Case Report

Dermatomyositis associated with adenocarcinoma of the lung: A case report

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Abstract

The association of dermatomyositis and underlying malignancies, especially small cell and squamous cell carcinoma of the lung, has been recognized a long time ago. We report the case of a 63-year-old male chronic smoker with adenocarcinoma lung associated with dermatomyositis. The occurrence of dermatomyositis should be considered as a potential presentation of paraneoplastic syndromes and physicians encountering similar clinical situations should perform extensive diagnostic work-up to exclude underlying neoplastic lesions.

Keywords: Dermatomyositis, lung cancer, paraneoplastic symptoms

Introduction

Dermatomyositis (DM) is a rare multi system autoimmune disorder, often triggered by drugs, infectious agents or underlying malignancy and clinically manifest with dermatological signs such as Gottron's papules, heliotrope rash, a diffuse violaceous erythema and systemic features, mainly, proximal muscle weakness.[1] The association of DM in adults with underlying malignancy is well documented and it is seen in upto 15% of all cancer patients.[2] Here, we report a case of a 63-year-old male patient with dermatomyositis secondary to underlying adenocarcinoma of the lung.

Case Report

A 63-year-old male chronic smoker was hospitalized with difficulty in getting up from sitting position and raising arm above head for the last eight months. On examination, there was redness over face, both shoulders and neck and weakness in the muscles of the upper and lower extremities. Blood investigations were unremarkable. Dermatological evaluation for the redness revealed gottron's papules, heliotrope rash, and a diffuse violaceous erythema [see Figure 1]. Dermatomyositis was diagnosed clinically and skin biopsy was sent for histopathological examination. The skin biopsy indicated atrophic epidermis with areas showing basal cell vacuolation and deem with mucin-like material in upper dermis with mild perivascular lymphocytic infiltration and few scattered melanophages, consistent with dermatomyositis. The muscle biopsy results showed features of inflammatory myopathy with vasculitis. Electromyography (EMG) showed short duration low amplitude MUAPS suggestive of myopathic potential.

The chest radiography obtained at the time of diagnosis showed right upper lobe mass with right hilar and paratracheal lymphadenopathy. The chest computed tomography (CT) confirmed the presence of a small irregular nodular soft tissue density lesion with speculated margins, surrounding ground glass densities, and interloper septal thickening in posterior segment of right upper lobe with mediastinal lymphadenopathy,

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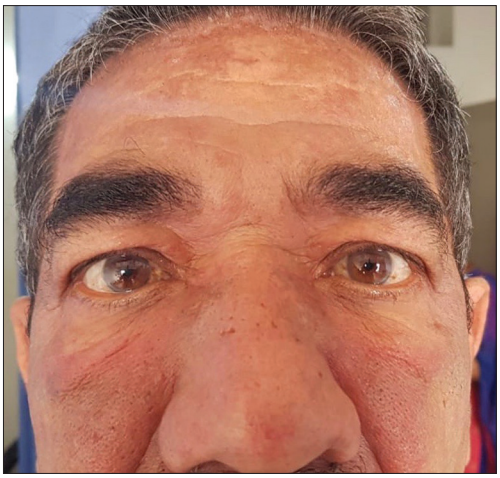

Figure 1: Typical skin rash on the face of the patient with dermatomyositis

possibly neoplastic process. Endobronchial Ultrasound (EBUS) needle biopsy of the lesion confirmed poorly differentiated adenocarcinoma. Patient was managed symptomatically and referred to oncology center after the confirmation of the diagnosis.

Discussion

The association of DM with malignancies has been consistently reported since 1916 with standardized incidence ratios (SIRs) typically ranging from 3 to 6.[3,4] DM is more commonly reported in older patients with small cell and squamous cell carcinoma of the lung when compared to other malignancies and age group.[5] The other malignancies commonly found to be associated with DM were colorectal, cervical, ovarian cancer, and non-Hodgkin lymphoma.[6] The symptoms of DM usually precede the diagnosis of cancer and the usual interval between DM and the diagnosis of internal malignancies was less than 1 year.[3]

In the current case, DM presented as a para-neoplastic syndrome accompanied by poorly differentiated adenocarcinoma of the right upper lobe. A histological classification of 48 cases of DM-associated lung malignancies reported in the literature till 2017 found small cell carcinoma in 43.8%, followed by squamous cell carcinoma (16.7%), adenocarcinoma (16.7%), neuroendocrine (6.3%), undifferentiated (4.2%), and others (12.5%).[4]

The immune complex deposition in the capillary beds, inflammatory cell accumulation, T cell-mediated myotoxicity, complement-mediated microangiopathy, and resulting structural changes of the myocytes characterize DM.[7] The pathophysiology of the relationships between neoplastic process and DM is not completely known. However, the role of the depolymerized glycosaminoglycans released from the extracellular matrix surrounding the tumor, substance secretion by neoplastic cells, cross-reaction to newly presented 'exposed' antigens, angiopathy, viral infections facilitating abnormal immunologic response and humoral and cell-mediated immunological processes have been hypothesized.[7]

The diagnosis of DM is made by careful clinical history, examination of proximal muscle weakness, skin, and muscle biopsy and laboratory investigations such as serum muscle enzyme concentrations and autoantibody tests (Anti-Jo-1).[1] According to Bohan and Peter, the presence of 3-5 of the following symptoms confirms the diagnosis of dermatomyositis: progressive symmetric muscle weakness of the inferior and superior extremity girdle with dysphagia (or not) or with respiratory muscle involvement, muscle biopsy confirming myositis, increase in the muscle enzyme serum levels, electromyogram (EMG) abnormalities indicating a primary muscle damage and characteristic skin lesions.[8] Our patient had progressive symmetric muscle weakness, characteristic skin lesions, muscle biopsy finding constant with myositis and EMG finding suggestive of myopathic potential. However, CPK value was closer to normal. A past study found that a lack of creatine kinase elevation in patients with dermatomyositis to be a poor prognostic sign.[9]

The treatment of DM is largely based on controlling the likely autoimmune component of the disease, in our case, the treatment of the underlying malignancy and prognosis largely depends on the prognosis of the malignant diseases. However, high-dose steroids, methotrexate, azathioprine, and mycophenolate mofetil are also commonly used in the management of DM.[1]

In conclusion, our case highlights the association of DM with adenocarcinoma lung in an elderly male chronic smoker. Physicians encountering similar clinical situations should perform extensive diagnostic work-up to exclude underlying neoplastic lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Shen C, Che G. Dermatomyositis as an antecedent sign of lung cancer in an elderly patient: A case report. J Thorac Dis 2014;6:E15-8.
2. Heinemann S, Zabel P, Hauber H. Paraneoplastic syndromes in lung cancer. Cancer Ther 2008;6:687-98.

3. Fujita J, Tokuda M, Bandoh S, Yang Y, Fukunaga Y, Hojo S, *et al*. Primary lung cancer associated with polymyositis/dermatomyositis, with a review of the literature. Rheumatol Int 2001;20:81-4.
4. Beel AJ, Demos DS, Chung A, Liao C, Lui NS. Ground-glass opacity heralding invasive lung adenocarcinoma with prodromal dermatomyositis: A case report. J Cardiothorac Surg 2018;13:20.
5. Antiochos BB, Brown LA, Li Z, Tosteson TD, Wortmann RL, Rigby WF. Malignancy is associated with dermatomyositis but not polymyositis in Northern New England, USA. J Rheumatol 2009;36:2704-10.
6. Hill CL, Zhang Y, Sigurgeirsson B, Pukkala E, Mellemkjaer L, Airio A, *et al*. Frequency of specific cancer types in dermatomyositis and polymyositis: A population-based study. Lancet 2001;357:96-100.
7. Zhang X, Wang Y, Ma G, Zhang L, Jing H, DU J. Dermatomyositis as a symptom of primary lung cancer: A case report and review of the literature. Oncol Lett 2016;11:3413-6.
8. Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). N Engl J Med 1975;292:344-7.
9. Fudman EJ, Schnitzer TJ. Dermatomyositis without creatine kinase elevation. A poor prognostic sign. Am J Med 1986;80:329-32.