

Primary splenic red pulp diffuse large B-cell lymphoma with anaplastic features

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Received: 01 March 2016; Accepted: 17 March 2016; Published: 06 April 2016.

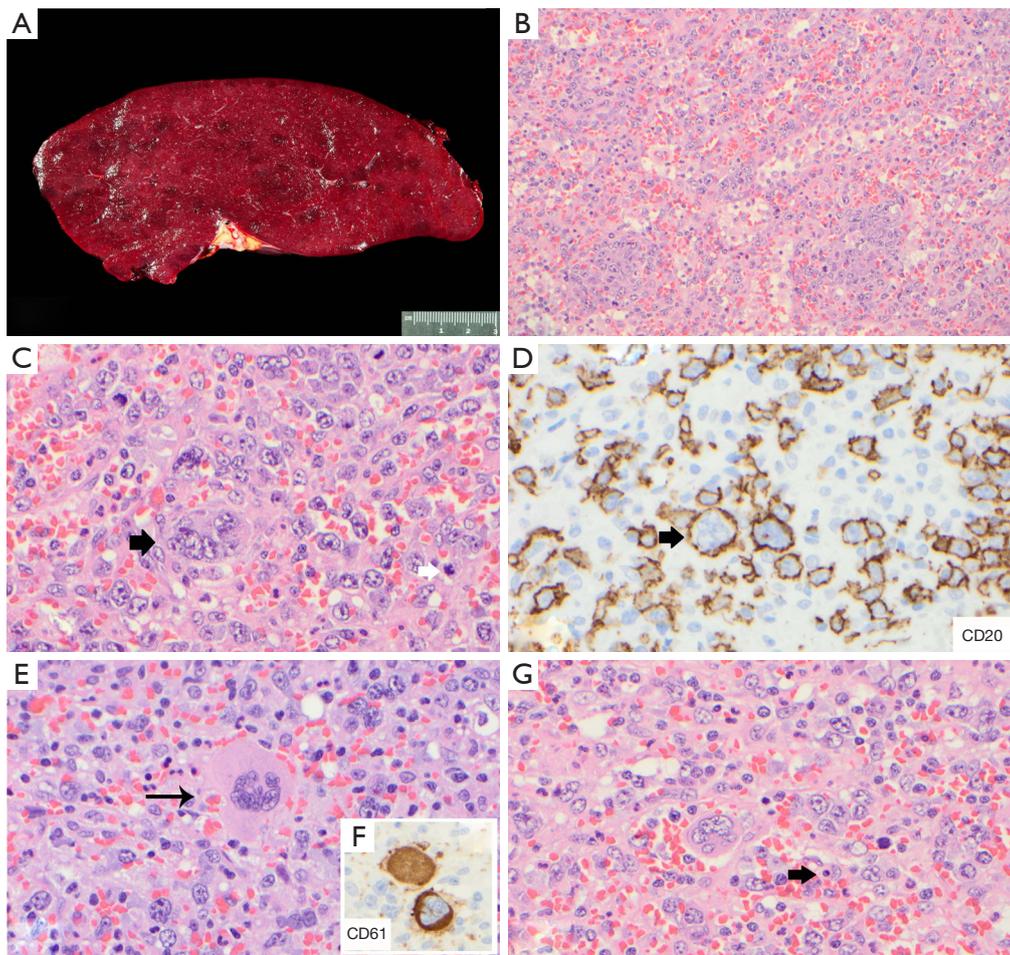
doi: 10.21037/sci.2016.03.04

View this article at: <http://dx.doi.org/10.21037/sci.2016.03.04>

A 76-year-old man presented with B symptoms, anemia (Hb 11 g/dL), and thrombocytopenia (96,000/ μ L) with elevated serum LDH (1,916 U/L) and ferritin (4,148 μ g/L). Radiology imaging showed hypermetabolic splenomegaly without lymphadenopathy. Bone marrow biopsy showed no evidence of lymphoma or other malignancy. The patient underwent splenectomy. Grossly, the spleen measured 19.0 cm \times 14.0 cm \times 8.5 cm and 1,410 g, with a red-brown homogenous cut surface without any discernible nodules (A). Microscopically, the splenic architecture was replaced by a diffuse proliferation of neoplastic cells involving the red pulp arranged along splenic cords (B). Splenic sinuses were mostly patent and occasional neoplastic cells were identified within sinuses. Residual and atrophic white pulp was present in the background. The neoplastic cells were large, pleomorphic, with eosinophilic cytoplasm, large round to irregular nuclei, vesicular chromatin and prominent nucleoli (C). Some of the neoplastic cells were very large with multinucleation, simulating megakaryocytes (C & D, black arrow). Scattered megakaryocytes (E,F), nucleated erythrocytes and occasional hemophagocytosis (G, arrow) were also recognized. In contrast to megakaryocytes, the multinucleated neoplastic cells showed vesicular chromatin with distinct nucleoli. By immunohistochemistry, the lymphoma cells were positive for CD20 (D), PAX5, BCL2, c-MYC (60%), MUM-1 (40%),

and negative for CD3, CD10, CD30, and BCL6. Ki-67 proliferative index was 90%. EBER-ISH was negative. Flow cytometry demonstrated a surface kappa-restricted large B-cell population, positive for CD19, CD 20, and negative for CD5 and CD10. Conventional karyotyping analysis showed: 78,XX,Y,del(10)(q24q25),+11,add(12)(p12),+15,+15,+18,+4mar[cp2]/46,XY[18]. Fluorescent in situ hybridization study demonstrated BCL6 rearrangement, without MYC or BCL2 rearrangement. The patient was diagnosed with primary splenic red pulp diffuse large B-cell lymphoma with anaplastic features and died two weeks post-splenectomy due to sepsis, before the initiation of chemotherapy.

Involvement of spleen by diffuse large B cell lymphoma (DLBCL) is not uncommon and patients frequently have systemic DLBCL. Primary splenic DLBCL is rare, usually derived from splenic white pulp; presents as >1 discrete nodules grossly. Primary red pulp DLBCL as we described here is extremely rare and has not been included in the current 2008 WHO classification. Characteristic features include diffuse red pulp involvement, hemophagocytosis, extramedullary hematopoiesis in spleen, with frequent liver and bone marrow involvement and absence of systemic lymphadenopathy. The cell of origin is unknown. Whether it is a distinct entity or a transformation from other low-grade lymphomas remains to be explored.



Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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Cite this article as: Khandakar B, Wang W, Li S. Primary splenic red pulp diffuse large B-cell lymphoma with anaplastic features. *Stem Cell Investig* 2016;3:9.