



Gardner-Diamond syndrome: psychogenic illness with immuno-inflammatory component

Hanane ATARGUINE^[1*], MD, Ouafa HOCAR, MD^[1b], PhD, Nadia AKHDARI^[1c], MD, PhD, Said AMAL^[1d], MD, PhD

^[1*]Department of Dermatology, Arrazi Hospital, CHU Mohamed VI, Marrakech.
Faculty of Medicine and Pharmacy-Cadi Ayyad University Marrakech

ABSTRACT

Gardner-Diamond syndrome also known as auto erythrocyte sensitization syndrome is an extremely rare disorder characterized by the spontaneous appearance of painful bruises that may interest all parts of the body. We report a rare case of this syndrome in association with Sjögren syndrome in a 24-year-old woman, who had an angioedema during the follow-up. Although Gardner-Diamond syndrome is not classified as self-inflammatory, it has been described in association with certain auto immune or inflammatory diseases: systemic lupus erythematosus, immune complex nephritis, hypocomplementaemia, idiopathic thrombocytopenic purpura, lymphadenopathy and the presence of anticardiolipin antibodies. The association of this syndrome with immunological and inflammatory abnormalities, as suggested in our case, requires further study.

Key words: Gardner-Diamond, Gougerot-Sjögren, angioedema.

INTRODUCTION

Gardner-Diamond syndrome also known as Autoerythrocyte sensitization syndrome (AES), painful bruising syndrome, painful blue spots, and psychogenic purpura, is an extremely rare disorder first described about the case of four young women with recurrent spontaneous painful bruises. The exact pathogenesis is not known. Through this observation, we report an unusual association of this syndrome with Sjögren's syndrome and angioedema.

MATERIALS AND METHODS

Observation:

A 24-year-old woman with history of loss of teeth since the age of 9. She was admitted to our department for recurrent painful spontaneous bruises at both lower limbs (Fig.1), appeared for the first time three years ago. She also reported the Raynaud syndrome, a dry eye and mouth syndrome and depressive mood. The psychiatric interview concluded a chronic depression on a histrionic personality.

The rest of physical examination was unremarkable.

The erythrocyte sedimentation rate was 10 mm in the first hour and the number of platelets was 254,000 per cubic millimeter. Other hematological tests were normal, including hemostasis tests. The immunological tests (antinuclear antibodies, SSA, SSB, anti-DNA antibodies, ANCA, anti-phospholipid antibodies) were normal. The Schirmer's test was negative. Punch biopsy of an ecchymotic lesions showed a non specific chronic dermatitis with extravasation of erythrocytes without vasculitis. The biopsy of the salivary glands showed a lymphocytic sialadenitis grade 4. Gardner-Diamond syndrome was confirmed by performing skin tests in three adjacent sites: The following fluids were injected (autologous erythrocytes (0.1 ml), saline solution and autologous plasma) (Fig.2). A bruise appeared in the part exposed to autologous erythrocytes. There was no bruise in the two other sites (Fig.3). We came to a conclusion of Gardner-Diamond syndrome being associated with Sjögren's syndrome. Our patient was treated with prednisone (20mg/ day) in combination with a serotonergic antidepressant (20mg/ day). The outcome was positive with regression of the bruises. During the follow-up, our patient had soft white and recurrent subcutaneous edema concerning the members, worsening during menstruation without any similar family history. The dosage of the complement factors has regained normal levels of C3 and C4, with a functional deficit of inhibitor C1 and an increased rate to its dosage 0.3 g / l. An angioedema type II is compatible with this clinical and paraclinical table.

RESULTS AND DISCUSSION

Gardner-Diamond syndrome is an extremely rare disorder first described in 1955 [1]. This syndrome is characterized by the spontaneous appearance of painful bruises that may interest all parts of the body. Various signs can be associated with this syndrome: headache, digestive disorders, arthralgia, Raynaud syndrome. [2]. This set of clinical signs can simulate a systemic disease, collagen or vasculitis, as in our case.

As to our patient, the diagnosis was made with the rich clinical symptoms associated with the context of chronic depression, the normality of the biological test, the absence of signs of vasculitis, and it is confirmed by the positive result of the intradermal test. Although Gardner-Diamond syndrome is not classified as self-inflammatory, it has been described in association with certain autoimmune or inflammatory diseases: systemic lupus erythematosus, immune complex nephritis, hypocomplementaemia, idiopathic thrombocytopenic purpura, lymphadenopathy, angio-immunoplastique and the presence of anticardiolipin antibodies [3-7].

Ratnoff described the lesions of Gardner-Diamond syndrome as inflammatory bruising [8]. In our patient, the low dose of corticosteroids associated with antidepressant has achieved stabilization of disease. This observation leads us to think that there is an inflammatory component in the Gardner-Diamond syndrome. In three cases of Gardner-Diamond syndrome, erythrocytes showed morphological abnormalities [9]. Among the pathophysiological hypotheses that are currently proposed: depression may be responsible for alterations in the erythrocyte membrane and the vascular endothelium with a secondary increase in vascular permeability [10-12]. In our case, Gardner-Diamond syndrome and angioedema are two separate entities whose common denominator is the psychological stress. Although the hypocomplementaemia has been described in association with Gardner-Diamond syndrome in some comments; when bruises, some patients had decreased serum complement levels. [5]

CONCLUSION

The Gardner-Diamond syndrome is part of psychosomatic disorders that may willingly impressive clinical expression of which the best therapeutic approach remains both omatic and psychological. The association with immunological and inflammatory disorders is suggested in many observations and deserves further study.



Figure 1 .Recurrent painful spontaneous bruises on lower parts.



Figure 2 .Intradermal tests: injection of autologous erythrocytes, saline and autologous plasma in three different sites.



Figure 3 .Appearance ofbruisingin theareaexposed toautologouserythrocytes.

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