Gardner-Diamond Syndrome

GARDNER-DIAMOND SENDROMU

İlgül ZEREN*, Cengiz BIÇAKÇI*, Güngör YILMAZ", Ahmet KARAMAN

* Uz.Dr.izmir Atatürk Devlet Hastanesi Dermatoloji Kliniği,
" Dr.izmir Atatürk Devlet Hastanesi Dermatoloji Kliniği,
*** Doç.Dr.Izmir Atatürk Devlet Hastanesi Dermatoloji Kliniği, İZMİR

SUMMARY

The autoerythrocyte sensitization (Gardner-Diamond syndrome) is characterized by painful ecchymotic lesions of the skin on various parts of the body. The disease occurs most frequently in young and middle-aged females and emotional upsets are generally believed to be precipitating factors.

18-year-old woman with a 5-year history of recurrent easy bruising is described. Diagnosis was confirmed by intradermal injection of autologous blood. She described more frequent recurrences shortly after a copper-containing intrauterine device had been placed. Taping a copper penny to her forearm caused similar rash. It seems Gardner-Diamond syndrome might be worsened by exposure to copper.

Key Words: Gardner-Diamond syndrome, Autoerythrocyte sensitization

T Klin J Dermatol 1995, 5:95-97

ÖZET

Otoeritrosit sensitizasyon (Gardner-Diamond sendromu), vücudun değişik bölgelerinde ağrılı, ekimotik lezyonlarla karakterizedir. Genellikle genç ve orta yaş kadınlarda oluşan hastalığı emosyonel olayların tetiklediğine inanılmaktadır.

***5** yıldır tekrarlayan ekimozları olan 18 yaşında bir kadın hasta tanımlandı. Tanı, otolog kanın intradermal enjeksiyonu ile kesinleştirildi. Hasta, bakır içeren rahim içi araç yerleştirildikten sonra daha sık atakları olduğunu belirtti. Hastanın önkoluna bantlanan bakır para benzer lezyona neden oldu. Gardner-Diamond sendromunun bakıra maruz kalmakla şiddetlendiği düşünülmektedir.*

Anahtar Kelimeler: Gardner-Diamond sendromu, Otoeritrosit sensitizasyon

T Klin Dermatoloji 1995, 5:95-97

The autoerythrocyte sensitization (Gardner-Diamond syndrome- GDS) described in 1955 by Gardner and Diamond Is characterized by painful ecchymotic lesions of the skin on various parts of the body (1). The disease occurs most frequently in young and middle-aged females and emotional upsets are generally believed to be precipitating factors (2-6). **A** male-to-female ratio of 1:20 is reported, with 80% of patients being between the ages of 14 and 40 years (7).

Geliş Tarihi: 26.7.1995

Yazışma Adresi: Dr.ilgül ZEREN
İnönü Cad. 149 sok. No: 1/16
35360 Hatay, İZMİR

* *Bu yazı 6-9 Haziran 1995 tarihlerinde istanbul'da yapılan Deri ve Zührevi Hastalıklar Derneği ve International Society ot Dermatology 'nin düzenlediği birleşik toplantıda poster olarak sunulmuştur.*

GDS is typified by recurrent bruising, most often on the extremities (2,4,6), and sometimes on the face and trunk (2,6,8). Often bruises occur spontaneously most likely at times of stress, but may also follow minor trauma or surgery (4,7). **A** case whose lesions were worsened by the copper-containing intrauterine device (IUD) was reported in 1987 (4).

A prodrome consisting of a painful or burning sensation in the skin is followed after 1 or 2 hours by the acute onset of all the clinical signs of local inflammation. The severity of pain may warrant narcotics and swelling is followed by bruising. Once the bruise has appeared, pain gradually subsides, and involution of the entire lesion takes place over a period of 1-2 weeks (7,9). Systemic symptoms of fever, muscles pains, headache, abdominal cramps, gastrointestinal bleeding, malaise, nausea and vomiting may accompany the onset (2,7-9).

The diagnosis may be confirmed by intradermal injections of patient's own erythrocytes (1,3,8,10,11).

No therapy seems to be effective, but psychotherapy directed at emotional problems has helped in some cases (2-4,9).

CASE REPORT

18-year-old woman with a 5-year history of recurrent easy bruising is described. Ecchymotic episodes accompanied by sharp pain lasted about 10-15 days and almost always precipitated by emotional stress. She noted that the lesions exacerbated during her pregnancy.

The patient had a history of syncope at the age of 9 and gastrointestinal bleeding five years ago. She described abnormal uterine bleedings after an ISJD had been placed a year ago. During her hospitalization she suffered from headache, dyspnea, tachycardia, and she slept unconscious one night.

On physical examination painful ecchymotic lesions were present on the arms (Figure 1) and the right leg. The remainder of the physical examination revealed no abnormal findings. Extensive laboratory testing was done, with normal results for bleeding, prothrombin time, partial tromboplastin time, and platelet count. Results of CBC, erythrocyte sedimentation rate, urinalysis, plasma glucose, blood urea nitrogen, serum creeatinine were within normal limits. Cranial and thorax computed tomography scan showed negative findings. Abdominal ultrasonography showed multiple ovarian cysts. Psychiatric consultation revealed she had anxiety. Owing to psychological instability, the possibility of autoerythrocyte sensitization was considered.

FIgure 1. Ecchymotic lesions on her right arm and a large bruise developed under the coin on her left forearm.

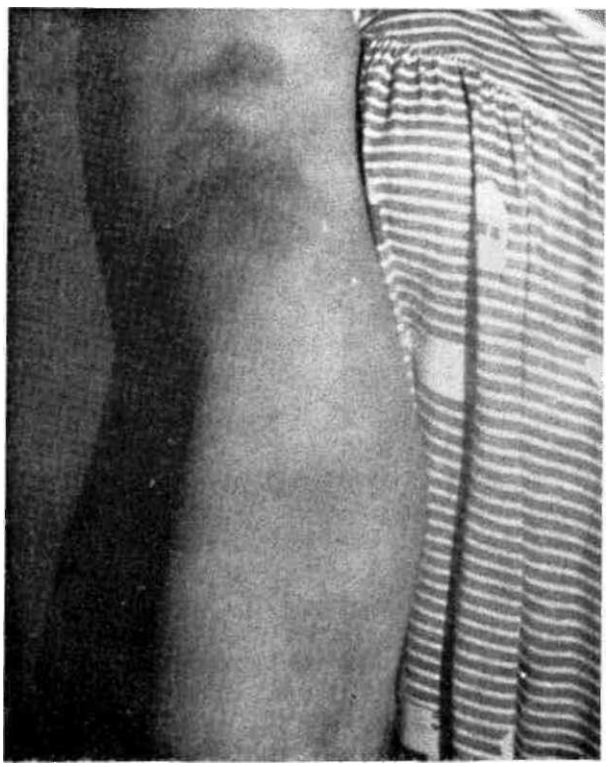

Figure 2. Bruise that developed after intradermal injection of autologous blood on the inner surface of her right forearm.

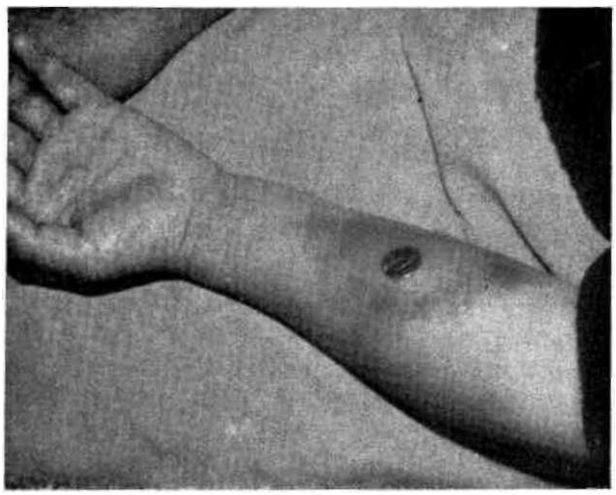

Figure 3. Close-up view of the bruise developed after taping a copper penny.

Intradermal injection of autologous blood caused local tenderness, induration arid later a bruise develo-

ped on the inner surface of her right forearm (Figure 2).

A biopsy taken from the mature lesion showed a proliferation of extravasated red blood cells in the subcutaneous fat.

Investigations showed that her IUD was containing copper. As a test for copper sensitivity, we had the patient tape a copper penny to her forearm. Although she was requested to leave it in place for 24 hours, she had such a severe reaction that she removed it in an hour. She developed a large bruise under the coin similar to those that developed with autoerythrocyte sensitization (Figure 3). The ecchymosis disappeared when the copper-containing IUD was removed. There was no recurrence for about six months.

DISCUSSION

GDS causes painful ecchymoses, and usually occurs in young women (4-6). Until 1985 eight cases of GDS in men were published in the literature (10), The etiology of GDS is unknown (4,9,10). This rare syndrome was initially attributed to sensitization to autologous blood. Although autoerythrocyte and auto DNA sensitization variants, and hysterical purpura have been described, numerous conflicting reports appear in the literature. It seems that both immunological and nonimmunological patho-physiologic pathways are existing (7), It is probable also that in some cases the lesions are factitious (7-9,11).

Psychologically, hysterical mechanisms play a part in many patients (2-9). The patients (exclusively female) suffering from this disorder are described as "angry young women". They are considered hysterical, masochistic, depressive, hostile and timid (5). Psychiatric symptoms were present in 21 of 30 cases reported (3).

Our patients had anxiety and she noted that emotional upsets precipitated her lesions.

The skin manifestations of GDS are often associated with systemic features such as fever (2,7-9), muscle pains (8,9). headache (2,7.8), abdominal cramps (2,3,8,9), gastrointestinal bleeding (3,9). nausea and vomiting (2,9), syncope (2,4,9), artralgia (2,9), abnormal uterine bleeding (3,4,9), dyspnea (9), tachycardia (9).

Our patient described syncope, gastrointestinal and uterine bleeding, headache, dyspnea and tachycardia.

Same with the patient reported by Grossman (4), our patient also had copper sensitivity and her lesions faded when the copper-containing IUD was removed. It seems GDS might be worsened by exposure to copper.

The derrnatologic management of GDS is confined to supportive measures. Warm soaks and mild analgesics may be prescribed. Psychiatric consultation should be requested as soon as the diagnosis is established. For most patients, antidepressant drugs, environmental manipulation to reduce stress, and supportive therapy are indicated (7).

The course is chronic, characterized by remissions and exacerbations of variable length (7).

Being aware of this disorder will avoid unnecessary and costly investigations.

REFERENCES

1. Merten JG. Ecchymotic patches of the fingers and Gardner-Diamond vascular purpura. Phlebologie 1987 Apr-Jun; 40(2):473-87.
2. Arnold HL, Odom RB, James WD. Andrews' disease of the skin, 8th ed. Philadelphia: WB Saunders Co, 1990: 958-9.
3. Champion RH. Purpura. In: Champion RH, Burton JL, Ebling JG eds. Textbook of dermatology, 5th ed, Oxford: Blackwell Scientific Pub!, 1992: 1891.
4. Grossman RA. Autoerythrocyte sensitization worsened by a copper-containing IUD. Obstet Gynecol i987 Sep; 70(3 Pt 2):526-8.
5. Plassmann R. Borderline structure and painful skin hemorrhages: a case of Gardner-Diamond syndrome. 2 Psychosom Med Psychoana! 1985; 31 (2):118-28.
6. Bagot M, Bagot JL, Besserman O, Bertrand JC. The Gardner-Diamond syndrome or painful recurrent ecchymotic purpura. Rev Stomatof Chir Maxiitofac 1984; 85(1):66-9.
7. Koblenzer CS. Psychocutaneous disease. In: Moschella SL, Hurley HJ eds. Dermatology, 3rd ed. Philadelphia: WB Saunders Co, 1992: 2038-38,
8. Braun-Falco O, Piewig G, Wolff HH, Winkelmann RK, Dermatology, 3rd ed. Berlin: Springer-Verlag, 1991: 657.
9. Frantzen E, Voigtlanger V, Gerhardt H. Gardner-Diamond syndrome. Hautarzt 1990 Mar; 41 (3): 168-70.
10. Ingber A, Alcalay J, Feuerman EJ. Autoerythrocyte sensitization (Gardner-Diamond syndrome) in men: a case and review of the literature. Postgrad Med J 1985 Sep; 61(719):823-6.
11. Du Toit JM, Jacobs P, Bird AR, Dickson DN. Autoerythrocyte i,onsltization-the Gardner-Diamond syndrome. A case report. S Afr Med J 1989 Dec; 2.76(11):629-30.