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RESEARCH ARTICLE

DERMATOMYOSITIS IN MALES AND IN ASSOCIATION WITH CARCINOMA OESOPHAGUS

Dr. Madhu Kaushal[1], Dr. Bimal Kumar Singh[2], Dr. Priyanka Tolani[3] and Dr. Sunil Nagar[4]

1. MBBS, M.D (Medicine) ACHD (Physician) Northern Railway Central Hospital (NRCH), New Delhi.
2. MBBS (Hons) DNB Medicine, Chief Physician Head of Department NRCH, New Delhi.
3. DNB Medicine Department of General Medicine NRCH, New Delhi.
4. Senior Consultant and Unit Head of Department of Interventional Pulmonology and Sleep Medicine, Accord Hospital, Faridabad.

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Abstract

Dermatomyositis is a **rareinflammatorymyopathy**, frequently associated with ovarian, breast, lung and nasopharyngeal cancers. A very few cases have been reported of dermatomyositis in males and in association with carcinoma oesophagus. We report one of such rare presentation.

Introduction:-

Dermatomyositis is an inflammatory disorder of skin and muscles. Its incidence is found to be 2-19 cases per million. **Skin involvement** in DM usually manifests with characteristic papules over digits, erythema over the elbows and knees, a heliotrope rash around the eyes, periungual telangiectasias, and dystrophic cuticles. **Muscle involvement** usually manifests as proximal muscle weakness. There is a well-established association of DM with an increased risk of **internal malignancy** mainly ovarian cancer, breast and lung cancers. Rarely is it found to be associated with oesophageal cancers.

Case report:

We report case of a 67 year old male patient who presented to the outpatient department with thehistory of **rashes** over both hands and forearms, face, upper back and neck for the last 20- 25 days and **tightness and weaknessof both upperlimbs** for the last 15-16 days. The rashes red in colour scaly in nature were initially observed by the patient over dorsal surface of hands and interphalangeal joints which then spread to forearm, extensor surface of elbow, upper back, neck, forehead and eyelids. They were mildly pruritic. One week **post development of rashes** patient started complaining of tightness and weakness of both arms. The weakness was gradually progressing and the patient was now unable to lift his arm above head, comb his hair or pick up any object above the level of his shoulders. The patient could however hold his pen firmly, had normal hand grasp and could easily type on his phone. Patient also gave history of difficulty in holding his head upright which he developed later in the course of his illness. There were however no complaints of difficulty in walking, getting up from sitting or supine posture, holding his slippers, any numbness or tingling sensation, any abnormal heat or cold intolerance, any visual disturbance. Along with weakness he also developed swelling and stiffness of both upper arms and facial puffiness especially periorbital. In view of these complaints he presented to our OPD.

The patient is a post operative case of **carcinoma esophagus** (T3N1). He was diagnosed in December 2015 as a case of squamous cell carcinoma esophagus on endoscopy followed by biopsy done for complaints of dysphagia and

Corresponding Author:- Dr. Madhu Kaushal
Address:- MBBS, M.D (Medicine) ACHD (Physician) Northern Railway Central Hospital (NRCH), New Delhi.

weight loss. He was initially given neo-adjuvant chemotherapy, later operated (Mac Ewansesophagectomy and celiac lymph node dissection) and followed by adjuvant chemotherapy. His latest PET CT (Positron Emission Tomography- Computed tomography) done in September 2016 showed that the esophageal lesion and paraesophageal lymph node are no longer visualized. However, mediastinal lymph nodes and lung opacities were new findings and abdominal lymph nodes had increased in size compared to previous scan.

Our patient had vitiligo which he said was present since birth. He was conscious cooperative. His pulse was 92/min, blood pressure124/80 mm Hg, SpO2 98% at room air and was afebrile to touch. Erythematous and patchy scaly lesions were present predominantly over dorsal surface of fingers, extensor surfaces of both forearms along with elbow joint, over nose, malar prominences, forehead, periorbital, few over arms and posterior surface of neck and ears and back mainly upper. Few white coloured lumps subcutaneously were also present over both forearms. Mild periorbital puffiness was present. No pedal oedema. A 2*1 cm mobile lymph node was palpated in the upper cervical region right and 1.5 * 1cm left upper cervical region. No other lymphadenopathy.

Respiratory and cardiovascular system examination was within normal limits. On inspection of the abdomen two scars were present consistent with past history of oesophagectomy. Rest of the abdominal examination was also normal. On nervous system and locomotor examination grade 4 – power was present in muscles of left upper limb especially upper arm muscles biceps brachii, triceps and deltoid and 4 + in muscles of right arm. Tone, coordination was normal. Also power in rest of the muscles was also 5/5.Reflexes both superficial and deep were normal. Sensory system examination was within normal limits. No signs of cerebellar or autonomic dysfunction were noted.

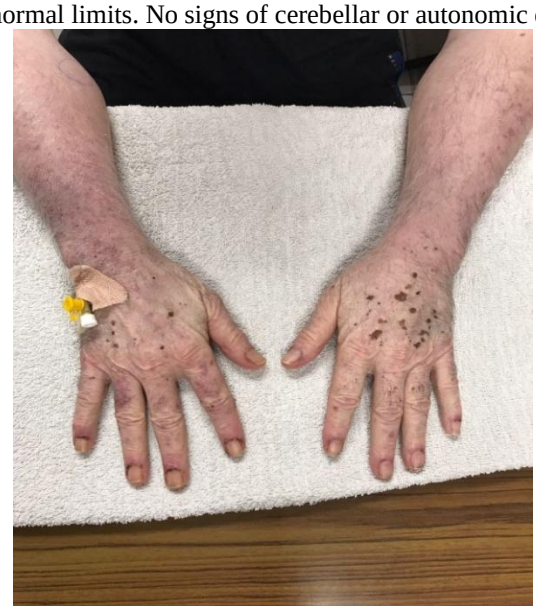

Figure 1:- Figure showing gottron's papules.

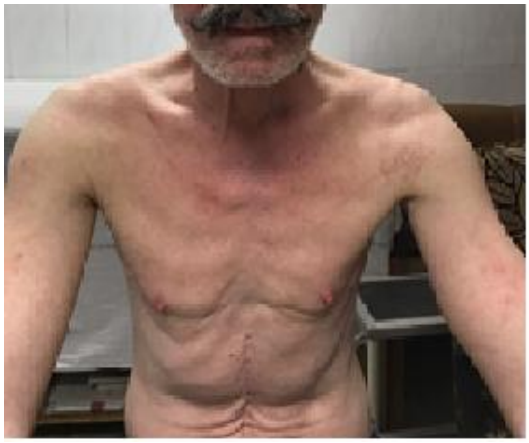

Figure 2:- Picture showing erythema over anterior chest (V sign).

Figure 3:- Gottron's sign.

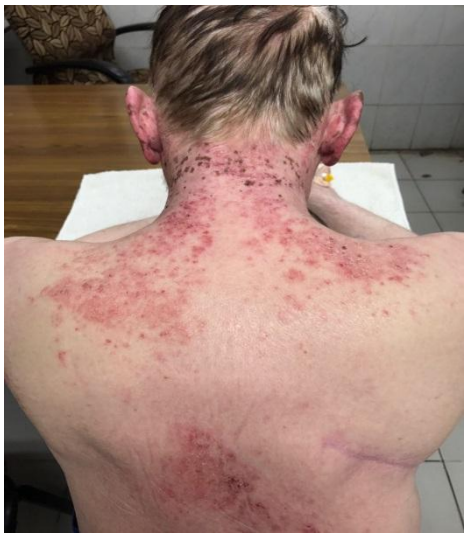

Figure 4:- Picture showing Shawl sign.

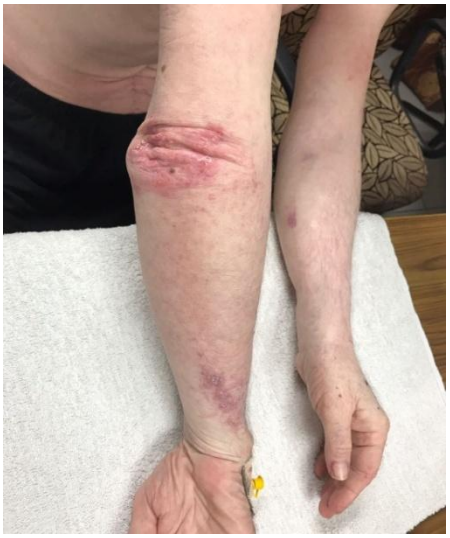

Figure 5:- Rough, cracked hands like mechanic's hands and gottron's sign.

On the basis of present and past history, general and physical examination, we made a probable diagnosis of **dermatomyositis as paraneoplastic syndrome of carcinoma esophagus.**

Hemogram was normal. S. urea was 47.7 and s. creatinine was 1.03 mg/dl. Total bilirubin was 0.45 mg/dl, SGOT/AST(serum glutamic oxaloacetic transaminase /aspartate aminotransferase) -632 U/l, SGPT/ALT (serum glutamic pyruvic transaminase /alanine aminotransferase)-107 U/l, ALP (Alkaline Phosphatase)-89 U/l, total protein- 6.62 g/dl, albumin-2.76 g/dl, globulin-3.86 g/dl and A/G (albumin/globulin) ratio 0.7. serum electrolytes within normal range. Serum CPK (creatinine kinase) -8832 U/L ,Serum LDH (lactate dehydrogenase)– 978 U/L. ANA (Anti-nuclear antibody) was positive in titre 1:160. Rest of the ENA (**Extractable Nuclear Antigens** Antibodies) profile was negative including Jo-1 antibody. Sonography of abdomen revealed multiple large retroperitoneal lymph nodes in porta, para-aortic and aortic region measuring upto 4.5 cm. Spontaneous activity from bilateral deltoid and supraspinatus with early recruitment in right supraspinatus and deltoid with polyphasic MUAPs suggesting myopathic pattern with fibrillations was detected on needle electromyography.

We gave three days of intravenous methyl prednisolone and patient responded well. His rashes decreased and power of muscles also increased. He is now planned for next cycle of chemotherapy for carcinoma oesophagus.

Figure 6, 7, 8:- All these figures depict clearance of rashes post three day course of intravenous steroids.

Discussion:-

Inflammatory myopathies represent largest group of acquired however potentially treatable causes of skeletal muscle weakness. They comprise of three major groups: polymyositis (PM), dermatomyositis (DM) and inclusion body myositis (IBM). The prevalence of inflammatory myopathies is estimated as 1 in 100000. These disorders present as progressive and symmetric muscle weakness except in IBM which can have an asymmetric pattern. Patients initially have proximal muscle weakness. Distal muscles are affected only late in the course of illness exception being

inclusion body myositis where distal muscles are affected fairly early. Ocular muscles are spared even in advanced untreated disease. Hence, the diagnosis of inflammatory myopathy must be questioned if ocular muscles are involved.[1]

Dermatomyositis is a rare type of inflammatory myopathy whose incidence is estimated to be between 2 and 19 cases per million population. It affects females almost twice as frequently as it does males. The age distribution is bimodal with two peaks , one at 10 years of age in case of juvenile DM and the other between 40 and 60 years of age in adult DM.[2,3]

It is a distinctive entity identified by characteristic rash which may accompany or more often precedes muscle weakness. The **skin manifestations** described in the literature are violaceous or blue-purple discolouration of upper eyelids with edema (Helioptrope rash), erythematous to violaceous papules and plaques over the extensor surfaces of the metacarpophalangeal and interphalangeal joints (Gottron's papules), erythematous macules and patches overlying the elbows and/or knees (Gottron's sign),V sign, Shawl sign, Mechanic's hands, Flagellate erythema, calcinosis etc.[4,5]**Muscle weakness**is characteristically initially proximal but later may involve the distal group also.Due to involvement of oropharyngeal striated muscles and upper oesophagus, patients may also experience dysphagia. Head drop also can occur as a result of involvement of neck flexor muscles. In severe cases respiratory muscles are also affected and respiratory failure occurs. Sensation and tendon reflexes however, are preserved. Myalgia and muscle tenderness is occasionally present in a small number of patients. There is a small subset of patients who clinically have neither muscle weakness nor significant muscle enzyme elevation. These patients are termed as having clinically amyopathic dermatomyositis (CADM).[1,4]

Pulmonary dysfunction mainly interstitial lung disease which develops in upto 10% patients of DM and may precede myopathy or occur early in the course of the disease and deforming arthropathy. Overlap syndrome occurs in patients of DM with features of systemic sclerosis or mixed connective tissue disease.[1]

In DM, humoral immunopathologic mechanism is implicated in contrast to PM and IBM in which T cell mediated cytotoxicity is likely. Perifascicular atrophy is the characteristic of dermatomyositis, even in the absence of inflammation.[1]

ANA (antinuclear antibodies)titers are found to be raised in sera of around 80 % patients of DM. The myositis-specific autoantibodies (MSAs) include anti-synthetase group, the most important of which is anti Jo-1 (histidyl-tRNA synthetase) antibodies, anti Mi-2, and anti-SRP antibodies. Antibodies to nuclear matrix protein NXP2 (MJ Ab), are MSAs found in juvenile as well as paraneoplastic DM. 75% patients with paraneoplastic DM also demonstrate the presence of new anti-TIF1γ (formerly termed anti-155/140) antibodies.Anti-MDA5 (or anti-CADM-140) antibodies are strongly associated with rapidly progressive interstitial lung disease in patients with amyopathic dermatomyositis.[6,7,8,9]

Paraneoplastic DM was first described in 1916 by Sterz who noted a tendency toward female predominance, particularly after their fifth decade of life. The most common associated underlying neoplasm is ovarian cancer. Other frequently associated malignancies include breast, colon, lung, prostate, pancreatic, and gastric carcinomas as well as non-Hodgkin's lymphoma. The tumor most frequently associated with paraneoplastic dermatomyositis in Southeast Asia is nasopharyngeal cancer.[10,11]

Very rarely has oesophageal cancer been found to be associated with dermatomyositis. Few case reports of this association have been published.[12,13] There have been a few reports of recurrence of oesophageal cancer presenting as dermatomyositis.[14] Dysphagia is a symptom common both to carcinoma oesophagus as well as dermatomyositis.In the setting of an underlying malignancy, these symptoms can be misleading and one can miss the diagnosis of dermatomyositis. However, recognition of the characteristic skin rash may provide a clue to the diagnosis.[12]

In cases of dermatomyositis that arise in patients previously diagnosed with cancer, the diagnosis of paraneoplasia is implicit in the diagnosis of the dermatomyositis, and screening studies are not indicated. However, in cases of adult dermatomyositis in patients with no previous diagnosis of cancer, the dermatologist is obliged to determine whether or not the patient has an associated neoplasm. It appears that the combination of the 2 concepts—the absence of

antibodies traditionally associated with myositis like anti-Jo-1, anti-Mi-2, and anti-U1-RNP and the presence of anti-155/140 antibodies—provides a sensitivity and a negative predictive value of almost 100% in dermatomyositis.[15]

Regarding the management of paraneoplastic dermatomyositis, our aim must always be to control the underlying neoplasm. Patients in whom tumour response was complete presented spectacular improvements or complete remission of the dermatomyositis. When oncologic treatment is not possible or does not achieve a complete response, and in those cases in which anticancer therapy is delayed, the treatments most widely used to control the dermatomyositis are oral corticosteroids at a dose of 0.5-1mg/kg/d. If there is no adequate response or if the treatment has to be prolonged, methotrexate can be beneficial at doses between 7.5 and 15mg per week.[15]

Conclusion:-

Dermatomyositis is a rare inflammatory myopathy. It can present in various forms like classic, amyopathic, paraneoplastic, associated with connective tissue disease (overlap syndrome) etc. Paraneoplastic dermatomyositis is mostly found to be associated with nasopharyngeal cancers, ovarian, breast, colon, lung, prostate, pancreas, cervical cancers and very rarely oesophageal cancers. We report this case of paraneoplastic dermatomyositis associated with oesophageal cancer. Usually dermatomyositis is common in females but our patient is male. Also we found that patient improved both symptomatically and in terms of blood reports which showed normalization after a course of corticosteroids.

Hence it is very important for physicians to be aware of dermatomyositis especially the paraneoplastic variant. Patient presenting with signs and symptoms of dermatomyositis has to be investigated for underlying malignancy or its recurrence. Early diagnosis and institution of treatment both in terms of steroids for dermatomyositis and therapy for underlying malignancy improves the outcome drastically. The prognosis of the disease and five year survival rates also increase.

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