

Life threatening Oropharyngeal Aphagia in a Patient of Dermatomyositis

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ABSTRACT

Dermatomyositis is an inflammatory myopathy characterized by proximal muscle weakness resulting from idiopathic chronic inflammation and typical cutaneous changes like heliotrope rash over eyelids and Gottron's papules. It is also characterized by multiple systemic symptoms like arthralgia, arthritis, dyspnea, dysphagia, arrhythmia, and dysphonia. In majority of adults it occurs as a manifestation of Para neoplastic syndrome. Here we present a 43 years old patient presenting with oropharyngeal aphagia. As a important manifestation of dermatomyositis

Introduction :

Dermatomyositis (DMS) is a rare inflammatory disease with musculoskeletal manifestations. It is characterized by progressive proximal symmetrical weakness, elevated muscle enzymes, abnormal electromyogram, abnormal muscle biopsy and classical cutaneous findings.

Case Report :

A 43 year old male, chronic alcoholic presented with 1 month history of gradually progressive weakness in proximal muscles in the form of difficulty in getting up from squatting position, difficulty in overhead abduction of arms, weakness was symmetrical and without any sensory involvement. Patient also gave history of progressive swelling and erythematous non-itching rash around eyes involving eyelids, dysphagia and rash over face and both upper limbs since 20 days. All the symptoms were progressively increased till the time of hospitalization. He gave history of stopping alcohol since last one month. There was no significant past history of any medical or surgical history or hospitalization.

On examination, he was conscious, oriented and his vitals were stable. He was pale but had no icterus,

cyanosis or lymphadenopathy. He had edema which was restricted to upper limbs.

He had a typical erythematous violaceous rash over both upper eyelids and peri-orbital region along with edema ("Heliotrope Rash"). Rash was non-itching, non-tender associated with some scaling of skin. (*Fig. 1 & 2*).

Figure 1



Figure 2



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He had erythematous macular non-pruritic rash around neck and shoulders, aggravated in sunlight (“Shawl Sign”) associated with edema over both arms (*Fig. 3*). In addition he had some form of vasculitic lesions over both shoulders and elbows. However there was no evidence of any papular lesions over fingers (Gottron’s Papules).

Figure 3



On neurological examination, his higher functions were normal. He had pooling of secretions in throat and nasal twang to voice. Motor system examination revealed normal power distally and grade III power at muscles acting at elbow, shoulder, knee and hip joints bilaterally. Weakness was accompanied by tenderness in both arms and hips. Tone and deep tendon reflexes were normal. Respiratory system examination revealed bilateral throat conducted sounds; cardiovascular and abdominal examination was normal.

Laboratory analysis showed elevated acute phase reactants (ESR: 45, CRP: 5.0), elevated creatinine phosphokinase (1274 U/L) and elevated serum transaminases. Electromyography showed increased spontaneous muscle activity with fibrillations, full recruitment, and low amplitude polyphasic units of short duration compatible with myositis. Skin punch biopsy revealed mild chronic inflammation with thinning of epidermis not consistent with DM, however muscle biopsy was avoided due to high risk of infection from high steroids. The anti-JO1 antibody titre was negative. Screening for neoplasm included chest x-ray, upper GI endoscopy, CT Neck+Thorax+Abdomen, all of

which were normal. On the basis of these findings diagnosis of dermatomyositis was finalized and he was started on oral Prednisolone 1mg/kg/day.

On the fifth day of hospitalization, during the diagnostic work-up, his swallowing difficulty worsened and he developed aphagia and respiratory compromise and started desaturating. Patient’s single breath count (SBC) was normal, SPO₂ was 84% at room air, arterial blood gases revealed mild hypoxemia with hypercapnia. Patient was intubated and observed on T-piece, steroids were stepped up with IV Methylprednisolone 1gm/day for three days, Methotrexate 10mg/week. Patient recovered within a span of 4 days, patient was extubated on 5th day post intubation, without requiring mechanical ventilation, dysphagia improved over few weeks duration with disappearance of skin rash.

Discussion :

Dermatomyositis is manifested by a symmetric proximal inflammatory myopathy and a characteristic violaceous, cutaneous eruption in classical areas. In addition to involving the skeletal muscles, dermatomyositis is known to involve the respiratory muscles, the myocardium and the gastrointestinal tract. Patients often present with progressive difficulty in activities that involve proximal muscles. Fine movements involving distal muscles are only involved late in the course. Ocular muscles are spared, even in advanced, untreated cases. However, pharyngeal and neck flexors muscles are often involved, resulting in varying degree of dysphagia and head drop¹.

It is estimated that 10-73% of patients with inflammatory myopathies suffer from dysphagia to some extent². However a study conducted in 2010 by Marie I et.al involving 73 cases established that ‘aphagia’ defined as dysphagia to both solids and liquid is relatively rare (~7%). As we know, most causes of aphagia are esophageal³.

Expecting a correlation between limb strength and severity of the oropharyngeal dysphagia can also be misleading. As in our patient, patient had grade III power even proximally in the limbs but had complete oropharyngeal aphagia; Kim and colleagues, who found no correlation between the

hyolaryngeal movement and the motor power of the limb muscles⁴, recently described the dissociation between the two.

Respiratory involvement in a patient with inflammatory myopathies is mostly in the form of interstitial lung disease, or in rare cases resulting from respiratory muscle weakness. What was peculiar in our case is the fact that there was no lung pathology detected on CT scan of chest, patient's single breath count never decreased; despite of this patient had severe respiratory compromise which was easily managed just by securing the airway by endotracheal intubation without the need of mechanical ventilation; to add to the insult was the aspiration of secretions which patient was unable to clear due to oropharyngeal weakness. Among factors associated with poor survival in dermatomyositis are advanced age, malignancies, delayed initiation of corticosteroid treatment, respiratory muscle weakness, lung and myocardial involvement. Our patient had none of the above yet oropharyngeal aphagia had proved almost fatal for him.

In conclusion, in cases of dermatomyositis, awareness of possible oropharyngeal aphagia as an early life-threatening complication of dermatomyositis is essential and may justify the switch from oral to IV medication delivery as soon as possible and is vital for patient survival.

References :

1. Harrison's Principles of Internal Medicine, 19th edition.
2. Oh TH, Brumfield KA, Hoskin TL, Stolp KA, Murray JA, Bassford JR. Dysphagia in inflammatory myopathy : clinical characteristics, treatment strategies, and outcome in 62 patients. *Mayo Clin Proc* 2007; 82 (4): 441-7.
3. Marie I, Menard JF, Hatron PY, et al. Intravenous immunoglobulins for steroid-refractory esophageal involvement related to polymyositis and dermatomyositis : a series of 73 patients. *Arthritis Care Res* 2010; 62 (12): 1748-55.
4. Kim SJ, Han TR, Jeong SJ, Beom JW. Comparison between swallowing-related and limb muscle involvement in dermatomyositis patients. *Scand J Rheumatol* 2010; 39: 336-40.