Autoerythrocyte Sensitization Syndrome

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Autoerythrocyte sensitization syndrome (Gardner-Diamond syndrome), also known as painful bruising syndrome or psychogenic purpura, is a distinctive unusual bruising disorder. It occurs primarily in young to middle-aged women who usually have psychogenic problems. To confirm the diagnosis, an intradermal injection of a patient's own whole blood was used. We present a 41-year-old woman with autoerythrocyte sensitization syndrome, confirmed by a positive test on her blood. She was emotionally unstable due to a past medical history of breast cancer. To our knowledge, this is the first reported case in Korean dermatologic literature. (Ann Dermatol 17(1) 27~29, 2005)

Key Words: Autoerythrocyte sensitization syndrome, Gardner-Diamond syndrome, Painful bruising syndrome, Psychogenic purpura

INTRODUCTION

Gardner and Diamond first described autoerythrocyte sensitization syndrome (ASS) in 1955, when they hypothesized that these patients have an abnormal response to bruising, characterized by recurrent crops of painful ecchymotic lesions[1]. ASS is generally associated with emotionally disturbed women, and most commonly occurs on the arms and legs[2]. Fever, abdominal pain and external bleeding are possible. The pathogenesis of ASS remains unknown. Hematological and immunological findings are usually normal. Early diagnosis and appropriate psychiatric management has been reported to be effective[3]. We present a 41-year-old woman who was emotionally unstable due to a past medical history of breast cancer.

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CASE REPORT

A 41-year-old woman presented with a recurrent painful, hemorrhagic bruise on the left thigh. She reported that she had suffered several similar episodes on the legs over the last 8 months. These had regressed spontaneously within 1 to 2 weeks. The lesion was heralded by a burning or tingling sensation. She denied any antecedent trauma or self-induced injury. Physical examination revealed an ill-defined, extremely painful bruise on the left thigh (Fig. 1).

Laboratory examination revealed normal hematological parameters including complete blood cell count, bleeding time, prothrombin time, partial thromboplastin time, factor VIII, antithrombin III, protein C and S, and fibrinogen. Past medical history included a modified radical mastectomy, chemotherapy and radiotherapy due to treatment for cancer on the left breast 2 years before. In addition, she had been suffering intermittently from recurrent attacks of migraine and insomnia. During the enquiry, she seemed to be depressed and hypochondriacal.

On the basis of the characteristic clinical picture and unstable emotional state, a diagnosis of ASS was considered. To confirm our suspicion, 0.1 ml each of the patient's own whole blood and normal saline

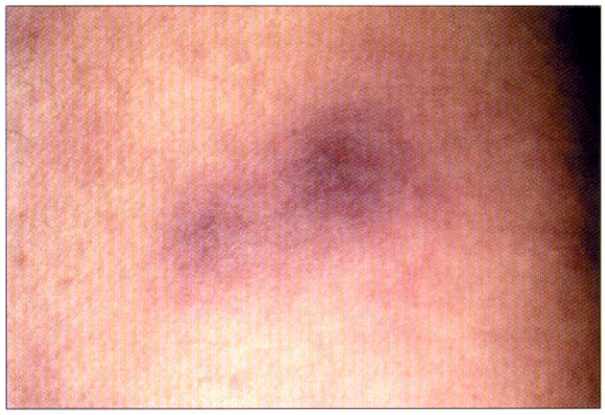

Fig. 1. An extremely painful bruise on the left thigh.

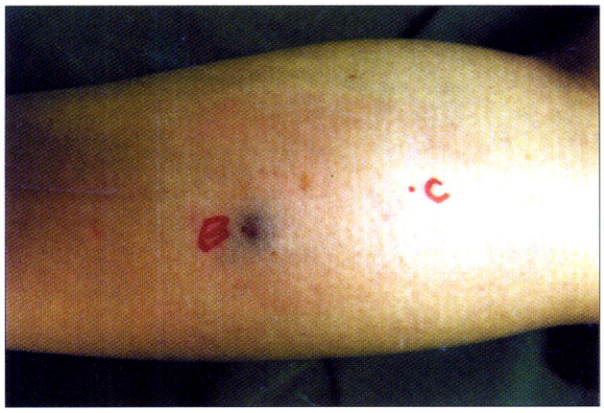

Fig. 2. A painful purpuric papule which developed at the site of an intradermal injection of patient's own whole blood (B), but not at that of a saline control site (C).

was injected intradermally into the left forearm. After 24 hours, a painful purpuric reaction had developed at the site of the blood injection but no change was seen at the normal saline injection site (Fig. 2). These findings led to a diagnosis of ASS. She was referred to the neuropsychiatric department, and given supportive psychotherapy for 2 months. After 4 months of treatment, no recurrent painful bruising was observed.

DISCUSSION

ASS is a rare but well recognized condition. It is seen predominantly in young to middle-aged women. However, pediatric and male patients have also been reported[4,5]. ASS is also called psychogenic purpura in view of its association with a psychiatric problem in the majority of patients. The psychiatric problems include depression, anxiety, personality disorder, hysterical or masochistic character traits, difficulty in handling aggression and hostility, hypochondriasis, guilt feelings and obsessive-compulsive behavior[2,6]. In addition, a surgical procedure or other forms of trauma can precede the onset of ASS. There was even a case report in which copper from an intrauterine device caused a marked exacerbation of the condition[7].

The clinical picture is characteristic. A recurrent type of eruption, which is an extremely painful and tender bruising, and occurs most commonly on the extremities but rarely on the face or trunk. The lesions are often precipitated by emotional stress and usually resolved within 2 weeks. Patients characcteristically describe a burning or tingling sensation before the onset of cutaneous signs. Abdominal pain, diarrhea, nausea, vomiting and headache may occur[2].

Although the exact pathogenesis of ASS remains unknown, emotional upset is generally believed to be a precipitating factor[2]. The histopathology shows only extravasation of erythrocytes in the dermis, and inflammatory cells are not seen[2,6].

Diagnosis of ASS is confirmed by an autoerythrocyte sensitization test. This test consists of an intradermal injection of the patient's own whole blood, RBC, hemoglobin, or fractions of erythrocyte stroma[1,3]. A positive response shows painful purpuric lesions. Hematological and immunological findings are usually normal. Since our patient showed the characteristic clinical picture, we considered a diagnosis of ASS. Without a skin biopsy, we performed an intradermal injection of the patient's own whole blood and made a diagnosis of ASS. She was referred to the neuropsychiatric department without delay. Since then, she has been well.

The differential diagnoses include purpura simplex and purpura factitia. They are often very difficult to differentiate, but atypical locations, peculiar morphology, lack of precipitating factors, and positive autoerythrocyte sensitization test suggest this syndrome.

Various pharmacological treatments have been tried for ASS with limited success. A better response to treatment was observed when the patient also had psychiatric treatment, and this was most effective

when initiated early in the disease[3,8,9].

The prognosis of ASS is generally good. The duration of the disease has been reported to vary from 1 month to 38 years[6]. No deaths have been reported from ASS or complications of the condition[10].

It is important for the dermatologist to be aware of this rare condition to prevent unnecessary investigations and to allow early referral of patients for psychological management.

REFERENCES

1. Gardner FH, Diamond LK: Autoerythrocyte sensitization; a form of purpura producing painful bruising following autosensitization to red blood cells in certain women. Blood 1955;10:675-690. Cited from reference 3.
2. Ratnoff OD: Psychogenic purpura (autoerythrocyte sensitization): an unsolved dilemma. Am J Med 1989;87:16N-21N.
3. Berman DA, Roenigk HH, Green D: Autoerythrocyte sensitization syndrome (psychogenic purpura). J Am Acad Dermatol 1992;27:829-832.
4. Campbell AN, Freedman MH, McClure PD: Autoerythrocyte sensitization. J Pediatr 1983;103: 157-160.
5. Ingber A, Alcalay J, Feuerman EJ: Autoerythrocyte sensitization (Gardner-Diamond syndrome) in men: a case report and review of the literature. Postgrad Med J 1985;61:823-826.
6. Ratnoff OD: The psychogenic purpuras: a review of autoerythrocyte sensitization, autosensitization to DNA, "hysterical" and factitial bleeding, and the religious stigmata. Semin Hematol 1980;17:192-213.
7. Grossman RA: Autoerythrocyte sensitization worsened by a copper-containing IUD. Obstet Gynecol 1987;70:526-528.
8. Anderson JE, DeGoff W, McNamara M: Autoerythrocyte sensitization (psychogenic purpura): a case report and review of the literature. Pediatr Emerg Care 1999;15:47-48.
9. Settle EC Jr: Autoerythrocyte sensitization successfully treated with antidepressants. JAMA 1983; 250:1749-1750.
10. Uthman IW, Moukarbel GV, Salman SM, Salem ZM, Taher AT, Khalil IM: Autoerythrocyte sensitization (Gardner-Diamond) syndrome. Eur J Haematol 2000;65:144-147.