A CASE VIGNETTE OF AUTO ERYTHROCYTE SENSITIZATION (GARDNER–DIAMOND) SYNDROME PRESENTING IN THE OUTPATIENT DEPARTMENT OF A RURAL HOSPITAL

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ABSTRACT

The article deals with two rare cases of Autoerythrocyte sensitization Syndrome that presented in the outpatient department of psychiatry of a rural hospital over a period of time. Autoerythrocyte sensitisation syndrome or Diamond-Gardner syndrome is a rare disorder characterized by painful and spontaneous purpura occurring in women with mental health problems. In patients with mental health problems it should always be considered as a differential diagnosis whenever there is spontaneous, painful bruising, Awareness of this condition may prevent unnecessary investigations and allow early referral to psychiatry or psychology. We present the two cases of 28-year-old and 44 year old female patients who were referred to our psychiatry clinic with a history of recurrent episodes of spontaneous painful bruising. Each of them had been attending departments of different discipline for months and had been given ambiguous diagnoses. Extensive Haematological work up proved uneventful. The diagnosis and treatment of the primary psychiatric illness helped the patients to overcome the condition without further delay.

KEYWORDS

Autoerythrocyte Sensitization Syndrome; Psychogenic Purpura.

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INTRODUCTION

Autoerythrocyte sensitisation syndrome or Diamond-Gardner syndrome is a rare disorder characterized by painful and spontaneous purpura occurring usually in women with mental health problems. The disease is characterized by the appearance of a bruise preceded by tingling and a stinging sensation similar to the usual symptoms. Extensive investigations, including full blood count, coagulation screen, factor VIII complex and connective tissue screen are usually found to be normal. The entity warrants the importance of good rapport establishment, good mental state examination and identification of a primary psychiatric diagnosis. In the case vignette concerned, we will be discussing one early adult and one peri menopausal woman presenting with similar symptoms over a different time frame.

CASE VIGNETTE

A well groomed, healthy looking 28-year-old woman was referred to the dermatology clinic for investigation of a six-year history of intermittent spontaneous bruising associated with joint pain. She had previously been investigated by rheumatologists and clinicians without any significant relief. There were recurrent episodes of bruising over her forearm and anterior aspects of thigh over a short period of time. The lesions continued to persist for 4 to 6 weeks without any other secondary manifestations. There was associated sudden bursting pain lasting for 10 to 15 minutes, followed by swelling. The bruise appeared self-limiting and subsided spontaneously over a period of time.

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There was no prior history of any excessive bleeding of menorrhagia. She was unmarried and denied having sexual encounter yet. There was no definite history of trauma preceding the lesions. She was not taking aspirin or non-steroidal anti-inflammatory drugs regularly and was occasionally given multivitamins by a local physician. Detailed mental status examination revealed she was the only child of her parents who separated when she was 8 years old. Since then she had been staying with her maternal grandparents. She had recurrent bouts of loneliness and had suffered from adjustment related issues with her mother's fiancé. Her personal life consisted of a brief relationship with a guy that lasted for 3 months. It was not intimate and she terminated it 2 years ago on grounds of her academic career.

The post break up period was associated with brief depressive spells which subsided after 6weeks.She was never put on any psychotropic medication and only went to a psychotherapist who suggested to initiate a course of cognitive behaviour therapy which she failed to comply. Of late, she admitted to have felt depressed for the last 8 weeks on her failed venture to put up a computer training set up in her village that had incurred financial losses. She was having reduced sleep and appetite and rarely felt any interest to participate in social gatherings. However there were no associated passive thoughts of death or suicidal ideations.

Her psychometry was normal. Her psychological interviewing did not show any signs of first rank symptoms related to psychosis. Haematological examination revealed normal blood count, coagulation profile (PT, APTT and TCT), factor VIII complex (Factor viii, Von Willebrand antigen and vWF RICOF) and thrombophilia screen, including lupus anticoagulant and anti-cardiolipin antibodies. Her purpuric lesions reappeared on intramuscular insult but resolved spontaneously after a few days. She was given a course of Escitalopram gradually escalating to !0 mg per day and observed on regular follow up visits. The symptoms resolved

without recurrence within 3 months along with improvement in her mental state.

A 44 year old divorced female was referred to the department of psychiatry with history of two deliberate self-harm attempts over the last 2 months. Her informant was her brother. She was receiving antihistaminics and paracetamol as an ancillary medication. She complained of recurrent low mood with feeling of loneliness, helplessness, worthlessness for the past 6 weeks. Her marriage terminated 1 year before on grounds of incompatibility. She was also having difficulty in initiating sleep with a significant reduction in appetite. She denied having any hallucinatory experience or to have consorted to any drug abuse. Her first attempt was by consuming 20 tablets of alprazolam 0.25mg 8 weeks back and the second attempt was by ingesting 14 tablets of cetirizine 10 mg. However along with the mental symptoms she was complaining of recurrent ecchymotic purpuric patches over her left thigh of variable size for the last 6 weeks. She denied to have undergone any trauma.

There was associated joint pain without fever. Her menstrual periods were within normal limits. She denied having suffered from any abnormal bleeding manifestations in the past. Her obstetric history revealed that her first pregnancy was aborted forcefully without her consent by her husband and in laws that initiated her process of maladjustment in the family that ultimately culminated in a divorce. The operative period was uneventful. She had been operated for appendicectomy 25 years back without complication.

Haematological investigations including blood count, coagulation profile (PT, APTT and TCT), factor VIII complex (Factor viii, von Willebrand antigen and vWF RICOF) and thrombophilia screen, including lupus anticoagulant and anti-cardiolipin antibodies remained normal. She had normal glycaemic status and thyroid profile. She was not having any regular medication prior to this consultation. She was put on an integrated plan on gradual escalating dosage of venlafaxine with carefully monitored benzodiazepine over 6 months and her depressive symptoms subsided. The cutaneous manifestations resolved after a period of 3 months without any intervention.

In both the above cases a diagnosis of autoerythrocyte sensitisation syndrome (AES) was suspected and an AES test was performed. However, considering the fact that the disease entity lacks a specified standardized test for establishment we resorted to the method described by Vun and colleagues in 2004,[1] with minor modifications. A peripheral blood sample from the patient was collected in a sterile tube containing citrate solution as an anticoagulant. The sample was centrifuged to isolate the erythrocytes, which were rinsed twice with normal saline. A 75% haematocrit was achieved by mixing the erythrocytes with normal saline. Using a 26-gauge needle, 0.1 ml of the solution was injected intradermally into the patient's right forearm.

The patient's left forearm served as a control into which was injected 0.1 ml of normal saline. The patients were blinded to the constituents of the injections. There were no immediate reaction in either arm on both the case. Over the next few hours both reported a tingling sensation at the red cell injection site and noticed a faint bruise. The bruise was 15 mm and 20 mm respectively in diameter at the 24-hour review. There were no alteration to the size of the bruise or the symptoms for the next 24–48 hours. The bruise started fading thereafter. Meanwhile on the control forearm there were no reaction or bruising at the needle puncture site.

DISCUSSION

Autoerythrocyte sensitisation syndrome or Diamond-Gardner syndrome is a rare disorder that was first described in 1955 in four women manifesting abnormal responses to bruising. It was initially characterized by the development of painful ecchymosis at the site of trauma, followed by progressive erythema and oedema.[2,3,4,5] Gardner and Diamond proposed that this represented a syndrome in which patients were probably sensitive to their own red cell stroma. They further suggested the present nomenclature for the entity. Autoerythrocyte sensitization syndrome is typically seen in adult females, although paediatric and male patients have also been described.[2,6,7] The largest series of 71 AES patients was reported by Ratnoff in 1989 presenting with similar symptomatology.[8]

The authors termed the syndrome 'psychogenic purpura' in view of the association with psychiatric problems in the majority of patients described. Further psychological analysis revealed that the cases had an increased incidence of comorbid depression, aggressive spells, profound guilt feeling, anhydonia, suicidality associated. Hysterical and borderline personality traits were also found to be associated with some of the cases. There has been a few reported case of obsessive compulsive disorder associated.[2,8,9.10,11] In children, Munchausen's syndrome by proxy is an important differential diagnosis to consider. The skin lesions vary in size from 1–2 cm in diameter to the involvement of an entire limb. The lesions typically occur on extremities and rarely on the less accessible locations such as the back.

In particular, lesions on the fingers have been reported.[12] Lesions are preceded by paraesthesia .[10] Other associated complaints in patients with AES include headache, syncope, blurred vision, epistaxis, gastro-intestinal bleeding and pain in the abdomen, chest, muscles and joints. The onset of AES may occasionally be preceded by surgical procedures or by other forms of trauma. In absence of a specific and standardized method autoerythrocyte sensitization test is used to formulate the diagnosis of AES.[2] It has been suggested that a positive skin test may indicate unidentified antigenic components of the erythrocyte membrane.[10,13] A number of haematological and immunological abnormalities like thrombocytosis, morphological abnormalities in RBC and functional platelet defects have been investigated to have been related to this disease entity.

A case of psychogenic purpura with abnormally increased tPA-dependent cutaneous fibrinolytic activity has also been published.[14] Antihistamines, albumin infusions, corticosteroids, chemotherapy, antidepressants, hormones, vitamin C and splenectomy are some of the proposed mode of management of this entity.[1] However the treatment of the underlying psychiatric illness also holds paramount importance in the therapy. The prognosis of AES is favourable with relatively low rate of mortality.[8,10] In some cases, the entity remits within months or years. However, the symptoms may recur on reaggravation of psychiatric illness.[8] The two described case in this vignette are unique because of the fact that they appeared separately as a clinical entity in two different age groups without their prior occurance.

Further, none of the subjects were adolescents as to the common occurrence. The disease characteristically appeared with the advent of psychological stressors and subsided spontaneously over a course of time without any further intervention. Absence of any other comorbid haematological entity further makes the case unique. The necessity of a proper psychiatric history taking and management is also emphasized.

CONCLUSION

In view of the above discussion and case vignette, it appears that the need for a general psychiatric examination in all patients presenting with haematological complaints with associated normal coagulation parameters can never be underestimated. However, although the terminology of psychogenic purpura has been found to be associated with a few issues relating to stigma[15],a clear association remains between this dermatoclinical entity and psychogenic illness. The vignette further explores the possibility of new diagnostic measures and further research on designing diagnostic formulation for the disease entity.

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