Recurrent painful ecchymosis in an adolescent female

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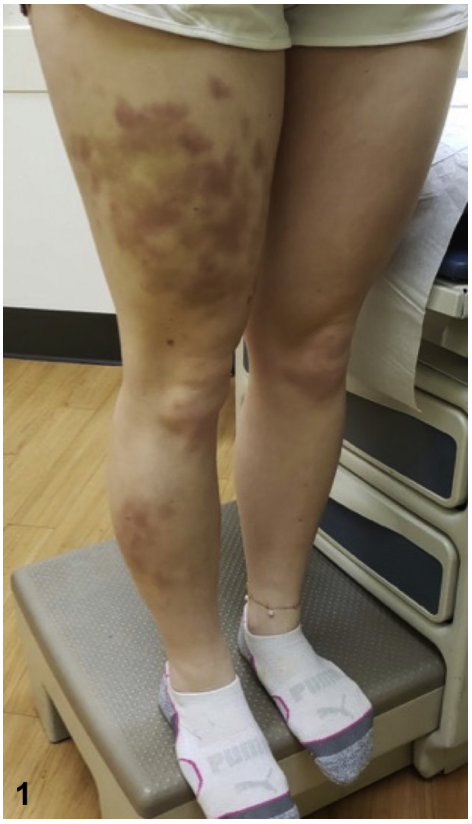

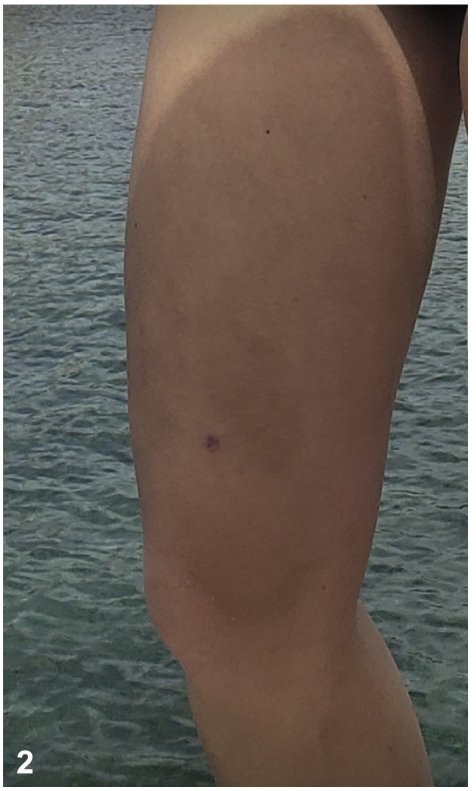

CLINICAL VIGNETTE

A 16-year-old girl presented to clinic with a 2-month history of painful, waxing and waning unilateral erythematous ecchymotic patches on the right lower extremity (Fig 1). She was otherwise healthy and of normal affect, and she denied any history of trauma to the area. She also denied any psychiatric disturbance or history. Two punch biopsies were obtained that both showed a mild perivascular lymphocytic infiltrate. Topical corticosteroids were of limited help, and the lesions spontaneously resolved within 1 week (Fig 2).

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Question 1: What is the most likely diagnosis?

A. Psychogenic purpura (PP)

B. Urticarial vasculitis

C. Thrombotic thrombocytopenia purpura (TTP)

D. Livedo reticularis

E. Livedo vasculopathy

Answers:

A. PP — Correct. PP, also known as *Gardner-Diamond syndrome* or *autoerythrocyte sensitization*, is a rare syndrome of recurrent painful spontaneous ecchymosis that can be associated with a prodromal phase of itch, pain, and warmth at the site of ecchymosis.[1]

B. Urticarial vasculitis — Incorrect. Urticarial vasculitis is characterized by wheals lasting more than 24 hours accompanied with burning and pain. Wheals can resolve with hyperpigmentation or ecchymosis. However, lesions are typically bilateral and symmetrical.[2]

C. TTP — Incorrect. TTP presents as a pentad of renal failure, fever, hemolytic anemia, thrombocytopenia, and neurologic symptoms. Petechial and purpuric lesions are common cutaneous manifestations.[3]

D. Livedo reticularis — Incorrect. Livedo reticularis classically presents on the lower extremities as a violaceous-erythematous and reticulated pattern caused by slow venous flow. It commonly presents as a physiologic response to cold temperatures but can be secondarily caused by a luminal obstruction or systemic disease.[3]

E. Livedo vasculopathy — Incorrect. Livedo vasculopathy is associated with hypercoagulability and presents as tender, punched out ulcers on a background of livedo reticularis on the lower extremities. Involved areas heal with stellate white atrophic scarring, telangiectasias, and hemosiderin deposition known as *atrophie blanche*.[3]

Question 2: Which of the following is correct?

A. PP can present after a triggering event such as trauma, emotional distress, surgery, or infection with ecchymosis developing in an unrelated site.

B. PP is a skin manifestation of an underlying coagulation disorder.

C. Psychiatric disease is not a common comorbidity present in patients with PP.

D. Systemic symptoms are not associated with skin lesions in PP.

E. Lesions of PP resolve with low rates of recurrence.

Answers:

A. PP can present after a triggering event such as trauma, emotional distress, surgery, or infection with ecchymosis developing in an unrelated site — Correct. A triggering event can often be identified in patients with PP. Postsurgical procedure is the most common, possibly because of the traumatic and emotional stress of recovering from surgery.[1]

B. PP is a skin manifestation of an underlying coagulation disorder — Incorrect. The diagnosis of PP is made in a patient with no underlying coagulation disorder that could explain the development of ecchymosis. Hematologic laboratory findings are usually normal.[4]

C. Psychiatric disease is not a common comorbidity present in patients with PP — Incorrect. Approximately 50% of patients with PP may have an underlying psychiatric disorder, most commonly depression and personality disorder.[1]

D. Systemic symptoms are not associated with skin lesions in PP — Incorrect. Appearance of new skin lesions in PP may be accompanied by systemic symptoms such as fever, arthralgia, and myalgia. Additionally, gastrointestinal hemorrhages and epistaxis have been reported.[1,4]

E. Lesions of PP resolve with low rates of recurrence — Incorrect. Lesions of PP resolve within 7 to 10 days. They become less painful and change colors from blue to green and then to yellow before resolving. Unfortunately, lesions do typically recur either in the same location or other parts of the body.[1,4]

Question 3: Which of the following is an appropriate intervention in the management of this condition?

A. Avoidance of skin biopsy

B. Psychiatric evaluation

C. Autoerythrocyte sensitization skin test

D. Referral for malignancy workup

E. Oral corticosteroids

Answers:

A. Avoidance of skin biopsy — Incorrect. Skin biopsy of ecchymosis will show extravascular

erythrocytes and edema in the epidermis or dermis. Histopathologic examination can be useful to help exclude vasculitis.[1,4]

B. Psychiatric evaluation — Correct. PP can present in patients with an underlying psychiatric disorder. A detailed evaluation should be considered, as this may lead to a psychoemotional diagnosis that may otherwise go untreated.[1] Because there is no confirmatory test, the diagnosis of PP is largely one of exclusion. Medication review and laboratory studies would be appropriate to rule out disorders that could predispose patients to ecchymosis.[1,5]

C. Autoerythrocyte sensitization skin test — Incorrect. An autoerythrocyte sensitization skin test is positive if an ecchymosis develops at the site of injected autologous erythrocytes. Historically, this test was used for diagnosis of PP; however, its utility is now controversial because of lack of standardization and data on its validity.[1]

D. Referral for malignancy workup — Incorrect. An association between PP and an underlying malignancy has not been reported. Hematologic malignancy may present with ecchymosis; however, prodromal of symptoms and lack of trauma in the area prior to the development of ecchymosis may not be present.

E. Oral corticosteroids — Incorrect. There is no standardized treatment of PP. Glucocorticoids, hormonal contraceptives, and antibiotics are ineffective. Investigators have reported improvement in skin conditions with treatment of the underlying psychiatric disease, if one is present.[4]

Abbreviations used:

PP: psychogenic purpura
TTP: thrombotic thrombocytopenic purpura

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