A case of Hypomyopathic Dermatomyositis which subsequently developed to overt Myositis with ILD

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ABSTRACT

Dermatomyositis (DM) is an inflammatory myositis. Clinically amyopathic dermatomyositis (CADM) and hypomyopathic dermatomyositis (HDM) are rare forms of DM in which skin manifestations are present with no and minimal clinical and laboratory evidence of myositis respectively. A common complication of CADM/HDM is interstitial lung disease (ILD), with the most common histopathologic variant of non-specific interstitial pneumonia (NSIP). This case highlights the development of ILD and severe muscle weakness, including proximal limb muscles, muscles of respiration and muscles of deglutition in a patient with previous diagnosis of hypomyopathic dermatomyositis.

Keywords: Clinically Amyopathic Dermatomyositis (CADM), Dermatomyositis (DM), Hypomyopathic Dermatomyositis (HDM), Interstitial Lung Disease (ILD), Non-specific Interstitial Pneumonia

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INTRODUCTION

A young female presenting with skin rashes, arthritis, shortness of breath, and weakness of proximal muscles strikes a physician with a handful of differentials in the spectrum of connective tissue disorders like SLE, dermatomyositis (DM), mixed connective tissue disorder (MCTD), undifferentiated connective tissue disorder (UCTD) or sarcoidosis. Diagnosis and treatment, if delayed, may lead to rapid deterioration with high morbidity and mortality. DM may involve multiple organ systems like skin, muscles, joints, respiratory system, cardiovascular system or gastrointestinal system. An atypical form of dermatomyositis is hypomyopathic dermatomyositis. Clinically amyopathic dermatomyositis (CADM) accounts for approximately 20% of dermatomyositis, which includes amyopathic dermatomyositis (ADM) and hypomyopathic dermatomyositis (HDM)[1,2]. The former refers to characteristic cutaneous manifestations without evidence of myopathy for two or more years. The latter refers to characteristic cutaneous manifestations of dermatomyositis with no clinical symptoms of muscle involvement, normal or elevated serum creatine kinase levels, subclinical evidence of muscle involvement determined by electromyogram (EMG), muscle biopsy and imaging examinations, and a medical history of more than six months[3]. Interstitial lung disease (ILD) is a common complication of amyopathic/hypomyopathic DM[4]. Here, we present a case of previously diagnosed hypomyopathic dermatomyositis which subsequently developed to overt myositis with severe weakness of proximal muscles of bilateral upper and lower limbs and features of ILD.

CASE DESCRIPTION:

A 30-year-old female was diagnosed as hypomyopathic dermatomyositis on the basis of inflammatory polyarthritis, skin rashes over face (malar rash and heliotrope rash) [Fig. 1], neck (shawl sign) and hands (gottron papules) and occasional mild proximal weakness of both lower limbs 3 months back. There was no objective evidence of muscle weakness and muscle enzymes, nerve conduction study (NCS) and EMG were normal at the time of diagnosis. She was then put on Methotrexate (MTX) 15 mg/week, titrated up to 25 mg/week and a NSAID with good response to skin rashes as well as joint pain.

After 6 months, she presented with an increase in muscle weakness, rapidly worsening shortness of breath, cough with sputum production, difficulty in swallowing and fever for one week. She noticed weakness of limbs on household activities, restricting her movement over the duration of 2-3 days. Shortness of breath, which was gradually progressive (MRC grade III) initially progressed to abrupt onset even at rest for 2 days associated with cough and sputum production. Dysphagia with nasal regurgitation was also present. There was a history of fever with the highest recorded temperature of 102˚F for 1 week. She also mentioned significant weight loss of ~5 kg over the duration of the last 3 months. There is no past or contact history of tuberculosis infection.

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On examination, the patient was dyspneic, anemia was present, and multiple hyperpigmented skin macules and patches were present in face. Pulse rate was 100 beats/min, respiratory rate- 30/min, temperature 100˚F, BP- 90/60 mmHg, and SPO2- 85% without oxygen. Chest examination revealed fine inspiratory crepitations over bilateral basal lung fields. Examinations of other systems were normal.

Investigations showed- Hb.- 13.2 g/dl, ESR- 45 mm in 1st hour, total WBC count- 12000 with neutrophils 75%, CRP was 0.378 mg/dl, Urea- 12 mg/dl, S. creatinine- 0.69 mg/dl, SGPT- 98 U/L, SGOT- 233 U/L, chest X-ray showed B/L pulmonary patchy infiltrations [Fig. 2], Montoux test was negative. Sputum for Gram's staining, acid fast bacilli and culture were normal. Blood culture was negative for any organisms. Renal function tests were normal; muscle enzymes were elevated with CK- 476 (previous reports were normal) and LDH- 331. HRCT showed ground glass opacities and was suggestive of diffuse parenchymal lung disease with volume shrinkage of the left lung and left pleural thickening [Fig. 3]. Cardiac screening with ECG and echocardiography were normal. NCS and EMG initially showed normal findings, and were not repeated. Muscle biopsy was not done. No biochemical or radiological evidence of malignancy was found.

Fig. 1

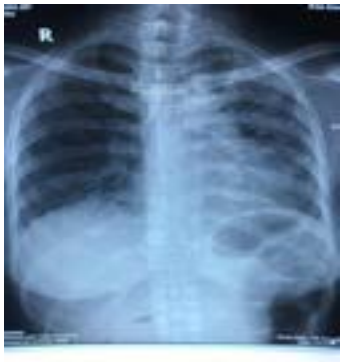

Fig. 2

The patient was further treated with oxygen, NG tube feeding, IV methylprednisolone 1 gm/day for 3 days followed by pulse cyclophosphamide 1gm/month followed by oral prednisolone 1mg/kg/day. Patient had significant improvement after getting first pulse cyclophosphamide therapy with decrease in symptoms and increase in oxygen saturation levels. She was discharged on oral prednisolone 1 mg/kg/day for 1 month followed by further tapering, and monthly IV cyclophosphamide for 6 months. On her next follow-up at 15 days, she had significant improvement of muscle power, dysphagia, nasal regurgitation, and shortness of breath. She was doing well till 4 months of her follow-up, when she had an increase in her symptoms and got herself admitted in our center again. Shortness of breath was present even at rest with dysphagia and nasal regurgitation reappeared. Supportive treatment with oxygen, NG tube feeding, and IV methylprednisolone 1 gm/day for 3 days was given. She denied ventilator support. After 1 week of hospital stay, the patient expired.

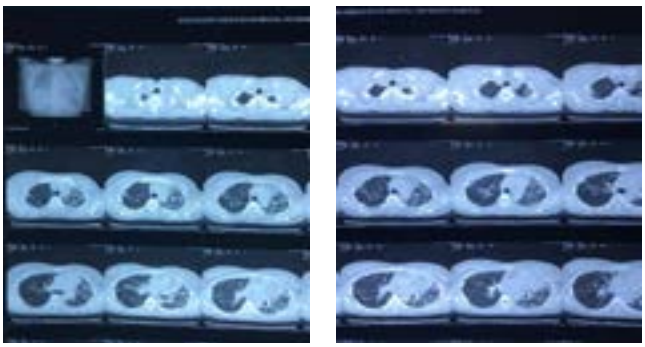

Fig. 3

DISCUSSION:

CADM is considered to be different from classic dermatomyositis, for its peculiarity for rapid development of interstitial lung disease[4]. However, there are certain degrees of muscular fiber damage in HDM subtypes. Characteristics of its pathological changes and whether it has a relationship with classic dermatomyositis remains to be determined.

Regarding development of shortness of breath on exertion with high grade fever, cough with sputum production and leukocytosis in this patient, we initially had some other differentials. They were : (a) ILD with superimposed pneumococcal pneumonia (b)ILD with pneumocystis carinii pneumonia and (c) MTX induced lung injury. Pneumococcal pneumonia was excluded after negative microbiological findings of sputum and pneumocystis carinii pneumonia was excluded after negative giemsa stain. MTX induced lung injury most commonly occurs during the first year of treatment and with high dose of MTX[5]. Presentation is with cough, dyspnea, fever and inspiratory crepitation within 1-5 months of therapy[6]. Investigations reveal hypoxemia with peripheral leukocytosis and eosinophilia in some patients. Patients on low-dose methotrexate are at increased risk for opportunistic infections, especially pneumocystis carinii pneumonia[7]. Although the findings were equally supportive of MTX induced lung injury, and the patient also fulfilled the criteria[8], we excluded this diagnosis on the basis of failure of improvement of symptoms even after withdrawal of MTX and starting pulse corticosteroid.

One of the other issues in this patient is regarding treatment of refractory DM. In patients with ILD and DM, combination of agent like cyclophosphamide or rituximab with initial glucocorticoid has shown good efficacy[9,10]. In this scenario, another option could be IVIG, which has a good efficacy in the improvement of muscle strength, however the effect is temporary and repeated transfusions may be required[11].

CONSENT

Written informed consent for publication of her clinical details and clinical images was obtained from the patient.

Place of study: Department of Rheumatology, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh

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