VOLUME - 11, ISSUE - 09, SEPTEMBER - 2022 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Dermatology

AUTOERYTHROCYTE SENSITIZATION SYNDROME.

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ABSTRACT Autoerythrocyte sensitization syndrome is characterized by lesions which are ill defined, tender, recurrent and spontaneous in nature . There are fewer than 200 case reports[1]. A 40 year old married female, agricultural labourer by occupation came to our OPD with history of recurrent painful petechiae and ecchymosis over both upper and lower extremities since 7 years. The episodes were frequent and were heralded by pain and edema of legs, followed by petechiae progressing to ecchymosis within a day. Lesions occurred at intervals of 7 to 10 days. Not associated with itching ,fever, joint pains or any other symptoms. She was excessively worried since her mother's demise and had low level of mood when visited. It typically occurs in middle-aged females with underlying psychiatric disorders such as depression, anxiety or obsessive compulsive disorders.

KEYWORDS : Autoerythrocyte Sensitization Syndrome, recurrent painful petechiae, depression.

INTRODUCTION:

Psychogenic purpura is also known as Gardner-Diamond syndrome or painful bruising syndrome or autoerythrocyte sensitization syndrome. It is first described by Gardner-Diamond. Though it is a rare disease, it should be considered in the differential diagnosis of recurrent painful purpura. Although lesions resolve spontaneously, remissions and recurrences are quite common. Though varied treatment options have been tried but with limited success.Prompt diagnosis helps to avoid exhaustive investigations and aggressive treatment.

Case Report :

A 40 years old married female, agricultural labourer by occupation came to our OPD with history of recurrent painful petechiae and ecchymosis over both upper and lower extremities since 7 years.

The episodes were frequent and were heralded by pain and edema of legs, followed by petechiae progressing to ecchymosis within a day. Lesions occurred at intervals of 7 to 10 days. Not associated with itching, fever, joint pains or any other symptoms. She was excessively worried since her mother's demise and had low level of mood when visited. Multiple violaceous non edematous non blanchable tender ecchymotic patches over thighs ,medial side of legs, and abdomen.

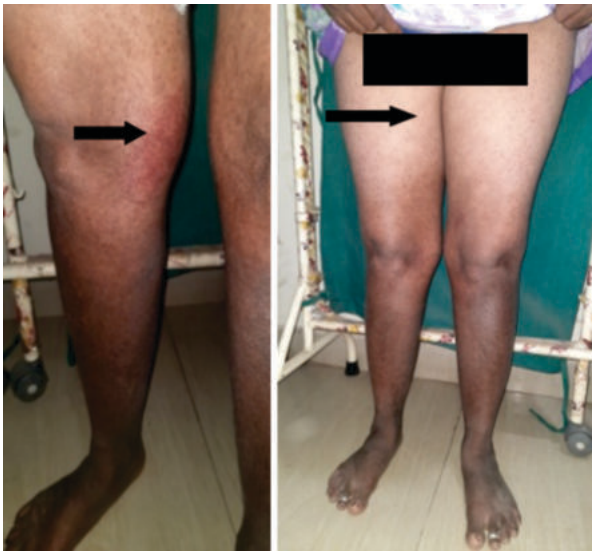


Fig 1&2 : showing multiple petechiae in the medial side of thighs and legs.

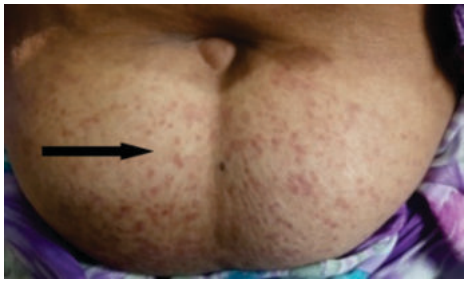


Fig 3:multiple purpuric lesions and petechiae seen over abdomen

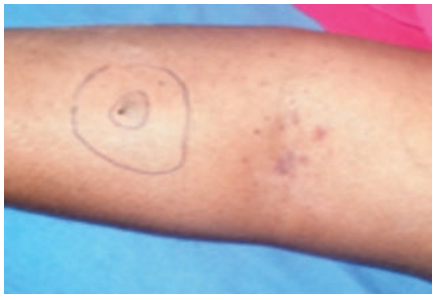
Workup And Management:

Blood parameters and coagulation profile :WNL. Liver and renal function tests: WNL

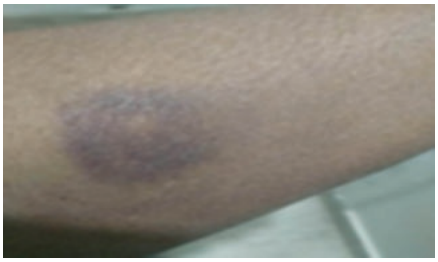
ANA profile: Negative.Dengue serology : Negative.Vitamin c & vitamin k: WNL.

Skin Biopsy : NonContributory .Autoerythrocyte Sensitization Test(AST): Positive (within 48 hours).

AST TEST



It is done by injecting 0.5ml of patient's own blood as a source of RBCs into her forearm.



After 48 hours she experienced the reddish discoloration : ecchymotic patch .

DISCUSSION:

Psychogenic purpura, first described by Gardner- Diamond and named by them as autoerythrocyte sensitization (1955)[2].It is a rare syndrome characterized by spontaneous, painful inflammatory ecchymoses. Because of the evidence that the occurrence of this syndrome is related to psychological factors, Ratnoff and Agle (1968) suggested that the condition be renamed "psychogenic purpura." They postulated a psychogenic basis for the etiopathogenesis of the disorder.[3] Psychological evaluation of these patients may show hysterical and masochistic traits, depression, anxiety, and inability to deal appropriately with hostile feelings.[4,5] However, the precise mechanism of this syndrome is not well-understood. Our patient displayed typical ecchymoses preceded by emotional stress, had a number of systemic complaints without any objective evidence of organic disease, had depressive symptoms, and had no aberration in a battery of laboratory tests. In addition to the cutaneous lesions, a large number of systemic symptoms have also been described in this syndrome, including abdominal pain, nausea, vomiting, joint pain, headache, and external hemorrhages such as epistaxis, gastrointestinal bleeding, and bleeding from ear canals.Cutaneous responses to the intra-cutaneous injection of erythrocytes are variable.Positive tests consisted of immediate itching and erythema around the injection site with the subsequent development over the next 48 hr of a typical lesion. Autosensitization of patients to their own blood, mainly to phosphatidylserine [phosphoglyceride of red blood cell (RBC) membrane] plays an important role in the pathogenesis[6] The disease follows an intermittent and irregular course with variable treatment responsiveness. Treatment consists of psychiatric therapy, which is most effective when instituted early in the course of the disease; so, early diagnosis will not only minimize the cost of the medical evaluation but will also benefit the patient. In our patient, psychiatric evaluation is done and she was diagnosed with depression and kept on escitalopram 10mg and clonezepam 1mg/day along with topical steroids & emollients.

CONCLUSION:

Auto-erythrocyte sensitization syndrome, also known as Gardner–Diamond syndrome. It typically occurs in middle-aged females with underlying psychiatric disorders such as depression, anxiety or obsessive compulsive disorders. Autosensitization of patients to their own blood, mainly to phosphatidylserine [phosphoglyceride of red blood cell (RBC) membrane] plays an important role in the pathogenesis. Though it is a rare disease, it should be considered in the differential diagnosis of recurrent painful purpura. Prompt diagnosis helps to avoid exhaustive investigations and aggressive treatment.

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