SEER Inquiry System - Report

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Question 20110017

References:

Heme & Lymph Manual & DB

Question:

Multiple primaries--Heme & Lymphoid Neoplasms: How many primaries are reported if a patient originally diagnosed with CLL is subsequently diagnosed several months later on a bone marrow biopsy with Richter's syndrome that transformed into a large cell lymphoma? See Discussion.

Discussion:

Per reviewed resources, the described condition is rare. Should the histology remain CLL or be changed to large cell lymphoma?

Answer:

For cases diagnosed 2010 and forward, access the Hematopoietic Database at http://seer.cancer.gov/seertools/hemelymph.

This case is accessioned as two primaries per Rule M10 which states to abstract multiple primaries when a neoplasm is originally diagnosed as a chronic neoplasm and there is a second diagnosis of an acute neoplasm more than 21 days after the chronic diagnosis. The first primary is CLL [9823/3] and it is a chronic neoplasm. The second primary is diffuse large B-cell lymphoma (DLBCL) [9680/3] and it is an acute neoplasm.

Richter syndrome (RS) is a complication of B cell chronic lymphocytic leukemia (CLL) or hairy cell leukemia (HCL) in which the leukemia changes into DLBCL. There is also a less common variant in which the CLL changes into a Hodgkin lymphoma. Richter's transformation affects about 5% of CLL patients. Richter syndrome is listed under the Alternate Names section in the Heme DB for diffuse large B-cell lymphoma [9680/3].

SEER*Educate provides training on how to use the Heme Manual and DB. If you are unsure how to arrive at the answer in this SINQ question, refer to SEER*Educate to practice coding hematopoietic and lymphoid neoplasms. Review the step-by-step instructions provided for each case scenario to learn how to use the application and manual to arrive at the answer provided. https://educate.fhcrc.org/LandingPage.aspx (https://educate.fhcrc.org/LandingPage.aspx).

History:

This is a new primary, diffuse large B-cell lymphoma (DLBCL) 9680/3. Richter syndrome (RS) is a complication of B cell chronic lymphocytic leukemia (CLL) or hairy cell leukemia (HCL) in which the leukemia changes into DLBCL. There is also a less common variant in which the CLL changes into a Hodgkin lymphoma. Richter's transformation affects about 5% of CLL patients.

Cancer Site Category:

Heme & Lymphoid Neoplasms

Data Item Category:

N/A

Other Category:

Multiple primaries

Year:

2011