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Case Report
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Long-Term Follow-Up of a Patient with Juvenile Dermatomyositis and Degos-Like Disease

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Abstract

Introduction: Degos-like disease has been associated with several rheumatologic conditions such as systemic lupus erythematosus, antiphospholipid syndrome, systemic sclerosis and rheumatoid arthritis.

Case Report: Seven-year-old girl with low weight, height and body mass index who presented with swollen eyelids with ptosis, suggestive cutaneous features of juvenile dermatomyositis along with the presence of scarce papules with hematic crusts and white atrophic scars with an erythematous rim (Degos-like), dysphagia, flaccid paralysis and proximal and distal weakness. With the use of several immunosuppressants the disease was controlled; and after 8 years of follow-up the patient's condition remains stable.

Conclusion: Herein we describe a unique case of juvenile dermatomyositis associated with Degos-like disease responsive to treatment and a long follow-up.

Keywords: Degos-like disease; Juvenile dermatomyositis; Atrophic; White papules; Telangiectatic rims

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Introduction

Degos' disease, Kölhmeier-Degos' disease, fatal intestinal-cutaneous syndrome or malignant atrophic papulosis is a rare vasculopathy of unknown cause, which primarily involves the skin, intestinal tract, central nervous system and occasionally other organs [1,2].

Kohlmeier in 1941, reported a case described as Buerger disease or thromboangiitis obliterans of mesenteric vessels. Later, Degos described a similar patient with "atrophic papulosquamous dermatitis". These patients died one year after the diagnosis due to intestinal perforations. The cutaneous component was then emphasized with the name of "malignant atrophic pustulosis" [1].

Degos-like disease has been associated with several rheumatologic conditions such as Systemic Lupus Erythematosus (SLE), antiphospholipid syndrome, Systemic Sclerosis (SS) and Rheumatoid Arthritis (RA) [3-5]. Herein we describe a patient with Juvenile Dermatomyositis (JDM) associated with Degos-like disease.

Case Presentation

A 7-year-old girl presented to the emergency department of our Institution with a 6-month history of progressive limb weakness, 2-month history of intermittent low fever, and facial skin lesions over the previous month. Her past medical history was unremarkable.

On physical examination her weight, height and body mass index were ≤ 3rd percentile (13.8 kg, 113 cm and 10.8 kg/m^2, respectively). She had swollen eyelids with ptosis, mild heliotrope rash, erythematous facial photosensitive patches, a fade violet color over her metacarpophalangeal joints (Figure 1a, 1b), and the presence of scarce papules with hematic crusts and white atrophic scars with a peripheral erythematous rim over her armpits, forearms and hands' dorsa (Figure 1b, 1c); in addition to dysphagia, flaccid paralysis, proximal and distal weakness (3/5) of the 4 limbs. Ophthalmologic and cardiologic examinations were within normal limits. The diagnosis of JDM was presumed, and a skin biopsy of the non-classic papules and an electromyography were performed. Further work-up showed hemoglobin 11.6 g/dl, white blood cell counts 9,700, lymphocytes 18%,

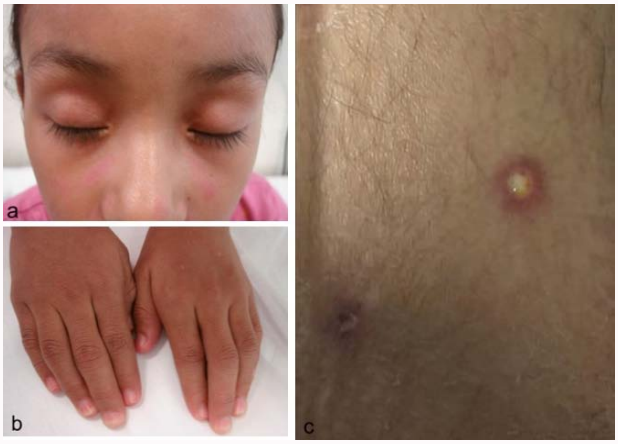

Figure 1: a. Mild heliotrope rash and erythematous facial photosensitive patches. b. Fade violet color over the metacarpophalangeal joints and scarce white atrophic scars on hands' dorsa. c. Papule with porcelain-white center and slightly raised erythematous telangiectatic rim on the forearm.

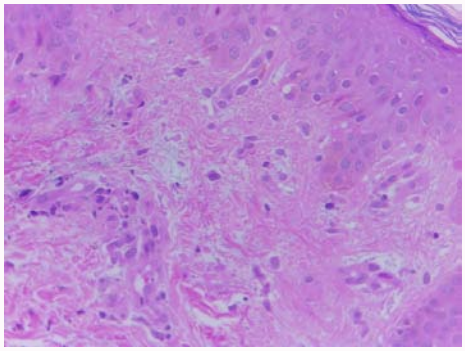

Figure 2: Slight basal layer liquefaction, dermal perivascular lymphocytic infiltrates and lymphocytic vasculitis, dermal edema and mucin deposition (H&E 40x).

neutrophils 75.6%, 449,000 platelets, creatine phosphokinase 194 IU/l, aspartate aminotransferase 28 IU/l, alanine aminotransferase 74 IU/l, lactate dehydrogenase 506 IU/l, C3 119 mg/dl, C4 32 mg/dl, positive antinuclear antibodies 1:180 and, negative double-stranded DNA, RNP, Jo-1, Sm and Ro antibodies.

Electromyography demonstrated myopathic features. Methylprednisolone (MPD) at a dose of 0.5 mg/kg/day and enteral feeding through a transpyloric tube were started. Due to the presence of non-infectious fever for three consecutive days, MPD was increased to 1 mg/kg/day and cyclosporine 3 mg/kg/day was added. The skin biopsy showed an atrophic epidermis with areas of necrosis, mild interface damage, dermal perivascular lymphocytic infiltrates as well as some blood vessel wall necrosis and mucin deposition (Figure 2), suggesting the diagnosis of Degos-like disease and JDM. Therefore, Intravenous Immunoglobulin (IVIG) was administered, acenocoumarin was started to reduce the risk of thrombosis, and a muscle biopsy was performed. The left thigh muscle biopsy reported perivascular and perifascicular atrophy, with mild fibrosis and inflammatory infiltrate, compatible with JDM.

Treatment was adjusted to Prednisone (PDN), methotrexate, acenocoumarin and cyclosporine with marked improvement in muscle strength and skin features, allowing hospital discharge and follow-up as an outpatient. Two months later, her clinical condition was remarkably improved, thus methotrexate, cyclosporine and acenocoumarin were discontinued; and mycophenolate mofetil was started with subsequent tapering of PDN until withdrawal. After 8 years of follow-up, the patient's condition remains stable, with no Degos-like disease manifestations or muscle involvement; however, she developed calcinosis in knees and arms.

Discussion

Dermatomyositis (DM) is characterized by several cutaneous features classified as pathognomonic (Gottron's papules and sign), characteristic (heliotrope erythema, periungual telangiectasias and macular violaceous erythema), compatible (poikiloderma and calcinosis cutis), and others [6]. The patient here described had Gottron's sign and heliotrope erythema as cutaneous findings of JDM, but also presented with papules with hematic crusts and white atrophic scars, suggestive of Degos' disease [1].

Degos' disease can be classified as classic or Degos-like disease [5]. Classic Degos' disease features typical skin lesions and multiple limited infarcts in other organs, such as bowel and central nervous system; approximately 15% of the cases are a benign form often limited to the skin. On the other hand, Degos-like lesions occur in patients with underlying diseases: SLE, antiphospholipid syndrome, SS, RA [3-5], and DM/JDM [4,7-13].

In both types, skin lesions are distinctively characterized by diffuse papules with porcelain-white centers and slightly raised erythematous telangiectatic rims [1,5]. On histopathology, the lesions demonstrate hyperkeratosis, epidermal atrophy, and an underlying wedge-shaped area of dermal ischemia and necrosis with a prominent lymphocytic infiltrate; vessels in the papillary dermis show fibrinoid necrosis and thrombosis. Mucin deposition is seen in all stages, and fibrin deposition may be also observed [8,9]. Several of these histologic features were seen in our patient's skin biopsy.

The etiology of Degos' disease is unknown, however a vaso-occlusive process, obliterating arteriolitis, necrotizing vasculitis, endovasculitis with secondary thrombosis, intravascular coagulation disorder, or a fibrinolysis disorder have all been considered. Several authors suggest Degos' disease actually represents a clinical and histological endpoint reaction pattern to vascular injury derived from different disease processes, instead of a distinct clinical entity, which could explain its association with autoimmune disorders [3-5].

There are only 10 previously reported cases of DM with Degos-like disease [4,7-13], 7 of them in adult patients (3 males and 4 females), with a median age at diagnosis of 37 years (20 to 50 years) and a fatal outcome in 4 (57%), as can be seen in Table 1. In this group of patients, cutaneous lesions typical of Degos were observed in 5 cases.

Regarding pediatric patients, Vadeh et al. [11] reported a 6-year-old-girl who developed bowel rupture 2 weeks after being diagnosed with JDM, and Day et al. [12] described the development of gastrointestinal perforation and cutaneous features of Degos in a 7-year-old-girl with known JMD. Besides, we believe that the child with JDM and livedoid vasculitis described by Olmos et al. [13] could actually have presented JDM with Degos-like disease, since the microcylinders of endoplasmic reticulum in histiocytes described in their patient were also seen in an adult with the classic features of Degos' disease. In our patient, armpit and upper limb lesions, not typical for JDM, were clinically and histopathologically compatible

Table 1: Degos-like disease associated with Dermatomyositis.

Author & Year	Gender & Age	Dermatomyositis features	Degos-like features	Treatment	Outcome
Adult Patients					
Tsao et al. 1997 [7]	Female 26 years	Muscle weakness Paresthesia's Dysphagia Heliotrope eruption Shawl-like poikiloderma Periungual telangiectasias Positive ANAs	6 months after DM diagnosis Numerous small atrophic porcelain-white papules with a thin rim of erythema Esophageal ulcer	Prednisone Methylprednisolone Methotrexate Azathioprine IVIG Aspirin	Good
High et al. 2004 [4]	Female 39 years	Shaw-like violaceous macular erythema Gottron's papules Tendon streaking Periungual telangiectasias Positive ANAs	5 years after DM diagnosis Crops of small erythematous papules that healed with white scars	Azathioprine Aspirin	Good
Magro et al. 2009 [8]	Male 37 years	Muscle weakness and pain Heliotrope rash Gottron's papules Reticular erythema Periungual erythema Positive ANAs	3 months after DM diagnosis Depressed porcelain atrophic macules in a reticulated background Gastric and duodenal ulcers & intestinal perforation	Prednisone Methotrexate IVIG	Fatal
Gupta et al. 2011 [9]	Male 50 years	Raynaud phenomenon Proximal muscle weakness Erythemato-edematous periorbital and neck rash, Positive ANAs, anti-Sm and anti-RNP antibodies	Before signs of DM Erythematous macules, papules, pustules, and small ulcers that healed with a white atrophic scar and a rim of erythema and telangiectasias Diarrhea and melena Diffuse cerebral atrophy Deep vein thrombosis	Dypiridamol Aspirin	Fatal
Burgin et al. 2014 [10]	Male 32 years	Heliotrope rash Gottron's papules Muscle weakness and pain Periungual edema	6 months after DM Erythematous macules and papules with white sclera-atrophic centers, violaceous borders and peripheral hyperpigmentation Gastric ulcers Ischemic colitis	Prednisone Azathioprine Rituximab IVIG Aspirin Dipyridamole Eculizumab	Fatal
Vardeh et al. 2016 [11]	Female 20 years	Polyarthralgias & myalgias Proximal muscle weakness Dysphagia Diffuse subcutaneous edema Positive ANAs	Simultaneous appearance Nausea/vomiting GI tract bleeding GI tract perforation On HP vascular changes resembling systemic Degos disease	Prednisone IV methylprednisolone IVIG Cyclophosphamide	Fatal
	Female 37 years	Gradual proximal weakness Petechial & maculopapular rash in a "shawl" distribution Negative ANAs	Bowel perforation & colectomy On HP vascular changes resembling systemic Degos disease	Corticosteroids Methotrexate IVIG	Good
PEDIATRIC PATIENTS					
Olmos et al. 1979* [13]	Male 8 years	Muscle weakness and pain Dysphagia Heliotrope eruption Scattered telangiectasias Livedo reticular	Simultaneous appearance Atrophic lesions	Not defined	Not defined
Vardeh et al. 2016 [11]	Female 6 years	Proximal muscle weakness Gottron papules Nail bed capillary changes	Two weeks after JDM diagnosis Bowel rupture & abscess formation On HP vascular changes resembling systemic Degos disease	IV methylprednisolone IVIG Cyclophosphamide	Good
Day et al. 2020 [12]	Male 7 years	Proximal muscle weakness Heliotrope rash Gottron's papules	9 months after JDM diagnosis Coalescing porcelain macules on upper eyelids Scattered hypopigmented depressed macules on the arms GI tract perforations Superior vena cava thrombus	Methotrexate Hydroxychloroquine Prednisone Folic acid Ranitidine Monthly IVIG Rituximab Cyclophosphamide	Good
Current case	Female 12 years	Muscle weakness Heliotrope rash Gottron's sign Dysphagia Positive ANAs	Simultaneous appearance Small papules with hematic crusts and white atrophic scars. Ptosis	Methylprednisolone Cyclosporine IVIG Acenocoumarin Prednisone Methotrexate Mycophenolate mofetil	Good

Notes: *Diagnosis was retrospectively presumed based on the description of the case

Abbreviations: DM: Dermatomyositis, JDM: Juvenile Dermatomyositis; ANAs: Antinuclear Antibodies; IVIG: Intravenous Immunoglobulin; IV: Intravenous

with Degos-like disease. Ptosis is not a feature of JDM which could be attributed to Degos' disease, since there are some case reports of Degos' patients with ocular manifestations involving the eyelids [14]. This clinical manifestation resolved completely after treatment.

Treatment modalities for Degos and Degos-like disease have included antiplatelet agents, anticoagulants, immunosuppressant drugs, and eculizumab with varied results [1]. Our patient responded to installed treatment and has remained free of new crops of white-porcelain papules and muscle involvement.

In conclusion, we present a unique case of JDM associated with Degos-like disease responsive to treatment and a long follow-up.

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