

## Case Report

# An intriguing case of Gardner Diamond Syndrome with conversion disorder

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### ABSTRACT

Diamond-Gardner Syndrome or autoerythrocyte sensitization is a rare syndrome characterized by spontaneous development of painful edematous skin lesions progressing to ecchymoses over the next 24 hours. Severe stress and emotional trauma always precede the skin lesion. We present here a case of 15 years old girl who presented with multiple ecchymotic patches over the body with conversion disorder. Baseline biochemical, hematological and immunological investigations were normal. Skin biopsy showed no evidence of vasculitis. All routine coagulation investigations were normal. Therefore the diagnosis of Gardner-Diamond Syndrome was made on purely clinical grounds. It was therefore a diagnosis of exclusion. A high index of suspicion and Psychiatric evaluation were necessary to make the diagnosis.

**Keywords:** Diamond-Gardner Syndrome, Severe stress, Ecchymotic patches

### INTRODUCTION

Gardner-Diamond Syndrome is a rare disease a type of autoimmune vasculopathy of little known aetiology. It consist of outbreaks of painful atraumatic or non-traumatic ecchymotic lesions in any part of body with relapse-remission behaviour, associated with the presence of psychiatric imbalance, suggesting emotional distress as the main trigger and perpetuator of such lesion.

### CASE REPORT

A young female 15 years of age presented with recurrent painful ecchymotic patches all over body since last 4 months. It started with feeling of severe pain and burning sensation in skin on anterior part of right thigh then eventually appearance of red patch over that area after few hours, which was painful, associated with severe muscular pain, attained a size of 7×4cm and became swollen, turned blue with yellowish hue in next 48 hours.

Due to pain patient was unable to move her lower limb, with time it became less tender and gradually disappeared in 1 week to involve other parts of the body. On examination multiple patches at various stages of development were noted of which largest one was on right forearm was bluish in colour, swollen, tender and measured 4×3cm. There was associated carpopedal spasm of right hand. During the stay in hospital patient had multiple alternating somatic complaints like pain in temporomandibular joint, locked jaw, head ache, abdominal pain, and pain in lower limbs with dorsiflexion of feet. There was no history of use of aspirin, NSAIDs, heparin, warfarin and steroids. And no family history of bleeding disorder.

Psychiatric evaluation and tactful history taking revealed that the patient is having anxious personality with borderline intelligence, undergoing exam related stress. Her social interaction had decreased. In addition, she had low level of mood, tension-type headache, easy

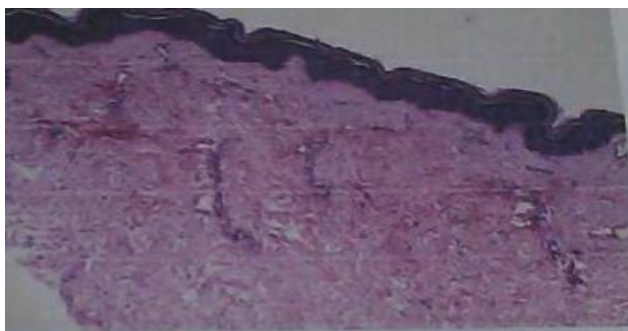
fatigability, poor concentration, hyper-acidity, and sleep disturbance. One episode of ecchymoses started just before her exams suggesting possibility of stress as a precipitating cause of illness. Moreover patient's alternating somatic complaints were relieved on distraction, was regarded as conversion disorder.



**Figure 1: Ecchymotic patches at different stages of development.**

General and systemic examinations were non-contributory. Lab investigations including CBC, platelet studies, ANA, ANCA, RF, Anti-CCP, APLA, Anti-TPO Ab, C3, C4, TSH, Prolactin, IgG, routine coagulation tests (BT,CT,PT/INR) and coagulation factor assay were carried out which helped to rule out ITP, VonWillibrand's disease, DIC, cellulitis, SLE, Polyarteritis nodosa, Henoch-Schonlein purpura.

Histopathological examination of skin biopsy showed extravasation of RBCs around blood vessels without significant inflammatory infiltrate. This helped us to rule out erythema nodosum and septal panniculitis.



**Figure 2: Histopathology of ecchymotic skin patch.**

1. Treatment with steroids showed no improvement.
2. Immunologic, hematologic, dermatologic investigations and consultation ruled out above mentioned conditions.
3. History of stress given by family members and psychiatric evaluation of the patient led us to diagnose this case as a case of 'Gardner-Diamond Syndrome'.

It was therefore 'the diagnosis of exclusion' and high index of suspicion was required to make the diagnosis. There is no specific treatment for Gardner-Diamond Syndrome, symptomatic therapy and psychotherapy is used in several cases with positive results.

## DISCUSSION

Gardner-Diamond Syndrome is an extremely rare condition typically noted in women with psychiatric comorbidities, first described by Diamond and Gardner as autoerythrocyte sensitization syndrome. It is regarded primarily as an autoimmune vasculopathy with sensitization to phosphatidyl-serine, a component of erythrocyte stroma, that is phosphatidyl-serine.<sup>1</sup> Ratnoff and Agle suggested that the condition be renamed as "psychogenic purpura" and postulated a psychogenic basis for the aetiopathogenesis of the disorder.<sup>2</sup> Psychological evaluation of these patients may show hysterical and masochistic traits, depression, anxiety and inability to deal appropriately with hostile feelings<sup>3</sup>. In addition to the cutaneous lesions, a large number of systemic symptoms include abdominal pain, nausea, vomiting, joint pain, headache, and external haemorrhages such as epistaxis, gastrointestinal bleeding, and bleeding from ear canals.<sup>4</sup>

Development generally follows severe stress emotional trauma or with certain psychiatric pathology. Frequency is highest in women but cases in men and adolescents have been reported. Our patient was also a young female of fifteen years, displayed typical ecchymoses preceded by emotional stress, had a number of systemic complaints without any objective evidence of organic disease, she also had depressive symptoms and no aberration in a battery of laboratory tests. Responses to the intra-cutaneous injection of patients own washed erythrocytes are variable.<sup>5</sup> Positive tests consists immediate itching and erythema around the injection site with the subsequent development of a typical lesion over the next 48 hr. A positive reaction was considered confirmatory by Gardner and Diamond but Ratnoff called the response erratic noting positive result only in 59 % of clinical patients. This finding suggests that a negative result may be unable to rule out diagnosis.<sup>2</sup> So this test was not carried out for our patient.

Clinical diagnosis has difficulty owing to the obscure nature of the disease, absence of standard laboratory

markers and the unreliable nature of many patients histories. Dermatologic, Hematologic, Immunologic and psychiatric evaluation are required. Association of psychological features is the mainstay for diagnosis and their presence is one of the essential diagnostic components of the syndrome. There is no specific treatment for Gardner-Diamond Syndrome, symptomatic therapy and psychotherapy is used in several cases with positive results.

## CONCLUSION

Gardner-Diamond Syndrome is a rare condition that should be considered in the differential diagnosis of conditions in which unexplained ecchymoses or purpuric lesions are noticed without any deranged lab investigations. Psychological evaluation is of vital importance to decrease unnecessary investigations and incorporation of appropriate therapy. Psychotherapy and psychiatric treatment provides the most effective treatment to relieve the triggering stress factor.

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