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A Case Report of an Aggressive Mantle Cell Lymphoma Masquerading as a Chronic Lymphocytic Leukemia With Catastrophic Treatment Complications

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Case Presentation:

Mantle cell Lymphoma (MCL) and chronic lymphocytic leukemia (CLL) account for majority of Small B-cell neoplasms (SBCNs) that express CD5 without CD10. We present a patient case with aggressive MCL presenting with leukocytosis and splenomegaly mimicking CLL initially on immunophenotypic features.

A 55-year-old presented with abdominal pain, drenching night sweats, early satiety, anorexia and significant weight loss for 8-month duration. On exam she had generalized lymphadenopathy and massive splenomegaly. Complete blood count showed pancytopenia with normal blood chemistry and LDH 778 U/L (Table 1). Differential count showed predominant lymphocytosis. Peripheral smear showed increased lymphocytes, condensed nuclear chromatin, smudge cells and occasional left shifted neutrophils. CT imaging showed diffuse lymphadenopathy and splenomegaly measuring 24 x 17 cm (Figure 1). Flow cytometry showed a large monoclonal B-cell population kappa light chain restriction, dim CD19, CD20, CD5, and variable expression of CD23. This population demonstrated moderate to bright CD38 and negative for CD10 and CD11c (Figure 2). The findings were most compatible with a diagnosis of chronic lymphocytic leukemia. Fish panel was pending at this point.

Due to B-symptoms and ongoing pain, decision was made to initiate FCR chemotherapy (Fludarabine, Cyclophosphamide, Rituximab). A bone marrow core biopsy (40x magnification) showing large lymphoid cells with slightly folded nuclei and prominent nucleoli (Figure 3A), was performed. Bone marrow aspirate (100x magnification) showed increased lymphocytes, condensed nuclear chromatin, smudge cells and occasional left shifted neutrophils. CT imaging showed diffuse lymphadenopathy and splenomegaly measuring 24 x 17 cm (Figure 1). Flow cytometry showed a large monoclonal B-cell population kappa light chain restriction, dim CD19, CD20, CD5, and variable expression of CD23. This population demonstrated moderate to bright CD38 and negative for CD10 and CD11c (Figure 2). The findings were most compatible with a diagnosis of chronic lymphocytic leukemia. Fish panel was pending at this point.

MCL is an aggressive non-Hodgkins lymphoma with poorer prognosis of all other lymphomas. Though not curable outside of transplant, frequent remissions (60%-90%) can be obtained albeit short lasting (1-2 years). MCL often misdiagnosed as CLL. Flow cytometric immunophenotyping often helps differentiate CLL from MCL, and a characteristic CLL phenotype is considered essentially diagnostic. There is significantly higher expression of CD23 in CLL. MCL typically is negative for CD23, but 25 % cases can be dim positive. On the other hand can occasionally be CD23 negative. Thus NHL often can be less obvious and confounds the diagnosis if a leukemic phase occurs at presentation. MCL is more specifically identified by the presence of the translocation t(11;14)(q13;q32), but cyclin D1-negative variants do exist. FISH assay is by far the most sensitive and specific technique for t(11;14) identification. This should be considered in newly diagnosed CLL patients with atypical features especially B grade symptoms at initial presentation, massive splenomegaly, immunophenotypic feature showing moderate bright CD20 and bright surface light chain expression. Given the importance of stem cell transplants for MCL patients, it is important to recognize this, as fludarabine therapy can impair successful collection of stem cells. Bone marrow biopsy though not required for diagnosis for CLL, but should be considered in atypical presentations, discordance in the immunophenotypic markers, presence of cytopenias and preferably prior to treatment initiation.

Rituximab induced CRS or splenic rupture is very rarely reported in literature. There is emerging evidence which implicates IL-6 as a central mediator in CRS. Toclilizumab (IL-6 blocker) is used as first line immunosuppressive therapy in clinical trials. Ophthalmic side effect of rituximab is reported including transient ocular edema, transient visual changes and severe loss of visual acuity; however these are very rare. The risk of reaction may correlate with the aggressiveness and burden of the disease. Hence we should consider aggressive hydration, pre-treatment with steroids, antihistamines and holding or dose reduction of rituximab with first cycle. Post-marketing surveillance database on rituximab indicates a mortality of 0.04–0.07% associated with the drug, however due to its increasing use, there should be high index of suspicion for these side effects as they are life threatening.

References: