Chronic Lymphocytic Leukemia as an Unusual Cause of Rapid Airway Compromise

1. Introduction

Chronic Lymphocytic Leukemia (CLL) is the most common form of adult non-Hodgkin's lymphoma (NHL). This mature B-cell neoplasm has an incidence rate of 4.6 per 100,000 people in the United States each year [1-5]. This disease process predominantly affects adults over the age of 65 and is more common in men. CLL has an extremely variable course of presentation ranging from asymptomatic lymphocytosis to painless lymphadenopathy, hepatomegaly, splenomegaly, cytopenias, and infections. Patients may also present with the typical “B” symptoms of unintentional weight loss, fever, and drenching night sweats [3-5]. Rapid progression with transformation into an aggressive, high-grade NHL is known as Richter syndrome and may occur in up to 10% of patients with CLL [6].

She was started on chemotherapy with bendamustine and rituximab (BR) with a dramatic anatomic response after two cycles (Figure 2) and near complete response (CR) after four cycles when tracheostomy was removed. She eventually completed 6 cycles of BR in October 2012 and achieved a CR with complete marrow recovery confirmed by bone marrow biopsy. She has been closely followed in the outpatient setting with no evidence of disease recurrence as of March 2017.

Although indolent in nature, CLL can present as locally aggressive extranodal mass resulting in symptoms depending on the location and extent of tissue involvement [7]. We present a unique case of a patient with known CLL and stable lymphocytosis that developed an enlarging lymphoid base of tongue (BOT) mass which was identified as nontransformed CLL.

2. Case Presentation

A 62-year-old woman with untreated Rai stage II CLL was initially diagnosed in January 2007. At that time she was found to have an absolute lymphocyte count (ALC) of 8 g/dL, mild splenomegaly, and mild abdominal lymphadenopathy. She was otherwise asymptomatic. Bone marrow biopsy confirmed a monoclonal B-cell population of lymphocytes with CD5 and dim CD23 coexpression. Results at that time also showed normal cytogenetics, FISH negative for 11q deletion, 13q deletion, p53 deletion, and trisomy 12. β2 microglobin was 2.4 mg/L and ZAP 70 was normal. She underwent close monitoring and continued her follow-up visits.
observation for several years. During this period of surveillance, she did not experience any recurrent bacterial infections, and serial monitoring of quantitative immunoglobulins demonstrated IgG levels greater than 500 mg/dL.

However, in December 2011, she presented to the emergency department for complaints of feeling a globus sensation over the span of forty-eight hours. This symptom was associated with dysphagia, diffuse myalgia, high-grade fevers, and shortness of breath. ALC at the time remained stable at 8.5 g/dL with no evidence of anemia or thrombocytopenia. Lactate dehydrogenase (LDH) was normal at 188 IU/L and β-2 microglobin was elevated to 3.3 mg/L. CT scan of the neck demonstrated a large heterogeneously enhancing 4.1 cm mass involving the palatine tonsils causing severe narrowing of the hypopharynx. She was admitted and quickly developed worsening shortness of breath and stridor prompting otolaryngology consultation for urgent tracheostomy placement.

The BOT mass was biopsied at the time of tracheostomy placement. The pathology specimen demonstrated squamous mucosa with acute inflammation and reactive hyperplasia most consistent with bacterial infection and lymphoid tissue hyperreactivity. No monoclonal lymphocyte population was identified. Furthermore, no pathogens were isolated from biopsy specimen, blood cultures, or sputum cultures, and CRP was normal at 0.06 mg/dL. However, the decision was made to empirically treat her with ten days of moxifloxacin. A superimposed infection could not be identified. Furthermore, no pathogens were isolated from biopsy specimen, blood cultures, or sputum cultures, and CRP was normal at 0.06 mg/dL. However, the decision was made to empirically treat her with ten days of moxifloxacin. A superimposed infection could not be identified. Furthermore, no pathogens were isolated from biopsy specimen, blood cultures, or sputum cultures, and CRP was normal at 0.06 mg/dL. 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Figure 1: Immunohistochemical staining demonstrating a monomorphic population of CD20 positive lymphocytes with coexpression of CD5 typical of CLL.

Figure 2: Interval change in size of BOT mass on contrasted CT scan of the head and neck before (a1 and a2) and after (b1 and b2) two cycles of bendamustine and rituximab.
considered for patients with stage IIIE/IV disease for palliative intent [7].

In summary, this case illustrates an unusual presentation of CLL causing airway compromise in a patient with a non-transformed indolent lymphoma. Prompt recognition is required to institute early intervention. Initial biopsy findings were concerning a possible bacterial infection in this patient with known CLL. However, a superimposed bacterial infection may have complicated the clinical picture. Nevertheless, cases of infection mimicking CLL progression have been described and accurate identification of the underlying etiology is essential in order to make appropriate treatment decisions. This patient responded very well and achieved a CR after completing six cycles of BR. She has maintained a CR for over four years after completing cytotoxic therapy.

Conflicts of Interest

No financial or nonfinancial conflicts of interest exist in the preparation of this manuscript.

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