over bony prominences usually metacarpophalangeal joints or interphalangeal joints. They may also appear over other bony prominences like the elbows, knees and ankles. Other cutaneous signs, which are common but not specific to dermatomyositis, include periungual erythema or telangiectasia, photosensitivity, calcinosis and poikiloderma which consists of hyperpigmentation, hypopigmentation, telangiectasia and atrophy.¹

Bullous dermatomyositis was first reported in 1902 by Christen who described a patient with dermatomyositis presenting with "pemphigus-like" lesions.² Since then, there has been several reports of bullous dermatomyositis, especially in the Japanese literature.³ Blisters and erosions have been described over the usual areas affected by dermatomyositis such as the extensors of the arms and upper chest/back. To the best of our knowledge, this patient is the first reported case of bullous dermatomyositis with oral ulcers as well. The histology of bullous dermatomyositis is not specific. Usually a subepidermal blister is seen along with dermal oedema,

mucin deposition and a superficial perivascular lymphocytic infiltrate. Direct immunofluorescence is usually negative. The pathophysiology is thought to be due to intense dermal oedema causing blister formation.⁴ In a review of 19 reported cases of dermatomyositis with vesicle or bulla formation, all but one patient had subepidermal blisters on histology.³ DIF was positive in four patients. Three patients had DIF findings of IgA deposition in normal skin. These were patients with dermatitis herpetiformis who subsequently developed dermatomyositis.^{5,6} This was thought to be a common genetic predisposition to autoimmune disease due to the association with HLA B8 and DR3. One patient had bullous pemphigoid following dermatomyositis.⁷ This was believed to have resulted from disruption of the basement membrane zone leading to the exposure of the BP antigen. Our patient had DIF findings of linear deposits of C3 and C1q along the dermoepidermal junction. The significance of this is unknown. We were unable to obtain a biopsy of a blister in this patient. The incidence of malignancy in vesicobullous dermatomyositis is 55%, most of which were gynaecological cancers.³ This is higher than the risk of malignancy in the typical or non-bullous dermatomyositis (about 20% to 30%).³ This suggests that bullous dermatomyositis may be associated with a higher risk of malignancy, especially gynaecological cancers and hence, a worse prognosis. Local studies have found a high incidence of NPC associated with dermatomyositis, so routine screening for NPC is indicated in all patients.⁸⁻¹⁰

We present the first case of bullous dermatomyositis with oral ulcers. Bullous dermatomyositis should be considered in the list of differential diagnoses of blistering disorders, especially in the presence of other suggestive clinical features. This would prevent a misdiagnosis and an unnecessary delay in the search for an underlying tumour. Bullous dermatomyositis appears to be associated with a higher risk of malignancy and therefore more aggressive investigations are warranted. In addition to routine screening for tumours in the lungs, gastrointestinal tract, gynaecological system, breasts and prostate, an otorhinolaryngological referral for NPC screening is indicated for our local population.

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