Medical Journal of the
Islamic Republic of Iran

Volume 16
Number 1
Spring 1381
May 2002

# AUTOERYTHROCYTE SENSITIZATION SYNDROME: REPORT OF A CASE AND REVIEW OF THE LITERATURE

L. DASTGHEIB,* P. PEYRAVI,** AND F.S. ASLANI***

*From the Departments of Dermatology and Pathology,*
*Shiraz University of Medical Sciences,*
*Shiraz, I.R. Iran.*

## ABSTRACT

Autoerythrocyte sensitization syndrome (AES) is a rare purpuric disorder of women characterized by inflammatory and painful ecchymotic lesions unrelated to blood clotting or vascular abnormalities. Gastrointestinal bleeding, hematuria, headache or syncopal attacks may also be observed.

Our patient is a 33-year-old woman presenting with recurrent severe painful ecchymotic lesions on both lower extremities associated with headache, palpitation and weakness. The diagnosis of AES was confirmed by skin testing with autologous washed red blood cells. No internal bleeding was detected. She was treated with vitamin C with some success.

*MJIRI, Vol. 16, No. 1, 51-53, 2002.*

**Keywords:** AES, Gardner-Diamond Syndrome, Painful Bruising Syndrome, Psychogenic Purpura.

## INTRODUCTION

In 1955 Gardner and Diamond described a condition characterized by recurrent crops of painful ecchymotic lesions in four emotionally unstable women in whom no hematological disorder was found. They postulated an autoimmune reaction to erythrocytes as further lesions were reproduced by intracutaneous injection of the patient's own erythrocytes or its stroma.[1]

Later, other internal manifestations of the syndrome were described including a wide range of hemorrhagic (hematuria, epistaxis, gastrointestinal bleeding) and non-hemorrhagic complaints including severe headache, paresthesia, repeated syncope and diplopia.[2]

Any case with autoerythrocyte sensitization syndrome (AES) is hardly ever forgettable because of the patient's bizarre and dramatic appearance. Also, since no effective remedy exists and the practitioner is confronted with pressure from colleagues, the patient and her family when only conservative measures are prescribed, the case becomes a distressing disorder to manage.[3]

## CASE REPORT

A 33 year-old married female presented in November 2000 with widespread spontaneous recurrent attacks of painful ecchymotic lesions on her lower extremities and buttocks. The first attack occurred spontaneously 5 months prior to her referral and was not preceded by any trauma. The lesions, developing within hours, were accompanied with chest pain, palpitation, dyspnea and weakness but without fever or arthralgia. There was gradual spontaneous resolution of the lesions in 5-7 days. The patient was admitted to the hospital on two occasions with no definite diagnosis and 30 mg of prednisolone per day was prescribed without benefit. She had undergone two operations in the past (cesarian section and tubal ligation) and was otherwise in good health.

Physical examination revealed raised tender ecchymotic lesions with irregular or annular borders in different sizes, more commonly on the extensor surfaces of the lower extremities and buttocks (Fig. 1). General physical examination was unremarkable. Platelet count,

*Assistant professor in Dermatology
**Senior resident in Dermatology
***Assistant professor in Pathology

bleeding time, fibrinogen level, prothrombin time and partial thromboplastin time were normal and other paraclinical work-up such as ESR, urinalysis, complete blood count, serum analysis for glucose, BUN and liver enzyme levels were also normal. VDRL, ANA, CRP and LE cell preparation were negative. Occult blood in repeated stool examinations was not detected. Chest roentgenogram showed no abnormality. Skin biopsies from the involved sites were performed twice and histopathologic examination revealed mild focal hydropic changes with mild perivascular mononuclear infiltration. No evidence of vasculitis was detected and direct cell immunofluorescence was negative.

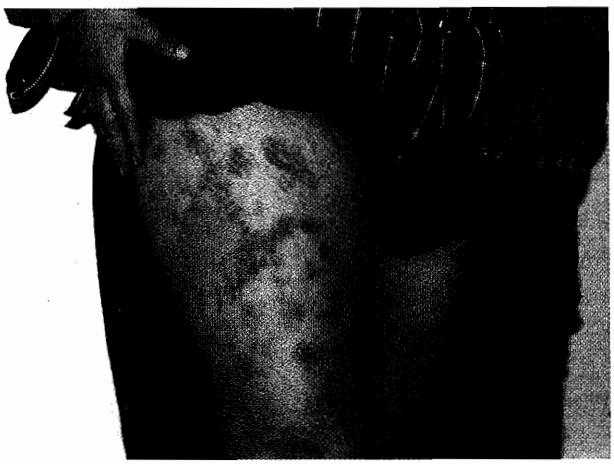

**Fig. 1.** Annular ecchymotic lesion on anterior thigh.

A hematology consultation was performed and no evidence of any bleeding disorder was detected. She appeared emotionally balanced and psychiatric consultation ruled out dermatitis artefacta as she only had mild depression. Due to the occasional cyclic nature of her problem and some correlation with her menses and also to rule out autoimmune progesterone or estrogen dermatitis, skin tests were done with both agents with negative results. Skin test with 0.1 mL 80% of autologous washed red blood cells and with whole blood was performed on the patient and also on two healthy volunteers. A few hours after injection, the patient noted pain and swelling which was then surrounded with purpuric discoloration (Table I, Fig. 2). None of the control subjects reacted to these tests.

Taking all of the above evidence into account, the diagnosis of AES was made and the patient was treated with vitamin C orally and some psychotherapy.

In follow-up, there was mild flare-ups of her symptoms.

## DISCUSSION

**Table I.** Results of skin testing in the patient.

| Material injected intradermally | Reaction |
|---|---|
| 1. Progesterone | - |
| 2. Estrogen | - |
| 3. PPD | - |
| 4. Isotonic saline | - |
| 5. Autologous whole blood | + |
| 6. Autologous washed red cells | + |

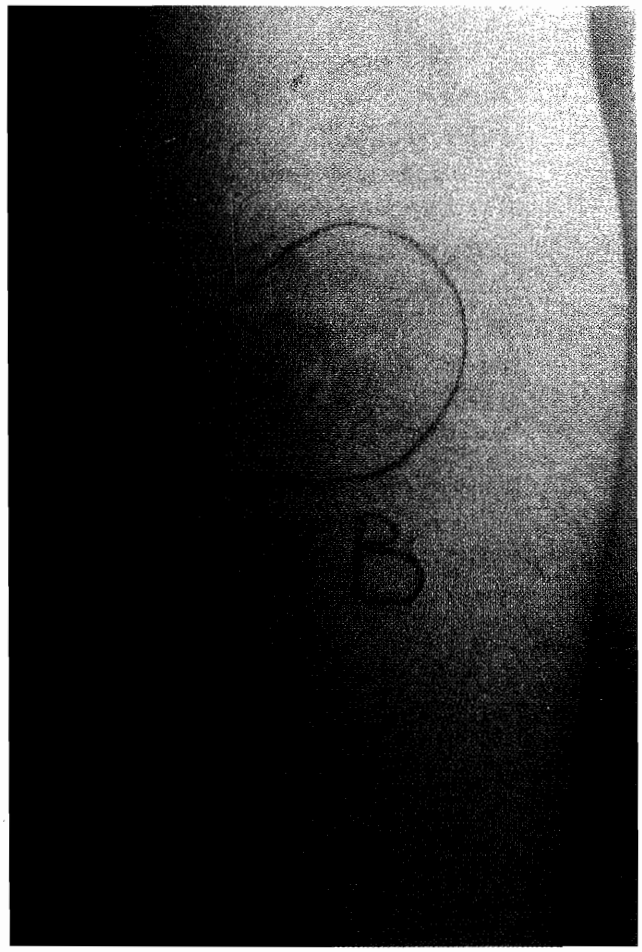

**Fig. 2.** Positive skin test with washed autologous red blood cells showing swelling and purpuric discoloration.

The clinical presentation of AES or psychogenic purpura is remarkably consistent. Almost all patients are women. Few exceptional cases of males with AES have been reported in the literature.[4] Groch and coworkers in evaluation of three patients with this entity found out that phosphatidylserine in red blood cell membranes plays a role in pathogenesis.[5]

The skin biopsy is characterized by extravasation of

red blood cells in the dermis, and in early stages may show edema and non-specific infiltration around blood vessels.

All laboratory findings are normal. An important diagnostic procedure is skin testing with washed red cells obtained from the patient or a competent donor and reproducing the clinical lesion within 24 hours.[6]

There are reports of association of AES with anticardiolipin antibodies[7] or multiple glomus tumors.[8] Psychogenic purpura must be differentiated from skin diseases associated with intravascular clotting defects by normal coagulation studies and the relative inflammatory and painful nature of the skin lesions.[6]

No specific treatment regimen is curative. Large doses of prednisolone or antihistamines are of no benefit except for decreasing the swelling. Attemps to desensitize patients with autologous erythrocytes have not been successful.

Therapy with antimalarials, antibiotics and vitamin C has been without favorable effects.[6]

There is a report of successful treatment with antidepressants.[9] It may be correct to state that the syndrome is not as rare as indicated by the number of cases reported and that there are numerous cases left undiagnosed.

## ACKNOWLEDGEMENT

We would like to thank Dr. Ahmady for her valuable technical assistance in performing the skin tests.

## REFERENCES

1. Gardner FH, Diamond LK: Autoerythrocyte sensitization. Blood 10: 675-90, 1955.
2. Ratnoff OD: Psychogenic purpura. An unsolved dilemma. Am J Med 87: 3-16, 1989.
3. Koblenzer PJ, Koblenzer CS: Psychogenic purpura. A most distressing case. Cutis 45 (1): 60-1, 1990.
4. Ingber A, Acalay J, Feuerman EJ: AES in men. Case report and review of literature. Postgrad Med J 61 (719): 823-6, 1985.
5. Groch GS, et al: Studies in the pathogenesis of AES. Blood 28: 19-33, 1966.
6- Gardner FH: AES (Gardner-Diamond syndrome). In: Demis DJ, (ed.), Demis Clinical Dermatology. Philadelphia: Lippincott, Unit 7: 25, 1998.
7. Sudy E, Urbina F, Vasques P: AES with anticardiolipin antibodies. Br J Dermatol 138 (2): 367-8, 1998.
8. Pearson B, Mazza JJ: Gardner-Diamond syndrome with multiple glomus tumors. Arch Dermatol 111 (7): 894-5, 1975.
9. Settle EC Jr: AES succesfully treated with antidepressants. JAMA 250 (13): 1749-50, 1983.