An unusual cutaneous of B-cell chronic lymphocytic leukemia presenting as a massive left-sided body ecchymosis

Lilia Bekel, MD,a Florie Dhaille, MD,a Guillaume Chaby, MD,a Henri Sevestre, MD, PhD,b Bruno Royer, MD,c Caroline Defossez-Tribout, MD,a Florian Lombart, MD,a Ali Dadban, MD,a and Catherine Lok, MD, PhDa Amiens, France

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INTRODUCTION

Cutaneous infiltration is rare in B-cell chronic lymphocytic leukemia (B-CLL). We describe a case of stage C B-CLL presenting as leukemia cutis (LC) in the form of a massive unilateral ecchymosis.

CASE REPORT

An 84-year-old man was monitored for stage A B-CLL (Binet classification) for 9 years. Past medical history was significant for chronic atrial fibrillation treated with vitamin K antagonists. He was admitted to our university medical center 2 months after the onset of a spontaneous, nonpalpable ecchymosis on the left side of his hemithorax and upper and lower limbs (Fig 1, A-C). Findings on neurologic examination of this right-handed patient were normal. The total lymphocyte count was 393,000/mm³ (1000-4000/mm³), the hemoglobin level was 8.5 g/dL (13-17.5 g/dL), and the platelet count was 259,000/mm³ (150,000-400,000/mm³). Coagulation test results and activated partial thromboplastin time were normal. The prothrombin time was prolonged because of vitamin K antagonist treatment with international normalized ratio within the therapeutic range.

A histopathologic examination of skin biopsies showed the presence of perivascular and perivascular infiltrates of leukemic cells without vasculitis. There was an abnormally high number of ectatic vessels within the papillary and mid-dermis. The leukemic cells were positive for CD20, CD79a, CD5, and CD23 (Fig 1, D-F). Whole-body computed tomography showed splenomegaly and cervical lymphadenopathy. Because of clinical presentation, lymphocytosis, and anemia, stage C B-CLL presenting as LC was diagnosed. After 8 cycles of treatment with bendamustine and rituximab, the cutaneous ecchymosis had completely regressed (Fig 1, G), and blood cell counts had normalized.

DISCUSSION

Leukemia cutis reportedly affects 4% to 20% of patients with B-CLL.1 It is most frequently associated with Binet stage A B-CLL and does not appear to influence disease progression. In fact, skin infiltration by neoplastic B lymphocytes may be caused by cutaneous recruitment of recirculating cells by antigenic stimulation rather than local proliferation of the malignant clone.2 The underlying molecular basis responsible for the migration of the leukemic cells to the skin is unknown.3

LC usually presents as maculae, papules, plaques, nodules, vesicles, and bullae. LC associated with B-CLL may have an ecchymotic, purpuric, or telangiectatic clinical presentation. This is encountered less frequently than in other leukemias, such as acute myeloid leukemia or acute lymphocytic leukemia.3 Plaza et al4 described an atypical presentation of LC in B-CLL in the form of a violaceous, circumferential,

Abbreviations used:
B-CLL: B-cell chronic lymphocytic leukemia
LC: leukemia cutis
 telangiectatic patch of the left lower extremity associated with a limb lymphedema caused by compressive lymphadenopathy. This presentation had similar clinical and histologic findings to those in our case report except for the lymphadenopathy. This phenomenon, by local inflammation, could be considered a stimulus to the local proliferation of the malignant B-cell clone in this site. We did not find such a triggering factor.

LC associated with B-CLL is reported to occur at sites of herpes zoster scars.1,5,6 Herpes zoster infection has unilateral involvement that may be caused by a lower activity of the cell-mediated immune system on the body side concerned. In our patient, serologic status for herpes simplex virus and varicella-zoster virus were positive in favor of remote infection. Except chicken pox in childhood, the patient did not have any history of herpes virus infection or recurrence in this part of the body. The ecchymosis did not have the characteristics of a dermatome and was not accompanied by fever or pain. Zosteriform distribution of LC in B-CLL without previous herpes zoster infection has been described once.6 In our patient, lesions seem to reproduce in a checkerboard mosaicism pattern reported in phacomatosis pigmentovascularis.7 Embryologic antigenic particularities of the involved skin may explain the reported pattern.

Moreover, recent work found relationships between hemispheric lateralization and immune system functioning, with the left hemisphere considered an immune activator and the right hemisphere considered immunosuppressive.8 This may explain some unilateral dermatoses in hemiplegic individuals.

Here, we report a unique case of extensive left-side body ecchymosis as an unusual cutaneous manifestation of B-cell CLL. The pathophysiology of this clinical presentation remains unclear and could be the expression of a skin mosaicism.

Fig 1. A and B. Extensive left-sided ecchymosis on the left thorax (A), and left upper (A) and lower extremities (B). C, Histologic sections show perivascular infiltration of leukemia cells in the dermis. Dilated superficial dermal vessels were noted. D-F, Immunohistochemical studies found that the leukemic cells were positive for CD20 (D), CD23 (E), and CD5 (F). G, Complete resolution of the left-sided ecchymosis 8 months after a combination treatment of bendamustine and rituximab. (A and B, Hematoxylin-eosin stain; original magnifications: ×100; C and D, immunohistochemical stains; original magnifications: ×200.)

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