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Cutaneous B-Cell Chronic Lymphocytic Leukaemia resembling a Granulomatous Rosacea

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ABSTRACT

B-cell chronic lymphocytic leukemia (B-CLL) is a low-grade lymphoproliferative disease. Cutaneous involvement of B-CLL is limited and, in most cases, it represents non-specific manifestations related to an impaired immune system. Leukemic skin infiltrates (leukemia cutis) occur in 4–20% of patients. Herein we report the case of a 65-year-old woman with B-CLL presenting with papular, nodular, and plaque skin infiltrates affecting the nose, mimicking granulomatous rosacea. We discuss several aspects of rare cutaneous manifestations of B-CLL involving the face.

Keywords: b cell, granulomatous rosacea, leukemia cutis.

INTRODUCTION

B-cell chronic lymphocytic leukemia (B-CLL) is a low-grade lymphoproliferative disease. It represents the most common adult leukemia in western countries. The etiology of the clonal proliferation of B lymphocytes is unknown. Cutaneous involvement of B-CLL is limited and, in most cases, it represents non-specific manifestations related to an impaired immune system. There is a wide spectrum of clinical presentations of B-CLL ranging from chronic and relapsing, isolated to grouped, erythematous to violaceous papules, plaques, or tumors. Herein we report the case of a 65-year-old woman with B-CLL presenting with papular, nodular, and plaque skin infiltrates affecting the nose mimicking granulomatous rosacea. We discuss several aspects of rare cutaneous manifestations of B-CLL involving the face.

CASE REPORT

A 65-year-old woman was first seen in our outpatient clinic because of swelling and redness of the nose. Her past medical history revealed that she had been diagnosed the B-CLL 5 years prior to presentation and had been closely followed up by the hematologist. Her course of illness had been relatively stable and no treatment had been necessary. During the last 2 years, she noted an enlargement of her nose with the appearance of red papules and nodules that tended to spread to the cheeks (Figure 1). Clinical examination showed no other skin lesions and no evidence of lymphadenopathy. Review of systems was unremarkable. A complete blood cell count showed mild lymphocytosis (WBC count, 9,800/mL, lymphocyte count, 3860/mL). Both serology and blood PCR for Borrelial infection were negative.

A punch biopsy specimen from the nose showed dense sheets of nodular lymphoid infiltrate in the dermis (Figure 3,4). The cells were composed of small lymphoid type cells. Immunohistochemical studies showed predominantly CD20-positive and CD79-positive lymphocytes. A proportion of the atypical lymphoid infiltrate co-expressed CD5 and CD23 with k-light chain restriction. The lymphocytes were CD10-negative. PCR studies for immunoglobulin heavy chain gene rearrangement showed a monoclonal population of B lymphocytes. The histopathologic findings were consistent with B-CLL. The involved area was irradiated with 36 Gy in 18 fractions. Starting from six months after the complete lesion regression up to now the patient has remained asymptomatic without development of systemic disease (Figure 2).



Figure 1. Granulomatous rosacea-like eruption on the nose



Figure 2. The clinical aspect after radiotherapy

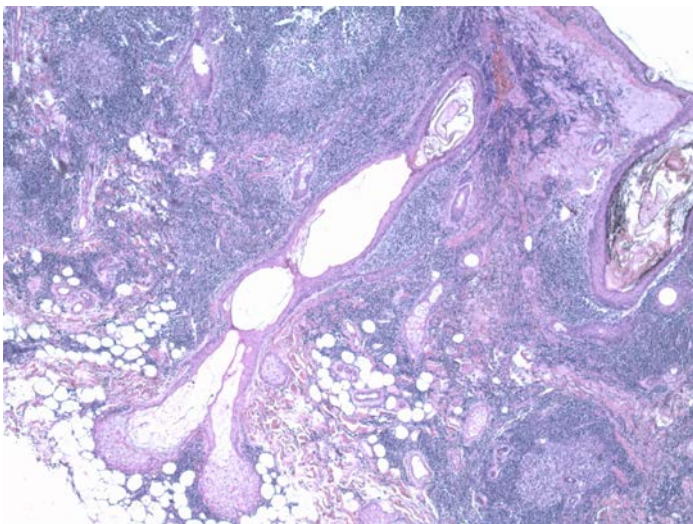


Figure 3. (H&E) The presence of dense, monomorphous, diffuse, and nodular infiltrates of lymphocytes, involving perivascular adnexal and subcutaneous structures.

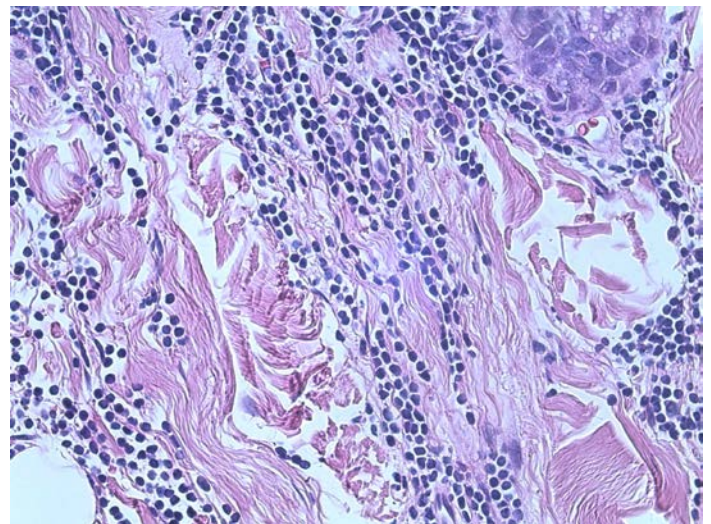


Figure 4. (H&E) The cells were composed of small lymphoid type cells.

DISCUSSION

Herein we describe a rare case of a well-defined skin manifestation, typical of B-CLL. A 65-year-old woman with a rosacea-like eruption of the nose had been misdiagnosed for years until these lesions were recognized as a clinical presentation of B-CLL.

Until recently, there were few studies about this extremely rare occurrence. Barzilai et al. addressed this topic with a case series [2]. Twelve cases of B-cell proliferative disorders mimicking granulomatous rosacea or rhinophyma were reported, two of which were B-CLL. These patients were two women, ages 67 and 74 years, respectively; they both presented with erythematous infiltrated papules and plaques on the nose. Their initial diagnosis was rhinophyma and the delay before diagnosis ranged from 3 to 10 years. The other patients exhibited different proliferative B-cell disorders, in particular PCMZL and PCFCL. Before these 2 cases, 7 cases of B-Cell neoplasm mimicking rosacea or rhinophyma were reported. Only two of them concerned B-CLL [3,4]. Even in these cases, the delay in diagnosis was significant because of the difficulty in distinguishing granulomatous rosacea and/or rhinophyma.

Leukemic skin infiltrates (leukemia cutis) occur in 4–20% of patients with leukemia. Any region of the body may be affected, most often with chronic and relapsing erythematous papules, nodules, plaques, ulcerations, and exfoliative erythroderma [5]. Leukemia cutis occurs mostly on the face but to mimic rosacea is considered extremely rare. Therefore, when a rosacea-like eruption is the unique clinical presentation it usually leads to a long diagnosis delay. These patients are often treated with topical or systemic antibiotics for at least a few months without any benefit before a biopsy specimen is obtained. Our patient was also treated for rosacea, which led to a delay in diagnosis of about two years. She was treated with topical and oral metronidazole, but her clinical manifestations did not improve until the definitive diagnosis.

In some cases the differential diagnosis would mostly include adnexal tumors, such as basal cell carcinoma, trichoepithelioma, or sebaceous hyperplasia. Furthermore, granulomatous diseases (infectious or noninfectious) such as sarcoidosis and T-cell lymphoma should be considered [2]. Other skin manifestations in patients with B-CLL include skin cancers (basal cell carcinoma, squamous cell carcinoma, malignant melanoma, Merkel cell tumor, and cutaneous T-cell lymphoma), viral infections (varicella-zoster, herpes simplex, and viral warts), and acute graft-versus-host disease [6]. Our patient, in complete absence of extracutaneous symptoms, reported that she had never suffered from any of these clinical manifestations.

Histological and immunohistochemical investigations are essential for diagnosis. In our case a biopsy confirmed the most salient characteristics of the skin manifestation of B-CLL. Histology may show either a patchy perivascular and periadnexal pattern or the presence of dense, monomorphous, diffuse, or nodular infiltrates of lymphocytes [7]. The subcutaneous fat is involved as a rule. The tumor is composed predominantly of small lymphocytes without atypical features. Small nodular areas with larger cells showing features of polymorphous or paraimmunoblasts (so-called ‘proliferation centers’) can be observed occasionally [7]. Immunohistology reveals the presence of B lymphocytes characterized by an aberrant immunophenotype (CD20+, CD5+) and monoclonal expression of immunoglobulin light-chains [7]. CD5 may be negative in some cases. A variable population of reactive T lymphocytes is usually present. The etiology remains unknown. Similar to descriptions of leukemic infiltrates of CLL localized to the sites of herpes zoster [8,9], lymphomatous/leukemic infiltrates localized to the face may represent the isomorphic phenomenon on the sites of a preexisting rosacea/ rhinophyma colonized by *Demodex Folliculorum*. In our case, however, there was no history of rosacea, but *Demodex Folliculorum* was detected in the biopsy specimen. It could be a trigger to the development of the disease. The prognosis seems not to be affected by skin involvement [7]. The treatment must be planned according to the hematological findings. Small solitary or clustered skin lesions may be removed surgically, but larger lesions may be treated by radiotherapy [10]. In our case, considering the extension and the localization of the skin involvement we decided to treat with radiotherapy. A good clinical and cosmetic result was obtained.

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