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Psychogenic Purpura with Hematuria and Sexual Pain Disorder: A Case Report

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Abstract

Psychogenic purpura (Gardner-Diamond syndrome) is the occurrence and spontaneous recurrence of painful ecchymosis following emotional stress and minor trauma. Although the exact mechanism of this syndrome remains unknown, apart from skin lesions, different types of hemorrhaging have been reported, such as epistaxis, gastrointestinal bleeding, and bleeding from the ear canals and eyes. We report a psychogenic purpura case that presented with hematuria in addition to skin lesions. Based on the psychiatric evaluation she was diagnosed with major depressive disorder, generalized anxiety disorder, and obsessive-compulsive disorder. Additionally, sexual pain disorder accompanied these disorders. With the help of antidepressant and supportive psychotherapy, the patient's ecchymosis and bleeding disappeared. During 8 months of follow-up the symptoms did not return. Vaginismus has not been reported in patients with psychogenic purpura. The presence of vaginismus, which is seen more frequently in eastern cultures and is thought to be related to sociocultural determinants, suggests that some cultural factors may be common to both psychogenic purpura and vaginismus. The aim of this case report was to call attention to a syndrome that is rarely seen and diagnosed, and to discuss its relationship to psychosocial factors. This syndrome should be considered in the differential diagnosis of not only ecchymotic lesions, but also various types of bleeding, including hematuria. Despite the fact that its etiology and treatment are not clearly understood, it should be noted that psychological factors play a role in this disease and therefore, psychopharmacological and psychotherapeutic approaches can be effective.

Key Words: Gardner-Diamond syndrome, psychogenic purpura, vaginismus, hematuria

INTRODUCTION

Psychogenic purpura was first described in 1955 by Gardner and Diamond as recurrent painful ecchymosis that occurs after emotional stress and minor trauma, based on the clinical observation of 4 female patients (Gardner and Diamond, 1955; Ratnoff, 1989). At first the lesions were thought to be a reaction against a patient's own erythrocytes and it was referred to as autoerythrocyte sensitization syndrome, as well as Gardner-Diamond syndrome. Later, Ratnoff and Agle (1968) noted the psychological factors underlying the disorder and renamed the syndrome psychogenic purpura; however, the mechanism of this syndrome remains unknown.

Ecchymosis usually appears on the arms and legs (Groch et al., 1996). In addition to skin lesions, neurological symptoms such as headache, temporary pruritus, paresthesia, and syncope, ocular symptoms such as blurred vision and double vision, pain in the stomach, chest, muscles and joints, and hemorrhaging in the nose, digestive system, ear canals, and eyes can occur (Koptagel-Ilal and Tuncer, 1986; Yücel et al., 2000; Vun and Muir, 2004).

The probability of seeing psychogenic purpura in routine psychiatric practice is quite low, as these cases are rare and they are referred for psychiatric evaluation at a later stage due to the difficulty of non-psychiatric doctors have recognizing the psychogenic origins. Nonetheless, given the psychogenic factors in the etiology of psychogenic purpura and the priority of psychiatric treatment,

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it is a clinical syndrome that psychiatrists need to recognize. Herein we present a case of psychogenic purpura that presented with a rare symptom, namely hematuria.

CASE

A is a 29-year-old female patient that was referred to our clinic from the Istanbul Faculty of Medicine, Hematology Clinic with the prediagnosis of psychogenic purpura following a 2-year follow-up for spontaneous, painful, and itchy ecchymosis, and sudden hemorrhaging. No organic factor that could explain the symptoms was detected. The patient reported that the lesions appeared suddenly for the first time on her arms, chest, and abdominal region about 3 years earlier. The lesions recurred every 2-3 months, on average; each time they lasted for 1-2 months and they appeared on other extremities as well. The patient reported that the lesions were related to emotional factors and that they appeared when she was anxious or upset.

Her symptoms first appeared when she moved in with her family after her divorce, in a period when she was distressed because of her father's critical attitudes. The lesions were sometimes as small as a coin, but at other times covered an entire leg. The ecchymosis was not directly or indirectly associated with physical trauma and was not formed by the patient herself. Additionally, A had 2 episodes of epistaxis bleeding lasting 1-2 months with recurrences every 2 days, on average, for the last year. Moreover, the patient had intermittent bleeding in her urinary tract that lasted about 2 months; the last occurrence was 6 months before she was diagnosed with hematuria.

History: A was born via normal delivery following a normal pregnancy. She had no problems with walking, speech, or other physical/motor development. She was a quiet, shy, well-behaved child that avoided demanding for the fear of being scolded. After graduating primary school she stopped going to school because her father didn't want her to. She went back to school 13 years later after convincing her father that she should. She is currently continuing her distance learning, taking high school and computer courses.

A has a step-brother 6 years older than herself and a sister 8 years younger. She reported having a conflictual relationship with her brother and a close one with her sister. Her father is 59 years old and retired, while her mother is a 50-year-old housewife. A describes her father as harsh and critical, lacking emotional warmth, and quite rigid concerning financial matters. She reported that her father describes her as, "someone who lives off the family's money," because she does not work and continues her education. She reported that her mother is more affectionate and caring, but that she has frequent health problems. She said that her parents have a conflictual relationship and that the conflict causes her mother to faint frequently and have physical complaints; A said that she was worried about her mother. Because of her mother's health problems, A lived with her grandmother for three 3 years and then with her uncle and aunt for a while in another city during her early childhood. She reported that since childhood she has been afraid that her parents would divorce. In addition to these fears, she reported that she cannot express herself in the face of her father's and brother's blaming and humiliating attitudes and that she cries secretly.

She reported that sexuality is a taboo within the family and that they do not speak about it. She had her first menstruation when she was 13 years old. She reported that she had been afraid because she had no previous information about menstruation and that she was relieved after her mother explained it to her. A does not masturbate and she feels uncomfortable thinking about it. Four years earlier A had an arranged marriage; her husband did not have a regular job and she divorced 5 months later. She had no emotional or sexual relationships before her marriage. She did not want to be married and asked for more time to get to know her husband, but her father insisted on the marriage and she complied. She said that her husband had unstable behaviors and had humiliated and abused her verbally. She reported that she was depressed during this period and that her menstrual cycle was irregular for about 3 months. She did not have sexual intercourse with her husband for the first 2 months of their marriage because she had involuntary vaginal spasms. When she did have sexual intercourse with her husband it was painful and difficult. Her father continually criticized her for getting divorced; during this time she felt like a failure and her ecchymosis first appeared.

Psychiatric history: A described having contamination obsessions that occupied her mind for 2-3 hours a day and compulsions concerning cleaning and checking the doors, windows, stove, etc. that lasted for a few years in her early twenties. After her divorce there was a period when she was not cheerful and felt mostly unhappy. She reported that she is anxious and has difficulty controlling her anxiety, even though she knows it is excessive. She can be overly anxious in response to taking exams, health problems, parents' conflict, and contact with peo-

Table. Hematological examinations.

Examination	Result	Normal Values
Hemoglobin	11.4 g dl^{-1}	12-16 g dl^{-1}
Hematocrit	35%	37%- 42%
Leukocyte	4.900 mm^{-3}	4.000-10.000 mm^{-3}
Platelet	166.000 mm^{-3}	150.000- 400.000 mm^{-3}
Bleeding time (Ivy method)	4 min	2-9 min
Prothrombin time	13 s	12-15 s
Activated-partial thromboplastin time	33 s	32- 40 s
Thrombin time	16 s	15-18 s
Platelet aggregation tests	Normal	Normal
Plasma fibrinogen level	325 mg dl^{-1}	250- 400 mg dl^{-1}
Factor VIII level	96%	80%-120%
D-dimer	230 µg dl^{-1}	< 550 µg dl^{-1}
Von Willebrand factor activity	102%	50%-160%
Protein C activity	88%	80%-120%
Protein S activity	102%	80%-120%
Direct Coombs test	Negative	Negative
ANA	Negative	Negative
Anti-ds-DNA	Negative	Negative
Plasma iron level	17 µg dl^{-1}	50-150 µg dl^{-1}
Iron binding capacity	355 µg dl^{-1}	210-450 µg dl^{-1}
Plasma ferritin level	8 ng ml^{-1}	12-30 ng ml^{-1}

ple she does not know. She reported that she has been feeling unhappy and having no desire and has not been feeling joy of life. Occasionally, she has insomnia, loss of appetite, fatigue, and difficulty concentrating. She had not undergone any previous psychiatric treatment. She does not smoke, does not have a history of alcohol or any other substance use, and she does not have a history of attempted suicide.

Medical Examinations: The patient's laboratory examination results are summarized in the Table. There was no abnormality in the morphological examination of her platelets. Skin biopsy and intradermal erythrocyte sensitization testing were not conducted upon the patient's request. The specificity of the intradermal erythrocyte sensitization test is contentious. Erythrocytes injected intradermally lead to ecchymosis; therefore, this test might have significance only in exaggerated reactions. Coagulation tests were analyzed twice, as a routine procedure. Hematological examinations indicated that primary homeostasis and coagulation systems were normal. Negative autoantibody test results led us to rule out a diagnosis of autoimmune vasculitis. Self-inflected purpura and Munchausen's syndrome were not ruled out completely, but there was no evidence to support these diagnoses when the patient's history, behaviors, and attitudes were assessed as a whole. ENT examination (performed due to mouth and nose bleeding) did not reveal any pathology, and urology and nephrology examinations showed that her hematuria was not menstrual. Additionally, no pathology was detected in dermatology, gynecology, or general surgery assessments. No focus of bleeding indicative of endometriosis was detected in these examinations. There weren't any reports of bleeding disorders in the patient's family history.

Mental Status Examination: A looked younger than her age and she had a childish expression. At first she was shy, inhibited, and tense. She was cooperative with the interviewer. She was conscious, cooperative, and her orientation to person, place, and time was accurate. Her thought flow was normal. Her mood was depressed, but she did not have suicidal ideation. Based on the psychiatric evaluation conducted using SCID-I, she was diagnosed with past major depressive disorder, dysthymic disorder, and obsessive compulsive disorder, as well as current major depressive disorder and generalized anxiety disorder. Dependent and avoidant personality disorders were diagnosed via SCID-II. The patient did not report childhood sexual trauma, but had a history of physical and emotional abuse (beating, humiliation, neglect, etc.).

Upon presentation to the psychiatry department, and in the 2nd and 8th months of treatment her Hamil-

ton Anxiety Scale (HAMA) score was 31, 11, and 9, respectively, while her Hamilton Depression Rating Scale (HDRS) score was 29, 7, and 6, respectively.

Her MMPI hysteria and hypochondriasis scores were high. On the "Draw a Person" test she drew an overweight woman about 20 years old whom she described as, "single, bored, and weary; in need of support from someone". Results of these tests conducted by the same person are summarized as follows: In general, these are immature, dependent people with low self-esteem who are in need of care and closeness; they experience psychological problems as somatic complaints, avoid confronting their anger, and act passive-aggressively. They do not accept the role of psychological factors in their somatic complaints; they experience physical symptoms under stress. It is difficult to establish relationships with the opposite sex, even though they desire to do so, and they usually fail.

Clinical Follow-up: The patient was followed-up at the Istanbul Faculty of Medicine, Psychiatry Department, Psychoneurosis Unit and was diagnosed with psychogenic purpura (Gardner-Diamond syndrome), as well as the above-mentioned disorders. Mirtazapine 30 mg/d was prescribed. In addition, she started weekly supportive psychotherapy sessions (45-minute sessions) with a fourth year psychiatric resident under supervision. A was given support throughout the psychotherapy process for coping with conflicts within her family, establishing social relationships, controlling anxiety, motivation about school and the course, and expressing emotions. In addition to an improvement of depression, the patient's symptoms related to purpura rapidly disappeared within the first month of treatment. During the third month of treatment she had minor ecchymosed lesions on her ankles that were as big as coins and lasted for a few days. The patient did not have any bleeding during the 8-month follow-up period. Diagnoses of major depression and generalized anxiety disorder disappeared during the second month of treatment. During the eighth month of the treatment her HAMA and HDRS scores were 9 and 6, respectively.

DISCUSSION

To date, only about 200 psychogenic purpura cases have been reported. Many psychiatric disorders, such as mood disorders, anxiety disorders, somatoform disorders, personality disorders, and dissociative disorders, are reported to be comorbid with psychogenic purpura (Ratnoff and Agle, 1968; Hallstrom, 1969; Yücel et al., 2000). In most cases, particularly, depressive disorders were comorbid (Klein et al., 1975; Lindahl, 1977; Ratnoff, 1989). In the assessment of our case using SCID-I, major depressive disorder, dysthymic disorder, obsessive-compulsive disorder, and generalized anxiety disorder were diagnosed. Frequent psychiatric comorbidity in these patients supports the view that the disorder might have a psychogenic etiology.

There was a period when our patient experienced vaginismus. Psychogenic purpura patients commonly experience difficulties in sexual lives (Klein et al., 1975; Lindahl, 1977; Ratnoff, 1989). Comorbid vaginismus, which is seen more frequently in eastern cultures and is thought to be related to sociocultural factors (Yetkin, 1999; Şahin, 2001), suggests that some cultural factors may be common into both psychogenic purpura and vaginismus. There are many reports indicating that sociocultural beliefs might play a role in psychogenic purpura (Koptagel-Ilal and Tuncer, 1986; Rheu et al., 1990; Yücel et al., 2000). The presented case had difficulty expressing her emotions and reported crying secretly, which bears some similarity to the feeling expressed by such common Turkish phrases as, "bleeding inside" and "pouring the blood inside". It was reported that psychogenic purpura patients have difficulty expressing emotions-anger in particular (Lindahl, 1977). Similar difficulty with emotional expression was reported in vaginismus patients (Şahin, 2001).

When we reviewed the available case reports, in addition to skin lesions, there were reports of bleeding in the mouth, ear, nose, uterus, and eyes (Ratnoff and Agle, 1968; Koptagel-Ilal and Tuncer, 1986; Yücel et al., 2000). Our patient had hematuria, as well as skin lesions, nose, and mouth bleeding. There are reports of hematuria in the international literature (Ratnoff, 1989), but to the best of our knowledge there are no previous reports in the Turkish literature. McIntosh et al. (1977) reported a case of immune complex nephritis comorbid with autoerythrocyte syndrome. Our case highlights the need to include psychogenic purpura in the differential diagnosis of patients with hematuria.

Although there is a general consensus concerning the importance of the role of psychological factors in the etiology of psychogenic purpura, its treatment remains debatable. No deaths have been attributed to psychogenic purpura, but it was reported that the disorder might recur frequently and last for years. To date, 6-mercaptopurine, albumin infusion, antibiotics, anticoagulants, antihistaminics, busulphan, chloroquine, corticosteroids, cypro-

heptadine, desensitization via erythrocyte suspension, hormones (progesterone/estrogen), immunosuppressive agents, meperidine, pentoxifyline, plasmapheresis, splenectomy, vitamin C, psychotherapy, and antidepressants have been used to treat psychogenic purpura (Vun and Muir, 2004). Treatment has limited success. Some researchers report that the most effective treatment is psychotherapy (Lindahl, 1977). The significant improvement observed in the presented case in response to mirtazapine and supportive psychotherapy supports the view that psychogenic purpura has a psychogenic origin and therefore, psychotherapeutic methods should be the primary treatment modality.

In conclusion, it is usually delayed to recognize and diagnosis of Gardner-Diamond syndrome, which is uncommon. Patients have to be seen by many doctors and referred to many different medical departments for a long time until diagnosis as psychogenic purpura. As such, we think that this case report will contribute to more widespread recognition of the syndrome and increase the number of patients correctly diagnosed and treated.

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