

Pernio as the presenting sign of blast crisis in acute lymphoblastic leukemia

Abstract

A previously healthy 5-year-old girl presented with acute onset of blue toes and red spots on the nose and fingers. The striking nature of these lesions, along with the finding of submandibular lymphadenopathy, prompted further evaluation. Laboratory findings were remarkable for anemia, high transaminase levels, and high blast count. Histopathologic findings were consistent with early pernio. Further examination revealed acute B-cell lymphoblastic leukemia. Treatment of the leukemia led to resolution of the pernio.

1 | CASE REPORT

A previously healthy 5-year-old girl presented in the spring with a 3-week history of blue toes and a 1-week history of spots on the nose and fingers. The lesions became more erythematous with exertion during outdoor play and were occasionally pruritic. Her mother denied that the patient had had similar lesions during the previous winter. She also complained of right leg pain that had been diagnosed as a "bone bruise." She was otherwise well.

On examination, she had a well-demarcated edematous pink plaque on the nasal tip (Figure 1A) and well-demarcated red to violaceous plaques with subtle pseudovesiculation on the dorsal fingers bilaterally (Figure 1B). All distal toes were red to violaceous, with scaling, and were cool to the touch (Figure 1C). She also had bilateral round, nontender submandibular lymphadenopathy. Because of the intense color and widespread distribution of her skin lesions, along with lymphadenopathy, further examination was performed. Laboratory studies were remarkable for macrocytic anemia (hemoglobin 8.5 g/dL, hematocrit 25.2%), absolute neutropenia (2000/ μ L), thrombocytopenia (90 000/ μ L), transaminitis (alanine transaminase 28 IU/L, aspartate transaminase 51 IU/L), and circulating blasts. She had negative antinuclear antibody and extractable nuclear antigen panels, cold hemagglutinins, and cryoglobulin screens.

Because of the abnormal laboratory findings, two punch biopsies of the digits were performed, both revealing superficial and deep perivascular and periadnexal mixed inflammation with neutrophils, mild papillary dermal edema, and red blood cell extravasation (Figure 2). Lymphocytes were predominantly CD3⁺ T-cells with rare

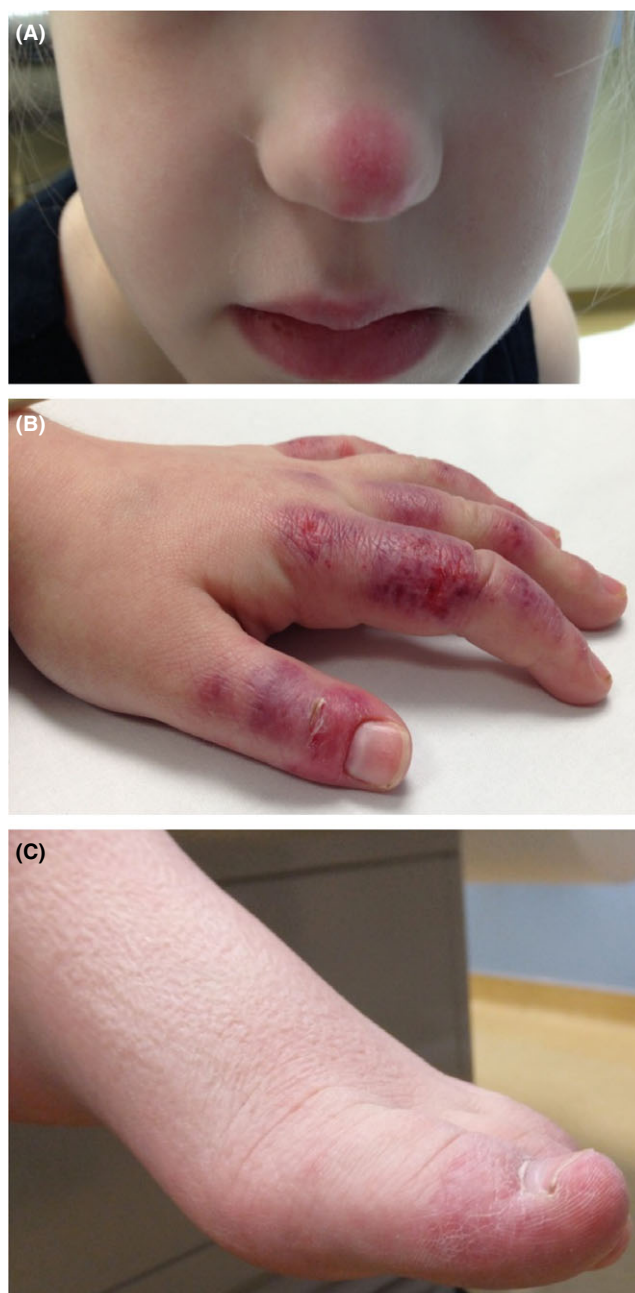


FIGURE 1 (A) Nasal tip with an edematous pink plaque. (B) Left fingers with well-demarcated red to violaceous plaques with subtle pseudovesiculation. (C) Red to violaceous left great toe with some scaling and cool to the touch

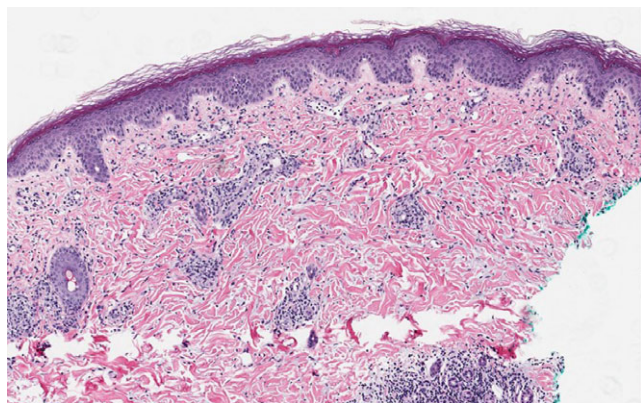


FIGURE 2 Punch biopsy specimen of a finger showing a superficial and deep perivascular and periadnexal mixed inflammation with neutrophils, mild papillary dermal edema, and red blood cell extravasation

scattered CD20⁺ B-cells. No CD34⁺ blasts were seen. Further testing including flow cytometry analysis of the peripheral blood and smears of bone marrow aspirate confirmed B-cell lymphoblastic leukemia with multiple chromosomal abnormalities. Chemotherapy with dexamethasone, vincristine, and intrathecal methotrexate led to significant improvement of her pernio over several weeks and eventual remission of her leukemia.

2 | DISCUSSION

Pernio, or chilblains, is an abnormal reaction to cold, manifested by localized inflammation and sludging of blood flow. It is most commonly observed in young women.¹ Acute lesions may appear 12–24 h after cold insult, whereas the chronic form is associated with repetitive cold exposure and can occasionally lead to scarring.¹ Pernio presents as erythematous violaceous edematous acral lesions that may be painful or pruritic. Cryoproteins (cryoglobulins, cold agglutinins), genetic interferonopathy, excessive cold exposure, and anorexia nervosa may lead to pernio in children. Pernio is typically self-limited, and prevention and rewarming measures are recommended.

The patient's history, clinical presentation, and histopathology were consistent with pernio. Although rare, chilblain-like leukemia cutis has been reported in chronic myeloid leukemia and juvenile myelomonocytic leukemia, and true chilblains has been reported in several cases preceding chronic myelomonocytic leukemia.^{2–5}

Evaluation of a child with pernio should include a thorough history, review of systems, and physical examination, followed by any clinically indicated laboratory tests, because pernio may be an unusual presentation of acute B-cell lymphoblastic leukemia or other

malignancy. Suspicion should be higher in cases with atypical presentation of pernio.

Keywords

acute lymphoblastic leukemia, chilblains, leukemia cutis, pernio

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