

A rare Para neoplastic manifestation associated with squamous cell carcinoma of bladder: dermatomyositis

Songchao Li¹, Tieding Chen²,
Xiaokun Zhao³, Zhaohui Zhong⁴

ABSTRACT

Dermatomyositis is a rare Para neoplastic syndrome, especially in urinary tract malignancy. We report a case of bladder squamous cell carcinoma in a 50-year old patient who presented with Para neoplastic dermatomyositis when tumor recurred. A radical cystectomy was performed for him and symptoms of dermatomyositis improved dramatically. The case shows that complete resection of bladder cancer may lead to resolution of Para neoplastic dermatomyositis and to our knowledge it is the first case of dermatomyositis in association with squamous cell carcinoma of bladder reported.

KEY WORDS: Dermatomyositis; Squamous cell carcinoma; Bladder; Malignancy.

Pak J Med Sci July - September 2012 Vol. 28 No. 4 738-741

How to cite this article:

Li S, Chen T, Zhao X, Zhong Z. A rare Para neoplastic manifestation associated with squamous cell carcinoma of bladder: dermatomyositis. Pak J Med Sci 2012;28(4):738-741

INTRODUCTION

Dermatomyositis (DM) is an uncommon inflammatory myopathy characterized by proximal muscle weakness and distinct cutaneous eruption. It has been confirmed that DM is associated with an increased risk of malignant diseases. The reported frequency of malignancy in DM has varied from 6%-60% with most large population-based cohort

studies revealing a frequency of about 20%-25%.¹The cancers most strongly associated with DM are ovarian, lung, gastric, colorectal, and pancreatic cancers, along with non-Hodgkin's lymphoma.²Few reports have linked DM to bladder cancer, notably transitional cell carcinoma. We present a case of Para neoplastic DM associated with squamous cell carcinoma of bladder.

CASE REPORT

A 50-year-old man was diagnosed with squamous cell carcinoma after transurethral resection of bladder tumor (TURBT) in October 2007. However, he refused radical cystectomy for him. Regular adjuvant chemotherapy was carried out for him.

The patient was admitted to our hospital presented with 1-month history of facial rash, proximal muscle weakness and pain in upper extremities in May 2008. Physical examination revealed diffuse violaceous erythema and periorbital oedema affecting his face (Fig.1). The erythema also involved his upper chest and arms. The patient had bilateral upper extremities weakness graded at 4/5. Laboratory tests revealed: creatine phosphokinase (CK) 13750 u/l (normal range 24-195u/l), alanine transaminase (ALT) of 134u/l (normal range: 0-40u/l), aspartate

1. Songchao Li, MD, Department of Urology, The First Affiliated Hospital of Zhengzhou University, Zhengzhou, China.
2. Tieding Chen, MD, Department of Urology, Lihui Li Hospital, Ningbo, China.
3. Xiaokun Zhao, Professor,
4. Zhaohui Zhong, Assistant Professor,
- 3,4: Department of Urology, Second Xiangya Hospital, Central South University, Changsha, China.

Correspondence:

Zhaohui Zhong,
Assistant Professor, Department of Urology,
Second Xiangya Hospital, Central South University, No 139,
Renmin Road, Changsha 410011, China.
E-mail: zhongzhaohui0616@163.com

- * Received for Publication: October 18, 2011
- * Revision Received: February 27, 2012
- * Accepted for Publication: June 6, 2012

transaminase (AST) of 735u/l (normal range: 0-37u/l), C-reactive protein 31mg/l (normal range 1-8mg/l), normal anti-nuclear factor and normal extractable nuclear antigens. Urinalysis revealed microscopic haematuria. Electromyography showed evidence of an active myopathic process. A biopsy of the right deltoid muscle showed minor muscle fiber degeneration and a little inflammatory cell infiltrate. Ultrasonography of abdomen revealed a large mass in the patient's bladder. Subsequent evaluation with positron emission tomography/CT (PET-CT) revealed abnormal hyper metabolic foci extending through the bladder wall into perivesical adipose tissue and no distant metastasis. Cystoscopy showed a broad-based tumor measured about 3.0 x 2.5cm on the posterior wall of bladder. Based on a multidisciplinary experts' discussion, a clinical diagnosis of T3bN0M0 bladder cancer and Para neoplastic dermatomyositis was made.

Considering recurrence and local progression of the bladder cancer, he agreed with radical cystectomy at this time. Radical cystectomy and ileal conduit was carried out for him. We used methylprednisolone 40mg daily in the five days following surgery. Doses of 60mg oral prednisolone was used after his gastrointestinal tract recovered and slowly tapered. The patient recovered from the surgery without any complication. The level of CK decreased to 123U/L and muscle power and skin rash improved dramatically. The histopathologic examination of the mass showed a poorly differentiated squamous cell carcinoma (Fig.2) that invaded perivesical adipose tissue. Until the last follow up, the patient is still alive without

local recurrence or distant metastasis and has no recurrence of dermatomyositis.

DISCUSSION

Numerous cases of DM associated with malignancy have been reported but this association is much less common in urinary system. Bladder cancer has rarely been reported associated with DM. To our knowledge, only nine cases of bladder cancer associated with DM has been reported in published English data. Squamous cell carcinoma of bladder cancer is a rare disease with poor prognosis and has not been described in association with DM. This is the first case. A case of small cell carcinoma of bladder has already been reported by Sagi.³ The other reported types of bladder cancer associated with DM are transitional cell carcinoma except a case can't identify the cancer's type. The characteristics of those patients are summarized in Table-I.

The pathogenesis of DM is still poorly understood, but there is evidence to suggest that both immune and nonimmune mechanisms are involved in the pathogenesis of dermatomyositis.⁴ The immune mechanisms involve immune cells and their products, such as cytokines and antibodies.⁴ More recently, a novel autoantibody recognizing a 155kDa protein had been found in 75% malignancy-associated DM and another novel autoantibody recognizing 155 and 140kDa protein had been found in 71% malignancy-associated DM.^{5,6} The possibility exist that certain cancers may be involved in initiating and perpetuating the autoimmune response in dermatomyositis.



Fig.1: Erythematous rash and periorbital oedema on the patient's face.

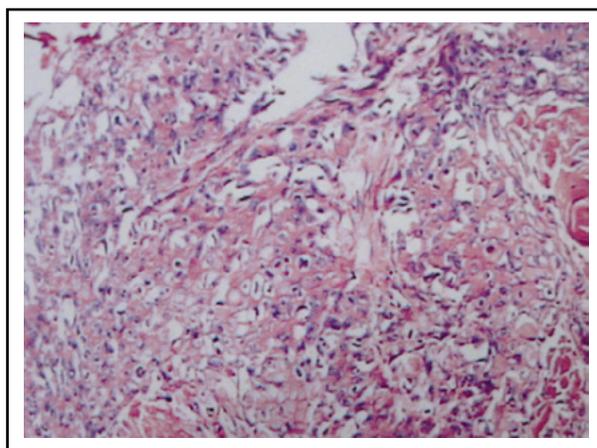


Fig.2: Pathological examination of bladder showed poorly differentiated squamous cell carcinoma. (H&E staining, original magnification×100)

Table-I: Characteristics of reported cases of bladder cancer associated with DM.

<i>Study</i>	<i>Age (years)/ Gender</i>	<i>Time of diagnosis of DM</i>	<i>Type of bladder cancer</i>	<i>Course of rash and muscles weakness</i>
Sabio	79/male	One year after Cancer	Transitional cell carcinoma	Significant improvement after TURBT and hormone therapy
Garcia-Donoso	60/male	Two weeks after cancer	Transitional cell carcinoma	Improvement within two months of chemotherapy and hormone therapy
Sagi	70/male	Simultaneous Carcinoma	Small cell	No obvious improvement after hormone therapy and chemotherapy
Federman	68/male	Thirteen months after cancer	Transitional cell carcinoma	Improvement of rash with hormone therapy
Hafejee	75/male	Seven weeks before cancer	Transitional cell carcinoma	Improvement after hormone therapy and immune therapy
Russ	71/male	Six months before cancer	Transitional cell carcinoma	Rash persisted after TURBT and hormone therapy
Talanin	62/male	Simutaneous	Sarcomatoid carcinoma? Or adenocarcinoma?	Improvement for a few days after radical cystectomy
Mallon	75/male	Three months before cancer	Transitional cell carcinoma	Significant improvement
Rankin	64/male	One year before cancer	Transitional cell carcinoma	Unknown
This study	54/male	Four months after cancer	Squamous cell carcinoma	Resolution of rash and weakness after radical cystectomy and hormone therapy

TURBT: transurethral resection of bladder tumor.

DM can be divided into 2 groups: Para neoplastic dermatomyositis and idiopathic dermatomyositis. Para neoplastic dermatomyositis is a kind of skin disorder that manifest an underlying internal malignancy. Appearance or worsening of a pre-existing DM indicating the relapse and extension of bladder cancer has been reported.⁷ Early recognition of this cutaneous presentation offers an opportunity for early diagnosis, treatment of the internal malignancy and monitoring for tumor recurrence.

It is now accepted that for a dermatosis to be recognized as a Para neoplastic manifestation; as suggested by McLean, it should satisfy two essential criteria: (1) The dermatosis must appear after the development of the malignant tumor and cannot be considered a Para neoplastic syndrome if it occurs before the tumor. (2) Both the dermatosis and the tumor should follow a parallel course.⁸

The diagnosis of DM can be done before, concurrently, or subsequent to bladder cancer (Table-I). Besides, it is not easy to demonstrate the parallelism between the bladder cancer and

DM unless the former has either been completely removed or has a relapsing stage. DM may follow the course of the malignancy (a Para neoplastic course) or follow its own course independent of the treatment of the malignancy.¹ The symptoms of DM persisted after the resection of bladder cancer has been reported.⁹ Consequently, not all the reported cases of DM associated bladder cancer belong to Para neoplastic syndrome.

In this case, our patient had several characteristics of a Para neoplastic manifestation. Firstly, DM appeared four months after the bladder cancer was diagnosed. Secondly, the DM relieved and has never recurred after the radical cystectomy. In the previous reported cases of bladder cancer associated with DM, only one patient reported by Talanin had radical cystectomy and the patient died about three months after the surgery because widespread bone metastases.¹⁰ The parallel course of bladder cancer and DM was sufficiently revealed in this case. According to McLean' criteria, the patient was considered to have paraneoplastic DM associated with bladder cancer.

The treatment of DM in a patient with cancer involves treating DM and treating the malignancy. Topical emollients and steroids are important in all patients and may control the skin lesions symptomatically until the tumour is treated and the dermatomyositis settles. However, if the tumour cannot be quickly treated radically, then the patient will probably require oral corticosteroids.¹⁰

In conclusion, we report a rare case of Para neoplastic DM associated with squamous cell carcinoma of bladder. Malignancies can manifest themselves on the skin. It is important for clinician to recognize DM as one of Para neoplastic dermatoses associated with bladder cancer and bladder cancer may be a rare cause of DM. Efforts should be made to detect underlying internal malignancy including bladder cancer to ensure early diagnosis and appropriate treatment. This case also shows that complete resection of the bladder cancer may lead to the resolution of Para neoplastic DM.

Source of funding: We declare that we had no financial support and conflicts of interest about this case report.

REFERENCE

1. Callen JP, Wortmann RL. Dermatomyositis. *Clin Dermatol* 2006;24:363-373.
2. Hill CL, Zhang Y, Sigurgeirsson B, Pukkala E, Mellekjær L, Airio A, et al. Frequency of specific cancer types in dermatomyositis and polymyositis: A population-based study. *Lancet* 2001;357:96-100.

3. Sagi L, Amichai B, Barzilai A, Shpiro D, Baum S, Neimushin A, et al. Dermatomyositis and small cell carcinoma of the bladder. *Can Fam Physician* 2009;55(10):997-999.
4. Zong M, Lundberg LE. Pathogenesis, classification and treatment of inflammatory myopathies. *Nat Rev Rheumatol* 2011;7(5):297-306.
5. Targoff IN, Mamyrova G, Trieu EP, Perurena O, Koneru B, O'Hanlon TP, et al. A novel autoantibody to a 155-kd protein is associated with dermatomyositis. *Arthritis Rheum* 2006;54(11):3682-3689.
6. Kaji K, Fujimoto M, Hasegawa M, Kondo M, Saito Y, Komura K, et al. Identification of a novel autoantibody reactive with 155 and 140 kDa nuclear proteins in patients with dermatomyositis: an association with malignancy. *Rheumatology (Oxford)* 2007;46:25-28.
7. Talanin NY, Bushore D, Rasberry R, Rudolph T, Tuli M, Friedman-Musicante R. Dermatomyositis with the features of inclusion body myositis associated with carcinoma of the bladder. *Br J Dermatol* 1999;141(5):926-930.
8. McLean DJ. Toward a definition of cutaneous paraneoplastic syndromes. *Clin Dermatol* 1993;11(1):11-13.
9. Russ BW, English JC 3rd, King DH, Halbach DP. Amyopathic dermatomyositis associated with transitional cell carcinoma of the bladder. *Cutis* 1999;63(2):73-76.
10. Chandiramani M, Joynson C, Panchal R, Symonds RP, Brown LJ, Morgan B, et al. Dermatomyositis as a paraneoplastic syndrome in carcinosarcoma of uterine origin. *Clin Oncol R Coll Radiol* 2006;18(9):641-648.

Contributions of each author:

Zhaohui Zhong and Xiaokun Zhao participated in the conception and revised this work; Songchao Li was responsible for collecting data and drafting this report; revising and follow up was done by Tieding Chen. All authors approved the version to be published.