PSYCHOGENIC PURPURA: A CASE SERIES AND BRIEF CLINICAL REVIEW

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ABSTRACT

Psychogenic purpura or Diamond-Gardner Syndrome is associated with painful bruising and purpura following an auto-sensitization to red blood cells along with imminent psychological and psychosocial factors. Patients with this disorder may present to the dermatologist from where they may be referred to the psychiatrist. A history of psychological stress may be elicited in some cases and psychiatric comorbidity may be seen in others. The psychiatrist, dermatologist and haematologist may have to assess the patient together to reach a diagnosis after a thorough evaluation. The treatment of this condition often involves a team approach involving various medical specialties. We present a series of three cases of psychogenic purpura and review the relevant literature.

Keywords: Bruising, Diamond-Gardner Syndrome, Psychogenic purpura, Stigmata

INTRODUCTION

Psychogenic purpura is a syndrome that is associated with painful bruising and purpura following an auto-sensitization to red blood cells along with imminent psychological and psychosocial factors[1]. It has also been called painful bruising syndrome, Diamond-Gardner Syndrome and auto-erythrocyte sensitization syndrome[2,3]. The syndrome was first described in four women by Diamond and Gardner in 1955, though in initial reports there was very little mention of psychological factors[4]. The intellectual climate with regard to autoantibodies is very different today from that in 1955 and their role in the disease process is now well established[5].

Since the first paper, over a 110 cases have been reported in literature with essentially similar clinical features[6-11]. Cases involving purpura have been divided by authors into five categories viz. the autoerythrocyte sensitization syndrome, autosensitivity to DNA, hysterical purpura, factitious bleeding and religious stigmata. There is a considerable overlap between these categories but in the present review we shall restrict ourselves to the first syndrome[12]. We present a series of three cases of this syndrome that were referred to us from the dermatology department. We requested for the consent of all the three patients for taking photographs of their lesions. Unfortunately all of them refused at the time when they followed up with us. Currently patients are on follow up but have no lesions.

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CASE 1

An 18 year old right handed girl, Muslim by religion was referred from the dermatology department for a psychiatric evaluation with chief complaints of multiple reddish-green coloured patches all over the body but more prominent on the arms, calves, thighs and trunk. The lesions would start as reddish-green patches around 0.5-3cm and then change colour over a period of 10-12 days to become greenish-brown or dark brown and then would slowly fade and disappear in 7-8 days. On examination the lesions were found on the arms, thighs and trunk in different phases of evolution. The mother mentioned that the first such lesion had appeared when the girl was 3 years of age. She had visited a paediatrician then who after investigations did not advise them any further treatment. The patient would have periods of 1-2 years which would be lesion free and they would reappear from time to time during her childhood and adolescence but no active treatment was sought for the same.

Presently the lesions were present since the past 3-4 months before presentation to our clinic. There was

a history of malaise, myalgia and lethargy without fever for a week prior to the appearance of the lesions. On enquiry she had a lot of anxiety regarding her condition and specifically the lesions which she felt were disfiguring in a way. She was known to be sensitive by nature and anxious in general. She was average in academics and preferred to be aloof and socially isolated. No history suggestive of social phobia or psychosis was elucidated. On investigation in the dermatology department, the autoerythrocyte antibody test was positive with the entire coagulation profile being within normal limits. As the history on probing failed to reveal major psychopathology, she was referred to the clinical psychologist for a detailed psychological and personality evaluation that revealed anxious personality type.

The patient was started on amitryptiline 50mg in divided doses and clonazepam 0.5 mg twice a day along with Cognitive Therapy. After a 2 week follow-up, the patient reported significant improvement in her anxiety symptoms and no new appearance of the purpuric lesions.

CASE 2

A 45 year old Muslim lady divorced from her husband since 5 years, working as a housekeeper since 2 years presented with reddish-brown lesions since the past 1 year appearing on multiple sites all over the body. The first lesion was on the right thigh approximately 2cm X 1cm and reddish brown in colour. The lesion disappeared without treatment within a month. Fresh lesions used to appear at monthly intervals and were associated with minimal tenderness at the site of the lesions. At the time of presentation the patient had two lesions – one on the right arm and one on the right thigh. There was a history suggestive of major depressive disorder since the past two years with symptoms like sadness of mood, easy irritability, lack of interest in day to day activities, occasional feelings of hopelessness and decreased sleep at night and reduced appetite. There was a stressful factor at home as her younger son had fallen into bad company and had conduct problems. He would fight with her often and there would be angry, aggressive and assaultive behaviour from him towards her. She had not taken any treatment prior to consultation with us and was referred to us from the dermatology department. On their evaluation, the blood counts and coagulation profile was normal. The patient was started on Escitalopram 10 mg and Cognitive Therapy. On subsequent follow-up, patient reported improvement in her mood and other depressive features. Her skin lesions had also subsided and there were no new lesions reported.

CASE 3

A 30 year old married female was referred from dermatology department for psychiatric evaluation to rule out psychogenic purpura.

The patient was apparently alright 2-3 months back when she developed mild fever with chills; fever rising every 2 days and subsiding on its own. Simultaneously she started having tender greenish black spots over left arm which remained for 1 week and then disappeared on their own. These spots would appear every 4-5 days and would involve multiple sites on the body like thighs, back and neck. On detailed inquiry she gave history of having similar spots 2 years back which occurred at intervals of 4 months. Frequency of these spots increased in the past one month only.

Further psychiatric history revealed a dispute over property in her family, financial problems and occasional alcohol consumption by the husband which was definitely concerning her when asked about it. On asking patient accepted of feeling sad on occasions due to these stressors, however she also claimed that she was able to cope up with them. She did not reveal any other depressive features.

Past history revealed genitourinary tuberculosis which was successfully treated with medications. Her pre-morbid personality did not reveal any significant pathology and was found to be socially well-adjusted.

This patient was also started on Escitalopram 10 mg and on follow-up she claimed to be better.

DISCUSSION

The lesions in Diamond Gardner syndrome are known to start with a prodrome that consists of painful stabbing, stinging or burning sensations experienced for 1-2 hours before the signs of inflammation like heat, erythema, induration, tenderness and pain appear. There is a characteristic reddish-blue discolouration of purpuric type in 1-2 days and the pain reduces once the bruise has appeared. There is devolution of the lesion with a fading over 1-2 weeks. The disorder has a very high female preponderance with a sex ratio of 1:20 in favour of the female sex. Age of onset may be as early as 5 years and as late as 54 years though the median range is 14-40 years[13]. There may be multiple lesions all over the body in various phases of development and may appear to occur as a result of repeated trauma. Such a picture in a child or adolescent may lead to a suspicion of physical trauma or child abuse – a clinical situation that has to be ruled out before other diagnoses.[14] The size of lesions is usually 1-2cm which are most commonly seen on the extremities followed by anterior trunk. Lesions are rare on the back or face[15]. It is clear that these characteristic cutaneous lesions are only a dramatic and visible component of a much more complex clinical picture. A careful enquiry into the past history will reveal that the patient had a variety of hemorrhagic manifestations that often started years before the painful bruising. Many patients claim that they always bruise easily and also have menorrhagia, gingival bleeding, bleeding after minor surgery and cuts or gastrointestinal bleeding; however none of these symptoms were

reported by any of our patients. It is interesting that this occurs in the presence of a normal coagulation profile[16].

A variety of non-hemorrhagic symptoms have been reported by many patients. These include episodic paresthesiae and tingling numbness, giddiness, diplopia, chest pain, hyperventilation and breathlessness, backaches, urinary frequency and sudden unexplained weight loss. It is seen that many of these symptoms may occur in isolation or just two to three of them may be found in a patient. Also noteworthy is the fact that most of the symptoms mentioned also figure in the diagnostic criteria of panic disorder, major depression and anxiety disorders[17,18]. In most cases laboratory studies and clinical investigations when undertaken turn out to be within normal limits[19].

Many researchers have explored the psychological connotations of this syndrome[20,21]. It is clear that a variety of pathophysiological mechanisms some of which may be mediated by the immune system and some of which are apparently not, result in a remarkably consistent clinical picture across cases. This consistency is reflected not only in past medical history and physical symptoms but also in the psychological background and psychosocial stressors if any. A history of previous psychiatric treatment is not uncommon, psychological disturbance is usually present and history of preceding emotional stress can be frequently elicited[22] and such history was found in both case 2 and case 3 from this review.

Although there may be a lack of experimental evidence in this syndrome, our knowledge in the area of psychoneuroendocrinology and psychoneuroimmunology does permit us to arrive at reasonable hypothetical explanations for the symptoms. The symptoms may be related to a stress induced activation of hypothalamo-pituitary axis hormones that in turn results in mobilization of immune mechanisms while in other cases the disease process may be mediated via beta endorphin, substance P and the kinins[23-25]. Many of these patients have been children in unstable homes where physical and emotional abuse are common. Unconscious guilt, depressive features, borderline personality traits, masochistic tendencies and obsessiveness are all part of the personality configuration[26]. Adult relationships often mirror the ones at home and are dependent, ambivalent, unstable and violent. Many patients have had conversion episodes in the past, dissociative features and unconscious masochistic pleasure tendencies[27].

The treatment of this condition often involves a team approach involving various medical specialties. The psychiatrist, dermatologist and haematologist may have to assess the patient together to reach a diagnosis after a thorough evaluation. There is no specific dermatological treatment available and certain authors recommend that one refrains from giving any treatment at all. Treatment must be directed towards the major area of psychopathology, rather than towards symptomatic relief of cutaneous lesions. It is prudent to avoid repeated laboratory tests and prolonged somatic treatments. These often create a mind set of physical illness and divert focus from various psychological issues at hand. One must seek psychiatric consultation as soon as the diagnosis is established; resist pressure from patients and family for repeated medical evaluation. The specific therapeutic modality used will depend on the nature, level and severity of psychopathology[28]. Patients with borderline personality may do well with a mood stabilizer or anti-depressant along with cognitive, psychodynamic or dialectical behaviour therapy[29]. Environmental manipulation to reduce stress factors may be essential. Along with drug treatment it is mandatory that some form of brief or prolonged psychotherapy be initiated by the treating doctor[30]. It is always better that a combination of various therapies be initiated in such cases than sticking to one approach alone.

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