PathMD™: Board Review Letter

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Case #1 These images are from a 300 gram spleen, which was noted to have multiple blood filled nodules. The cells of interest were positive for CD68 and CD31 but negative for CD34. Based on these findings, the best diagnosis is:

B. Littoral Cell Angioma

Answer: B. Littoral Cell Angioma is a vascular lesion, which resembles splenic sinuses. Interestingly, the cells express CD68 (histiocyte marker) and CD31 (endothelial marker), but they lack expression of CD34 (present in other vascular lesions). (Sternberg, 4th Ed., p. 869-870) The appearance of these lesions matches somewhat the immunoprofile. They are vascular, and therefore express CD31, but they also look and act like histiocytes (CD68 expression). These features make it easier to remember the immunophenotypic profile.

Case #2 The images for this case are of a spleen from a 70 year old man with a history of "leukemia." A splenectomy was performed for symptoms. Flow cytometry show dim positivity for CD20 and kappa light chains. In addition, CD5 and CD23 were positive and CD10 was negative. Based on these findings, the best diagnosis is:

A. CLL/SLL

Answer: A. CLL/SLL is not uncommon to involve the spleen. Like most lymphomas involving the spleen it expands the white pulp and then spills over to involve the red pulp. CD5 and CD23 expression are helpful in diagnosis. Some cases may be difficult to differentiate from Hairy Cell Leukemia. The absence of DBA.44 and cyclin D1 expression are helpful in this differential. Remember, in addition to mantle cell lymphoma, hairy cell leukemia can express cyclin D1. (Sternberg, 4th Ed., p. 852-854)

Case #3 During an operation, the spleen was noted to have multiple masses. Representative sections from these areas are show in the images for this case. Based on the morphologic findings, the best diagnosis is:

A. Hemangioma

Answer: A. Hemangioma Hemangiomas of the spleen share the same characteristics of hemangiomas in other locations. CD31 and CD34 are two common vascular endothelial markers that should be positive. Refer to the littoral cell angioma question for comparative features.

Case #4 The images for this case are from a spleen removed for "symptoms." It weighed 1,000 grams and had multiple small white nodules throughout the parenchyma. Special stains were positive for CD20 but negative for CD5, CD43 and CD10. Based on these findings, the best diagnosis is:

C. Marginal Zone Lymphoma

Answer: C. Marginal Zone Lymphoma (MZL) is a relatively new category. In the past many of these lymphomas were placed in the CLL/SLL or MCL category. MZL has a few defining characteristics. First, there is more abundant cytoplasm than in CLL/SLL or MCL. This is why the marginal zone is noticeable on H&E sections of the spleen. Second, there is more heterogeneity of the cell size and shape compared to MCL or CLL/SLL. The cells are small to intermediate in size, and there may be plasmacytoid differentiation. This can be so prominent at times, differentiation from a lymphoplasmacytic lymphoma may require clinical correlation. The may also take on a monocytoid B-cell appearance. Immunophenotypically, MZL is an exclusionary diagnosis. Meaning it is the lack of defining immunophenotypic characteristics that helps us classify a MZL (CD5-, CD10-, CD20+, light chain restricted). (Sternberg, 4th Ed., p.855-857)

Case #5 Multiple vascular lesions were found in a 70 year old male with no other significant past medical history. No other lesions in other organ were found. A splenectomy was performed, which showed multiple blood filled vascular areas. Representative images from the spleen are shown for this case. Based on the findings, the best diagnosis is:

D. Angiosarcoma

Answer: D. Angiosarcoma of the Spleen This is a rare case of angiosarcoma of the spleen. There are areas which resemble Kaposi's sarcoma. However, there is no history of immunodeficiency in the patient. In addition, note the areas of necrosis, pleomorphism, and anastamosing vascular spaces with large atypical endothelial cells. These features combined support the diagnosis of angiosarcoma with kaposiform features. There are only approximately 130 cases of primary angiosarcoma limited to the spleen reported in the medical literature.

Case #6 A calcified lesion was found incidentally in a 20 y/o. At surgery it was found to originate from the 400 gram spleen. Based on the morphologic findings, the best diagnosis is:

D. Epithelial cyst

Answer: D. Epithelial Cyst Cysts in the spleen are not common. If there is a true lining, then they are considered true cysts. Epithelial cysts with squamous lining are thought to arise from metaplasia of a mesothelial cyst. Mesothelial cysts are thought to be secondary to trauma. If there is no true lining, then the cysts are considered false cysts. (Sternberg, 4th Ed., p. 869)

Case #7 A 24 y/o male with a history of short stature and renal failure (s/p renal transplant) presented with multiple splenic abscesses. A splenectomy was performed. Representative sections from the spleen are shown for this case. Based on the clinical history and morphologic findings, the best diagnosis is:

E. Cystinosis

Answer: E. Cystinosis involving the Spleen Cystinosis is an autosomal recessive disorder that affects approximately 400 individuals in the United States. These individuals are usually of very short stature and look child-like. Renal failure develops early on, requiring transplant. Interestingly, the transplanted kidney is not affected because there is normal transport of cystine from the lysosomes. The classic finding in urine cytology are hexagonal crystals. These are not easily identified on H&E sections from other tissue sights. Oxalosis and

Case #8 A 45 y/o male presents to his physician with early satiety. Physical examination was significant for an enlarged spleen. A CBC & bone marrow examination was performed at an outside institution that was significant only for a monocytopenia. No flow cytometry was performed. The patient now undergoes a splenectomy due to continued symptoms. Representative histologic sections of the ____ gram spleen are shown on the website. Flow cytometry is performed and shows a monoclonal kappa population that expresses CD103, CD11b, CD25, and CD20. Based on these findings, the best diagnosis is:

E. Hairy Cell Leukemia

Answer: E. **Hairy Cell Leukemia (HCL).** This is a very popular topic to test. Probably because it is well described, has very characteristic features, and has an excellent long term prognosis. Three things should be emphasized in HCL. First, it is fairly unique among lymphomas in that it is associated with a monocytopenia. Second, there is a very unique flow cytometry immunophenotype (CD103, CD25, CD11b, and CD19/20 positive). Finally, it is notorious for being overlooked in specimens, particularly bone marrow biopsies. Histologically, HCL is described as having a fried egg appearance (i.e. it has more abundant cytoplasm and cell borders are more easily identified than in usual lymphomas). If one keeps these things in mind no HCL question should sneak by.

Question #1 Examination of a spleen shows dilated sinuses containing histiocytic appearing cells with occasional emperipoesis. Lympocytes are mixed in, and plasma cells are noted to cuff around small vessels. You suspect a diagnosis, which should have which of the following staining characteristics:

A. S-100 +, CD1a =

Answer: A. The description is of Rosai-Dorfman disease. The histiocytes are positive for S-100 and negative for CD1a. Dual positivity of S-100 and CD1a is c/w histiocytosis X (eosinophilic granuloma). PAS positivity raises the possibility of something like Whipple's disease.

Question #2 A patient is diagnosed with CLL/SLL. Which of the following are associated with a poor prognosis:

B. Del 11q23
C. Del 13q14
D. Trisomy 12
E. Both A and C are correct

Answer: D. Del 13q14 is a good prognostic marker in CLL/SLL. Del 11q23 and trisomy 12 are associated with a poor prognosis (I do not know of anything involving 11q23, which has anything but a poor prognosis). CD38 and ZAP-70 expression are also associated with a more aggressive course.

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)

