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A review on dermatomyositis

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ABSTRACT

Dermatomyositis (DM) is an autoimmune inflammatory idiopathic myopathy with distinctive skin lesions, unique autoantibodies and, most notably, symmetrical proximal muscle weakness and sub-acute onset muscle inflammation. Some forms of DM, referred to as clinically amyopathic DM, have only characteristic skin lesions without muscle disease. Characteristic lesions on the skin include violecious discolouration around the eyelids. The symptoms of this disorder include, but are not limited to, fatigue, diminished mobility and dysphagia. Due to impaired esophageal and respiratory muscles, symptoms of dysphonia and dyspnea have been recorded. Calcinosis is another major complication seen in DM. A calcium accumulation on soft tissue is Calcinosis.

Keywords: Dermatomyositis, Autoimmune inflammatory, Idiopathic myopathy, Fatigue.

INTRODUCTION

Dermatomyositis (DM) is a chronic skin and muscle autoimmune disease. While it is considered an autoimmune disease, concerns about etiopathogenesis remain with distinctive papules over digits, erythema over the elbows and knees, heliotrope rash around the eyes, hands or legs, periungual telangiectasia, and dystrophic cuticles [Figure 1]. Skin involvement in DM usually manifests. Muscle participation typically presents initially as proximal muscle fatigue, with or without myalgia or tenderness and it also cause heliotrope eruption on skin [Figure 2]**.** There has been a definition of an amyopathic variant with minimal to no muscle inflammation. The association of DM with an increased risk of internal malignancy is well known [1].

Figure 1: Heliotrope rash and Gottron papules

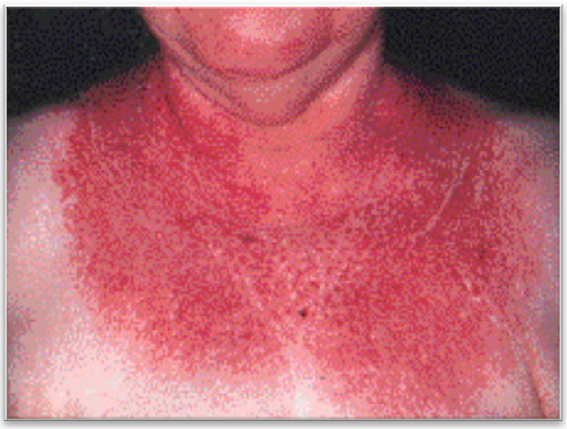

Figure 2: Heliotrope eruption

HISTORY

E.Wagner and P.Potain in the year 1863 (1875) has reported the first detailed descriptions of patients with rare muscle disease (acute myositis form) with cutaneous lesions. A new category of IIMs characterized by the damage of many skeletal muscles and by skin manifestations, were introduced to the medical community by these authors. Such conditions are uncommon, but they are increasingly recognized. They have a number of clinical symptoms, immunological disorders and courses, and they constitute a complex community of unknown causes of illnesses. Dermatomyositis (DM) and polymyositis are the most frequent types of these conditions. [1]

A description of a rare muscle disorder with fatigue and malaise, muscle pain and weakness, swelling of the face, and bluish eyelid lesions was published by Professor Hans Unvericht in 1887. A 27-year-old stonemason reported experiencing an acute onset of fatigue, stiffness, and pain in the proximal muscles of his arm, leg, and back. Diffuse swelling of the face and extremities emerged a week later, followed by low-grade evening fever and a bluish rash across his eyelids. [2]

DEFINITION

Dermatomyositis is one of a group of muscle diseases known as the inflammatory myopathies, which are characterized by chronic muscle inflammation accompanied by muscle weakness.[3]

PATHOPHYSIOLOGY

The primary process is attack on the endothelium of the capillaries of myofibers, with deposition of complement on the vessel walls and eventual formation of membrane attack complex 1.This causes perivascular inflammation and can eventually reduce the number of intramuscular small vessels. 2. This causes hypoxic change in the muscle, characterized by per fascicular atrophy, since these fibers are more distal to the vessel. 3. In

chronic disease, the number of capillaries can be significantly reduced in a biopsy. 4. There is up regulation of MHC-1 in myofiber and also increased expression of ICAM1. 5. No viral etiology has been associated with dermatomyositis. [4]

CAUSES

It is not known the exact cause, but potential causes include: Abnormal genes for which you are born, Cancer, particularly in the elderly, autoimmune disease, a type of disease that causes the immune system of the body to attack its own tissues leads to muscle damage [Figure 3].An illness, medicine or other exposure that causes the disease in your environment [5].

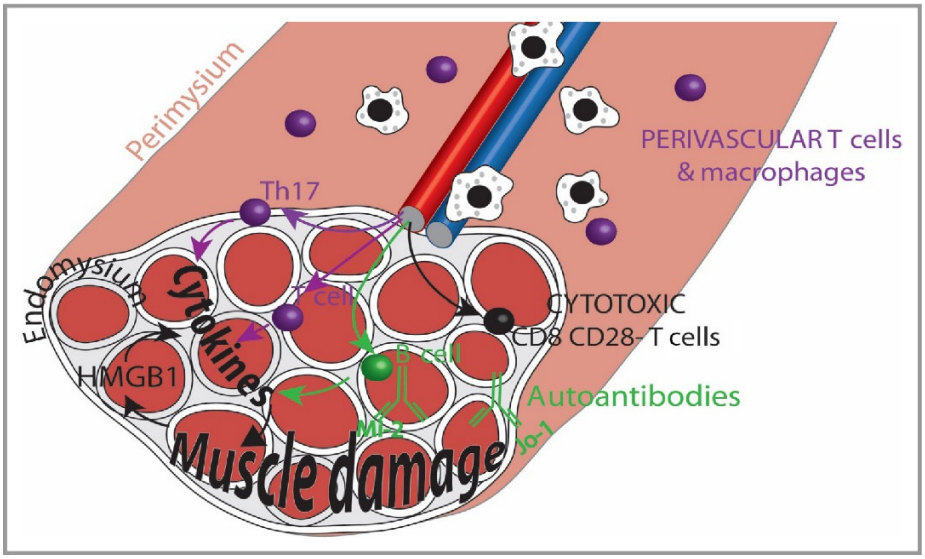

Figure 3: immune system muscle attack

SIGN AND SYMPTOMS

The signs and symptoms of dermatomyositis can appear suddenly or develop gradually over time. The most common signs and symptoms include: A reddish-purple rash around the eyelids [Figure 4],Red or violet bumps that form on the outside joints of the hand (Gottron papules), Red or violet bumps on the knees and elbows, Discolored skin on shoulders, neck, upper back (shawl sign), Muscle weakness starting in the arms and/or legs [Figure 5],Joint pain.

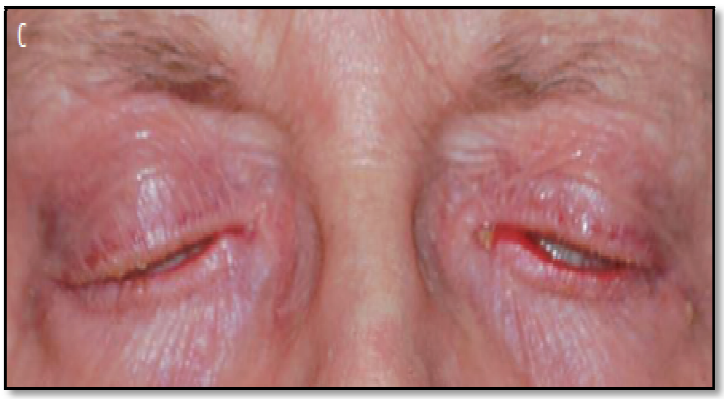

Figure 4: reddish-purple rash around the eyelids

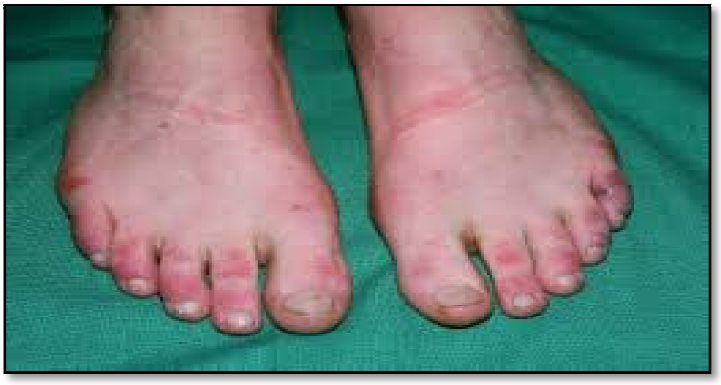

Figure 5: Muscle weakness in legs

HISTOLOGIC DESCRIPTION

The most accurate test for confirming the diagnosis of dermatomyositis and for removing other causes of muscle weakness or skin rash is muscle biopsy. To avoid a missed diagnosis, however, it is important to select the correct muscle for a biopsy. Muscle biopsy, as detected by physical examination or contralateral to irregular muscles, as recognized by electromyography, should be obtained on weak muscles. It is important to obtain a muscle biopsy from patients with suspected dermatomyositis but missing the characteristic skin findings. [6] The microscopic images of the inflammatory myopathies are shown in [Figure 6&7].

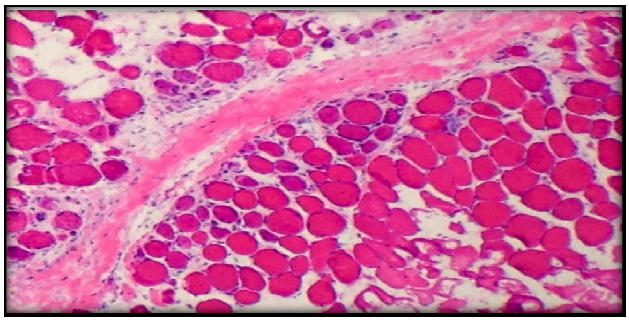

Figure 6: Microscopic image:inflammatory myopathy

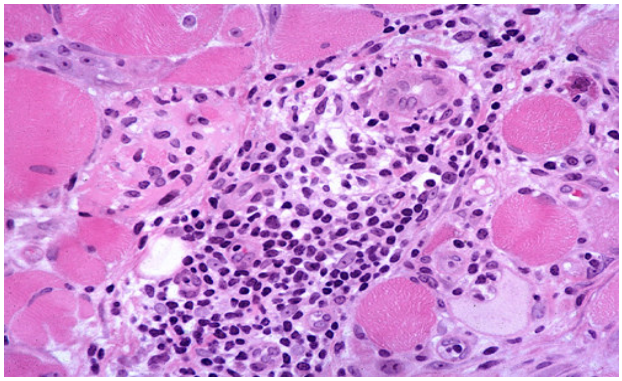

Figure 7: vacuoles or cracks, which contain basophilic granules [7]

DIAGNOSIS

The most commonly used criteria for the diagnosis and classification of DM and PM was defined by Bohan and Peter in 1975.In Dermatomyositis ,the pattern of cellular invasion that it's the blood vessels in the muscle .Muscle cells appear smaller than normal around the edges of bundles of muscle fibre and capillaries**.** A clinical-pathologic diagnosis, Skin and muscle biopsies can be administered at the same time, although the requirement for a skin biopsy can be overridden by a clinical history of skin rash. Magnetic resonance imaging can detect muscle edema or fluid retention in the muscles. [8] [9].

TREATMENT AND DRUGS

Dermatomyositis is almost totally resistant to treatment, particularly in the pediatric age group, although some patients have long-term or lifelong residual symptoms that require sustained immunosuppression. In the early stages, intensive therapy may help to reduce the occurrence of myopathy and Calcinosis. Antimalarial agents, topical steroids, and antihistamines are the first-line treatments in patients with cutaneous DM sun safety. Oral corticosteroids that can be tapered later and other immunosuppressive agents such as methotrexate, mycophenolate mofetil, and azathioprine are used in second-line treatment. Intravenous immunoglobulin may be given to patients who do not respond to these agents (IVIgs). The next line of treatment is Rituximab [10].

CORTICOSTEROIDS

Systemic corticosteroids are the first line of treatment when there are both cutaneous and muscle involvements, only muscle involvement, extensive skin disease.

Antipruritic

Patients of DM may have significant pruritus, and topical agents such as camphor, pramoxine, lidocaine, and menthol as well as oral antihistamines and amitriptyline can be given.[11]

Methotrexate

In DM, methotrexate is considered to be the first-line steroid-sparing agent. It is given in a dose of 7.5 to a maximum of 25 mg/week either orally or subcutaneously. Since it takes 6–8 weeks to show effect, it is not useful for rapid control of acute or progressive DM. Hepatic and pulmonary parameters should be monitored. Cyclosporine in a dose of 3–5 mg/kg/day is useful in refractory cases and has been used in combination with IVIg for relapsed or resistant disease. Common side effects include hypertension, infection, and renal toxicity.

Rituximab

Rituximab, given at a dose of 1 g for 2 weeks apart, was effective in treating recalcitrant or serious illnesses.

Intravenous Immunoglobulin

If there is no response to antimalarial drugs and other immune suppressants, it is possible to give IVIg at a dose of 2 g/kg for 2-5 days per month for 3 months. It is useful in the treatment of refractory myositis, especially in dysphagia patients. A limiting factor in this treatment is the cost [12].

Miscellaneous

The anti-inflammatory effects of dapsone may be useful in resistant cutaneous DM. Antiestrogens such as tamoxifen and anastrozole are useful in the improvement of skin lesions though there is relapse on withdrawal. Pulsed dye laser is useful in reducing the poikilodermatous erythema in DM. Efalizumab, which is a monoclonal antibody against CD11a, is useful in the reduction of erythema over the exposed areas. Leflunomide in a dose of 20 mg/day has been effective in the treatment of recalcitrant skin lesions of DM.

CONCLUSION

In this review we also highlighted the clinical presentation and assessment of DM. In the group of idiopathic inflammatory myositis with very characteristic skin involvement, it is important to note that DM is a rare disease. There are major clinical symptoms, including pulmonary involvement with ILD and malignancy that can be associated with DM. Recent research on CADM have also been checked, showing that these patients

can develop lung involvement and malignancies such as classical DM patients, and need to be monitored for muscle inflammation production in the long term. Finally, we reviewed the diagnostic assessment for DM patients, including an MSA examination.

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