**Spleen involvement in DLBCL**

Diffuse large B-cell lymphoma of spleen accounts for about 1/3 of all primary splenic lymphomas. Splenic lymphomas are the most frequent splenic malignancy. Diagnosis by non-surgical imaging studies is problematic; CT accuracy in depicting splenic lymphoma is approximately 22–65%. Diagnosis can be made by either splenectomy or ultrasonically guided percutaneous splenic tissue core biopsy.

- Primary (1%) and secondary (>30%)
- Differential diagnosis: splenic cysts, abscess, hematoma, and non-lymphoid neoplasms; if retroperitoneal lymphadenopathy is present on imaging, it suggests primary splenic lymphoma
- **Definition of primary splenic DLBCL**: The definition of splenic lymphoma in the literature has been inconsistent
  - Catherian et al: splenomegaly without peripheral lymphadenopathy, with or without involvement of regional lymph nodes, bone marrow or liver.
  - Das Guta et al: splenomegaly without any evidence of disease elsewhere; liver biopsy specimen, as well as para-aortic and mesenteric lymph nodes, should be free of malignant lymphoma.
  - Sharin et al: splenomegaly without significant lymphadenopathy and no hepatomegaly or peripheral blood involvement.
  - Ahmann et al: staging system in which stage I indicates disease limited to the spleen, stage II includes splenic hilar node involvement, and stage III involvement of liver or lymph nodes beyond the splenic hilum.
  - several authors favour inclusion as stage II, perhaps by the designation “E”, those cases with contiguous extension and invasion of adjacent structures as seen in seven of 10 cases with diffuse large cell pathology reported by Harris et al. Spleens are large (moderate to marked splenomegaly) with an average weight of 1 kg
- The prevalence of HCV infection is about 50%
- Two types: tumor mass and diffuse
- **Primary DLBCL that presents as a tumor mass**:
  - Patients have symptomatic splenomegaly
  - Presumed to be derived from white pulp
  - Age: >50, F>M
  - There is usually single tumor mass, which is large and it occupies more than 50% of the spleen; this pattern is not specific to primary DLBCL as secondary involvement by DLBCL will have the pattern
  - Bridge of splenic capsule and local invasion of neighbouring structures is common
  - In contrast, other NHL in the spleen usually show military pattern (tiny nodules reflecting white pulp involvement) or homogeneous diffuse infiltration
  - Bone marrow usually negative at the time of presentation
  - Both GC and ABC types were described as well as T/HRBCL
  - Necrosis is very common; secondary infection of necrotic tissue may further delay diagnosis as the symptoms and microbiological cultures will favor splenic abscess
  - Weight loss, fatigue, fever, night sweats are common
  - Labs: normochromic normocytic anemia, increased ESR, increased LDH
  - Left-sided pleural effusion may be seen in some patients
  - Ann Arbor stage II (also -/+E due to extranodal spread) in most cases
  - Complete response to chemotherapy for DLBCL is often achieved, but it is still more aggressive than equivalent stage DLBCL of most other sites
  - Multiple (confluent) nodules may be present – same histology, probably same as uninodular
Vascular invasion may be seen – different than intrasinusoidal distribution in red pulp type of DLBCL
Cytological features: centroblastic morphology is more common than immunoblastic; anaplastic cells may be scattered in the tumor

- **Primary diffuse DLBCL that is manifesting in the red pulp:**
  - Only <30 cases described so far
  - Involves splenic sinuses and cords
  - Age: >60 year old, M>>F
  - Labs: very increased LDH, cytogenetic findings: complex chromosomal aberrations
  - Average spleen size 1.3 kg.
  - Clinical stage 4B; PB involvement about 40%
  - Red pulp primary DLBCL is an aggressive disease
  - Often it is disseminated in the bone marrow (intrasinusoidal and interstitial) and liver; typically, LNs are not involved, but involvement of perisplenic LN has been reported.
  - Spread in LN different than intravascular DLBCL
  - Hemophagocytosis in spleen and bone marrow, but not hemophagocytic syndrome
  - Extramedullary hematopoiesis in spleen
  - Negative for HIV, HTLV-1, or EBV; about 80% CD5+ and in 30% CD10+

- **Other splenic lymphomas with large B-cells:**
  - T/HRBCL
  - Splenic B-cell lymphomas with >55% prolymphocytes in blood are considered to be a transformation of underlying low-grade B-cell lymphoma including SMZL or splenic diffuse red pulp small B-cell lymphoma and they are often p53+, de(7q)

**Literature:**