PathMD™: Board Review Letter

Author: Philip Ferguson, M.D. **Spleen – Part 1** Volume 1, Number 15

Case #1 The images associated with this case are from a moderately enlarged spleen in an individual with a WBC count greater than 150,000. The flow cytometry profile showed the population to express CD13, CD33, and CD117. Based on the morphology and flow cytometry, the best diagnosis is:

Answer: C. AML AML involving the spleen replaces the red pulp. Think of the spleen as a large lymph node where the blood and lymphoid tissue meet. Lymphomatous processes tend to start in the white pulp and spill over into the red pulp, and leukemia tends to start in the red pulp before spilling over into the white pulp. This is the first place to start when evaluating the spleen. Ask the question, "is this a red pulp process, or a white pulp process?" Then consider entities in the appropriate category first. This case of AML shows red pulp predominance by cells with granulocytic features. They appear arrested early in the maturation sequence, raising the suspicion of an acute leukemia over CML. Helpful stains include CD34 (immature marker), CD117, myeloperoxidase, Tdt, and B or T cell markers. These should help differentiate an ALL from an AML.

Case #2 A 4,000 gram spleen was removed from a man and has the histology presented for this case on the website. Special stains were performed. The cells of interest were positive for CD68 and negative for S-100, CD1a, and PAS. Based on these findings, the best diagnosis is:

Answer: B. Histiocytic Proliferations (Lipid/Ceroid Histiocytosis and Gaucher Disease) Ceroid is a general term used to describe benign histiocytic proliferations. These proliferations develop secondarily in several settings. ITP is one of the more common, and is due to the accumulation of lipid-rich membranes as the histiocytes phagocytose platelets. CML, light chain deposition disease, and inherited lipidoses are also other causes. Gaucher disease is similar to ceroid histiocytosis, but the spleen is often much larger (mean volume 19.8 times normal). It is due to a defect in lysosomal glucocerebrodase. In ceroid histiocytosis, histiocytes will react with acid fast stains and PAS/d, but in Gaucher disease the cells stain for iron. (Sternberg, 4th Ed., p. 863-864)

Case #3 The images for this case come from a 60 year old man with an enlarged spleen and a hilar mass. Based on the information, the best diagnosis is:

Answer: D. Hodgkin Lymphoma (HL) is much less common to be diagnosed in the spleen today because staging procedures are much less common. The diagnostic criteria for HL is the same in the spleen as it is in the lymph node. It is important to remember when staging a spleen for HL, the number of nodules of HL is important prognostically. Care needs to be taken when grossly evaluating the spleen because some lesions are as small as 1mm. Five nodules appears to be an important cutoff number for the number of nodules involved by HL. Subclassification of HL in the spleen is optional, and is not a consideration in treatment. (Sternberg, 4th Ed., p. 860-861)

Case #4 The images for this case come from an elderly female with a history of an autoimmune disorder. The spleen was 1,500 grams and noted to have "tumor nodules". Based on the findings, the best diagnosis is:

Answer: C. Kaposi's Sarcoma (KS) is a rare vascular tumor in the spleen. KS is now most commonly associated with patients with HIV. There is an additional association with HHV8. Morphologically, KS is characterized by spindle cells in short fascicles with red cells present in slit-like spaces. The mitotic activity is usually low, and there is little atypia of the cells. A minor infiltrate of plasma cells is also common. In comparison to other spindle cell soft tissue tumors, KS will mark with vascular markers (when the vascular component is difficult to recognize). (Sternberg, 4th Ed., p. 72-73)

Case #5 Images for this case come from a spleen in a 60 year old male with a ruptured spleen. Special stains showed positivity for CD20, bcl-1, and CD43. Based on these findings, the best diagnosis is:

Answer: B. Mantle Cell Lymphoma (MCL) has some similarities to CLL/SLL, including expression of CD5 and CD43 (think of CD43 as a surrogate to CD5). MCL does not usually express CD23. In addition cyclin D1 is positive and supports the underlying genetic alteration, t(11;14). Tricks to remember are that bcl-1 is the same thing as cyclin D1, and not all cases will be positive for cyclin D1 (a nuclear stain). Almost all cases are positive by FISH. The typical morphology of MCL is of small lymphocytes with dense chromatin and irregular nuclear membranes. The blastoid variant may look more like an ALL or sometimes even a DLBCL (pleomorphic variant). This is why we do a large panel of immunostains. (Sternberg, 4th Ed., p. 854-855)

Case #6 These images are of a 1,000 gram spleen from a patient with multiple episodes of hemolysis resistant to steroids. Based on the images and the patient's history, the best diagnosis is:

Answer: A. Splenic Congestion secondary to autoimmune hemolytic anemia The spleen can become engorged with RBCs in hemolytic processes. One may also see extramedullary hematopoesis within the sinuses. In passive congestion from cirrhosis or other process, the extramedullary hematopoesis is not present, and there is often increased reticulin fibrosis. (Sternberg, 4th Ed., p. 861-862)

Case #7 These images are from the spleen of a 30 y/o pregnant patient. The spleen weighed 2,000 grams. Stains for AFB and fungi were negative. The best diagnosis is:

Answer: E. Necrotizing Granulomatous Splenitis (NGS) NGS is a somewhat non-specific finding. They are well described in patients with lymphoproliferative disorders (childhood AML) and infectious etiologies (Bartonella Henselae, other bacterial and fungal infections). It is important to remember that negative stains do not rule out a disease category. A good note to place on the diagnostic line that infection must be clinically excluded.

Case #8 The images for this case are from a 1,000 gram spleen from a 75 year old man. Special stains were positive for CD45 and CD20, but negative for CD3 and CD15. Based on these findings, the best diagnosis is:

Answer: A. Diffuse Large B-Cell Lymphoma (DLBCL) is a common primary lymphoma of the spleen. Similar to Hodgkin lymphoma, but unlike other small cell lymphomas (MCL, MZL, CLL/SLL), it often produces discrete masses. They can also arise out of a background of a lower grade lymphoma. (Sternberg, 4th. Ed., p. 858-859) This case is an example of an immunoblastic DLBCL. The immunoblastic morphologic variant is characterized by >90% of the cells having a single centrally located nucleolus. Centroblasts represent <10% of the cells. Additionally, stains may be necessary to differentiate these cases from a plasmablastic plasma cell myeloma. (WHO, 2001, p. 172)

References:

Pathology and Genetics: Tumours of Haematopoietic and Lymphoid Tissues (World Health Organization Classification of Tumours), ES Jaffe, 2001.

Sternberg's Diagnostic Surgical Pathology, SE Mills, et al. Fourth Edition. 2004.

Notes for question set:[1]

[1] PathMD strives for the highest quality and accuracy. However, the *PathMD: Board Review Letter* is for review purposes and not meant for clinical decision making. It should not be used in place of review of primary reference texts and the current medical literature. If inaccuracies are identified, please notify us so that a correction may be published. (info@PathMD.com)

