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Benign dego's – not so benign: Spilling the beans-unveiling thrombotic micro-Angiopathy in systemic lupus erythematosus

Dr. Sood Vivek[1], Dr. Vandana[2], Dr. Joshi Arun[3], Dr. Dogra Manu[4], Dr. Dua Arun[5], Dr. Chauhan Parikshit[6], Dr. Tiwari Rohit[7], Dr. Gupta KL[8]

[1, 3-6, 8] Department of Nephrology, Postgraduate Institute of Medical Education and Research, Chandigarh, Army Hospital (Research and Referral), New Delhi, India

[2] Department of Internal Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh, Army Hospital (Research and Referral), New Delhi, India

[7] Department of Pathology, Postgraduate Institute of Medical Education and Research, Chandigarh, Army Hospital (Research and Referral), New Delhi, India

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Abstract
Dego's disease, is a rare multisystemic vasculopathy, usually benign with involvement of cutaneous vessels, however at times can turn malignant with widespread thrombosis particularly involving vessels of gastrointestinal tract and central nervous system. In the current manuscript, we describe a rare case of Dego's disease- the benign atrophic papulosis variant unmasking systemic thrombotic microangiopathy in a young patient of systemic lupus erythematosus.

Keywords: Dego's disease, thrombotic microangiopathy systemic lupus erythematosus

Introduction

Dego's disease is characterized by widespread thrombosis particularly of the vessels of skin, central nervous system and gastrointestinal tract. Although etiology is unknown; vasculitis, autoimmune disorders and coagulation disorders are considered as the underlying pathogenic mechanisms but none of these have been confirmed [1]. Dego's disease may manifest either as benign atrophic papulosis involving only the skin or malignant atrophic papulosis which may also involve several internal organs [2]. In this context, we describe a rare case of Dego's disease- the benign atrophic papulosis variant unmasking systemic thrombotic microangiopathy in a young patient of systemic lupus erythematosus (SLE).

Case summary

A twenty three years old un-married female presented with one year history of papules over bilateral arms which became erythematous for last two months. Five years ago, this patient was diagnosed as a case of SLE with cutaneous (malar rash, oral ulcer, photosensitivity, cicatricial alopecia), musculoskeletal (myalgia, arthralgia) and renal manifestations (lupus nephritis class II). She was managed with steroids and Mycophenolate mofetil for three years. In view of maintaining complete remission for over an year, her immunosuppressants were tapered and she was off immunosuppressant drugs for last two years. Currently, except for the skin symptoms, she denied any gastrointestinal, renal or neurological symptoms. There was no history of intake of any alternative medication or NSAIDS.

Dermatological evaluation was contributory with multiple hypopigmented scars surrounded by an erythematous rim, however general physical and systemic examination was unremarkable. Laboratory investigations showed hemoglobin-8.6g/dl, total leucocyte count-6000 cells/mm^3, platelet count 113000 cells/ mm^3, blood urea-40mg/dl, creatinine-0.61mg/dl, 24 hour urine protein – 2000 mg/day and urine microscopy revealed had 6-8 erythrocytes/HPF and protein 2+, without any casts. Further investigations were positive for anti-nuclear antibody, anti-ds-DNA antibody (100 IU/mL), lupus anticoagulant but negative for anticardiolipin antibody (IgG, IgM), anti-beta 2 glycoprotein antibody. Histopathological examination of the skin lesions revealed slight atrophic epidermis with mild hyperkeratosis. The dermis demonstrated edema and extensive mucin deposition leading slight alteration of collagen bundles with melanophages and melanin incontinence. Periodic acid Schiff stain highlighted mucin deposition in the dermis suggesting Dego's disease. Immunofluorescence (IF) of skin biopsy revealed presence of IgM and IgG 3+; C3- 2+ along the dermo-epidermal junction and upper dermis (band test positive) - features were consistent with Lupus. Renal biopsy was suggestive of Lupus nephritis class IV with TMA (6 out of 10 glomeruli (G), showed evidence of endocapillary proliferation with wire-loop lesions, and 2 G showed evidence of thrombotic microangiopathy (TMA) with interstitial inflammation on light microscopy and IF revealed full house pattern. Thus diagnosis of SLE flare (renal and extrarenal) with probable secondary APLA syndrome with Dego's disease was made. Patient was pulsed with methylprednisolone (750 mg/day for 3 days) followed by 1 mg/kg/day oral prednisolone, cyclophosphamide according to NIH protocol and started on warfarin. She responded well and attained remission after second dose of cyclophosphamide and

her skin lesions improved gradually by third month of follow up without any new lesions.

Discussion

Dego's disease was first described by Kohlmeier – as a case of malignant atrophic papulosis in form of thromboangitis obliterans in 1941. It was described as a specific entity by Dego's in 1942. Dego's disease is characterized by diffuse, papular skin eruptions with porcelain white centers and slightly raised erythematous telangiectatic rims [3]. The disease generally occurs in young adults, affects both the sexes and is occasionally familial (autosomal dominant inheritance has been reported). Etiology of the disease is unknown; however, autoimmune diseases, viral infections, coagulation disorders and vasculitis have all been considered as underlying pathogenic mechanisms. Vascular inflammatory changes similar to Lupus Erythematosus have been described but in most cases the inflammation is minimal. It is characterized by narrowing and occlusion of the lumen by intimal proliferation and thrombosis, which leads to ischemia and infarction of involved organs [4]. Skin lesions of Dego's disease consist of three different evolution stages: (a) erythematous papules of onset; (b) erythematous papules with purple or necrotic centers with or without central crust;(c) porcelain-white scars surrounded by an erythematous rim [5].

SLE is a chronic multiorgan auto-immune inflammatory disease and skin lesions are the second most frequent clinical manifestation [6]. Non-specific disease-related skin lesions are frequently seen, usually in the active phase of the disease. Further diagnosis is by laboratory and histologic findings. Vascular manifestations are well known to occur in SLE, and the incidence of TMA in SLE was around 25% however APLA work up was positive in only 10% of these patients.

There are no standardized guidelines for management of these patients and benefit of adding plasmapheresis to standard therapy is controversial [7]. However, index patient had lupus nephritis with TMA and Dego's disease with probable secondary APLA, responded well to standard therapy with no addition of plasmapheresis.

Currently there is no consistently effective therapy for Dego's disease and therapeutic intervention is empirical, since the pathogenesis is not clearly understood. Anticoagulants have been used without demonstrable effect. Other therapies without proven benefits include antibiotics, arsenic, chloroquine, methotrexate, azathioprine and interferon [8].

Conclusion

Dego's disease is a rare entity with clinically distinctive vasculopathy. Pathogenesis remains unclear. Presence of this in SLE is very rare and provides an important clue for the diagnosis of underlying systemic TMA.

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