Long-standing purpuric exanthema

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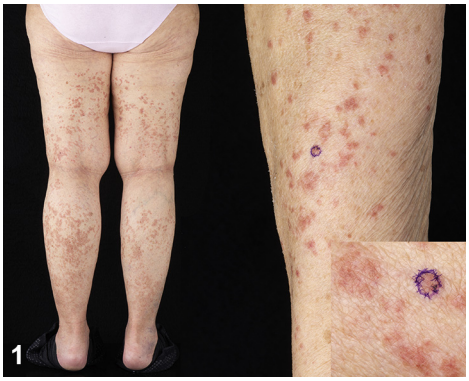
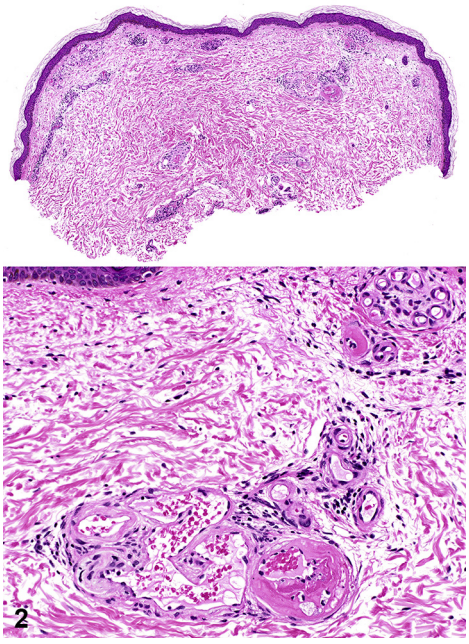
Key words: essential thrombocythemia; occlusive vasculopathy; purpuric exanthema.





A 68-year-old woman presented with a chronic, progressive, mildly pruritic erythematous-purpuric exanthema developing over the past 3 years. The physical examination revealed slightly infiltrated, telangiectatic papules and macules on her trunk, extremities, and on her cheeks (Fig 1). Histopathologic examination revealed an increase of telangiectatic dilated vessels in the dermis without relevant inflammatory infiltrates, as well as small fibrin thrombi within some of the vessels (Fig 2).

The patient had a 24-year history of chronic myeloproliferative disease with essential thrombocythemia treated with anagrelide for the last 18 years, as well as arterial hypertension, chronic kidney failure, and hyperlipidemia.

Question 1: What is the most likely diagnosis?

A. Schamberg disease

B. Occlusive vasculopathy secondary to essential thrombocythemia

C. Drug eruption

D. Immune complex vasculitis

E. Teleangiectasia eruptiva macularis perstans

Answers:

A. Schamberg disease—Incorrect. Schamberg disease is the most common condition among the pigmented purpuric dermatoses. The clinical presentation ranges from petechiae to brown-orange macules, favoring the lower extremities and sometimes involving also the trunk, buttocks, and arms.[1]

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Funding sources: None.

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JAAD Case Reports 2021;11:38-40.
2352-5126

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https://doi.org/10.1016/j.jdcr.2021.03.013

B. Occlusive vasculopathy secondary to essential thrombocythemia—Correct. Essential thrombocythemia is a chronic myeloproliferative condition characterized by an increase in the number of circulating platelets caused by the proliferation of megakaryocytes, resulting in recurrent thromboses of small and medium or large vessels. Cutaneous manifestations occur in almost one fourth of the patients.[2] Essential thrombocythemia can cause a wide range of dermatological manifestations including pruritus, hematomas, ecchymoses, petechiae or purpura, as well as other symptoms related to the occlusive vasculopathy (erythromelalgia, livedo reticularis, livedo racemosa, Raynaud phenomenon, and acrocyanosis).[2,3]

C. Drug eruption—Incorrect. Drug eruptions represent a broad spectrum of cutaneous adverse reactions to several drugs. The clinical presentation is protean, depending on the involved drug and type of cutaneous reaction. Our patient did not receive any new medication in the past several months, and the purpuric exanthema was present for 3 years, and therefore, a diagnosis of drug eruption could be ruled out.

D. Immune complex vasculitis—Incorrect. Immune complex vasculitis is a type of small-vessel vasculitis often presenting with palpable purpura, sometimes with necrosis and ulceration.

E. Teleangiectasia eruptiva macularis perstans—Incorrect. Teleangiectasia eruptiva macularis perstans is an uncommon form of cutaneous mastocytosis. It is characterized by telangiectatic macules and brown background color.[4]

Question 2: Which of the following is a possible cause of the exanthema in our patient?

A. Reactive teleangiectasia subsequent to occlusive vasculopathy

B. Side effect of long-term treatment with anagrelide

C. KIT D816V mutation

D. Viral infection

E. Autoimmune reaction

Answers:

A. Reactive teleangiectasia subsequent to occlusive vasculopathy—Correct. The purpuric exanthema in these patients is likely related to the reactive teleangiectasia subsequent to occlusive vasculopathy of the dermal small vessels.

B. Side effect of long-term treatment with anagrelide—Incorrect. Purpuric exanthema is not described as a long-term adverse reaction of therapy with anagrelide. In some cases, cutaneous ulcerations on the lower legs and dermatomyositis-like eruptions have been described as a side effect of long-term treatment with hydroxyurea in patients with essential thrombocythemia.[5]

C. KIT D816V mutation—Incorrect. The mutation in this gene is found in patients with mastocytosis.

D. Viral infection—Incorrect. Virus infections can cause a wide range of skin lesions, from vasculitic skin changes to classic virus exanthema.

E. Autoimmune reaction—Incorrect. Autoimmune processes play an important role in many dermatological diseases, such as autoimmune bullous diseases and connective tissue diseases.

Question 3: What other conditions can show fibrin thrombi on histopathology?

A. Immune complex vasculitis

B. Livedo racemosa

C. Schamberg disease

D. Drug eruptions

E. Teleangiectasia eruptiva macularis perstans

Answers:

A. Immune complex vasculitis—Incorrect. The histological criteria of immune complex vasculitis are an inflammatory infiltrate with neutrophils and eosinophils, leucocytoclasia, and fibrinoid necrosis of the vessel walls and extravasated erythrocytes.

B. Livedo racemosa—Correct. Livedo racemosa is characterized by a persistent violaceous or erythematous netlike patterning of the skin. It is the typical sign of Sneddon syndrome, and it can also occur by essential thrombocythemia, for example. Fibrin thrombi are one of the histological features.

C. Schamberg disease—Incorrect. Histologic signs, such as hemosiderin deposits and variable dense inflammatory infiltrates may be associated with Schamberg disease.

D. Drug eruptions—incorrect. Histologic features related to drug eruptions include eosinophil granulocytes as wells as neutrophil granulocytes and interface dermatitis.

E. Teleangiectasia eruptiva macularis perstans—Incorrect. Accumulations of mast cells on histology

indicate the presence of cutaneous mastocytosis. The disseminated presence of mast cell collections is found in teleangiectasia eruptiva macularis perstans.

Conflicts of interest

None declared.

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