Hodgkin Lymphoma in a Patient with Chronic Lymphocytic Leukaemia A Rare Presentation of Richter's Transformation

Abstract:

P De La Harpe Golden, C Egan, M Leader, PT Murphy, J Quinn
Beaumont Hospital, Beaumont, Dublin 9

Abstract

Richter's transformation of chronic lymphocytic leukaemia (CLL) to high-grade B-cell Non-Hodgkin lymphoma occurs in < 5% of CLL cases. Transformation of CLL to Hodgkin Lymphoma is a much rarer event and here we describe a patient who developed Richter's transformation into a Hodgkin Lymphoma presenting as rapidly progressive hepatosplenomegaly.

Introduction

CLL is the most common haematological cancer affecting adults with an incidence of 4/100,000 in Europe. CLL is a chronic disease often remains indolent for many years with anti-CLL therapy only commenced when there is evidence of advanced/progressive disease. Mostly CLL is a low-grade disorder, however in <5% of cases the disease undergoes transformation to an aggressive high-grade lymphoma, so-called Richter transformation (RT) with the majority of patients developing a high-grade B-cell Non-Hodgkin Lymphoma e.g. Diffuse Large B-Cell Lymphoma (DLBCL). In contrast, RT to Hodgkin Lymphoma (HL-variant RT) is a very rare occurrence.

Case Report

A 76 year old man presented with a short history of fever, weight loss, abdominal distension, and deranged liver function tests (Bilirubin 21 mol/L, alkaline phosphatase 401 IU/L, gamma-glutamyl transferase 148 IU/L) with lactate dehydrogenase also elevated at 602 IU/L on admission. He had been diagnosed with CLL 5 years previously and his disease was known to carry the 17p deletion which is associated with an adverse outcome. Previous treatment had included corticosteroids and a 12-week course of the anti-CD52 monoclonal antibody alemtuzumab last administered 3 months previously. Computed Tomography (CT) scan showed massive hepatosplenomegaly with multiple low-attenuation foci. In light of his recent immunosuppressive treatment infection was suspected, however liver biopsy displayed features consistent with infiltration by Hodgkin lymphoma with characteristic positive staining for CD15 and CD30. Despite immediate treatment with high-dose methyl-prednisolone, he passed away just 25 days following admission.

Discussion

In this case we describe a patient with poor prognosis CLL who developed a HL-variant RT. Unlike de novo DLBCL, RT carries a very poor prognosis with median overall survival of approximately 8 months'. Although, it has been suggested that the rarer HL-variant RT may carry a better prognosis than the typical DLBCL transformation", the disease may follow a heterogeneous course with widely varying median survival reported, ranging from 9 months to 39.5 months". HL is characterised by the presence of Reed-Sternberg (RS) cells and two variants of RS cell distribution have been described in HL-variant RT. The first consists of RS cells scattered on a background of CLL cells perhaps representing a ‘true’ transformation of the same clonally identical B cell as was previously present, whilst the latter shows RS cells separate to the CLL cells, in a polymorphous inflammatory background and may represent a de novo HL arising in a patient with CLL". The majority of HL-variant RT are thought to consist of the de novo variant6 unlike the more common variant of RT where approximately 20% of DLBCL lineages are clonally unrelated to the underlying CLL. This differentiation in DLBCL-RT has significant clinical implications with one study of 86 cases showing a median survival of 42.5 months for clonally unrelated DLBCL versus just 14.2 months for clonally related DLBCL. Although all HL histological subtypes have been described in reports of HL-variant RT', it is not known if prognosis varies according to subtype.

Overall, HL-variant RT is less well studied than the DLBCL variant, however, some studies suggest a more prominent role for Epstein Barr Virus (EBV) infection in the development of HL-variant of RT'. This differentiation in DLBCL-RT has significant clinical implications with one study of 86 cases showing a median survival of 42.5 months for clonally unrelated DLBCL versus just 14.2 months for clonally related DLBCL. Although all HL histological subtypes have been described in reports of HL-variant RT', it is not known if prognosis varies according to subtype.

Correspondence: P De La Harpe Golden
Beaumont Hospital, Beaumont, Dublin 9
Email: peter.delaharpe.golden@gmail.com

References


Hodgkin Lymphoma in a Patient with Chronic Lymphocytic Leukaemia A Rare Presentation of Richter's Transformation


