PathMD™: Board Review Letter

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Case #1 This patient has a 52-year-old female with a history of an enlarged right cervical lymph node. The patient also has hyper-gammaglobulinemia. Representative sections of the lymph node are shown in images for this case. Special stains performed but not shown, include CD21 which highlights follicular dendritic cells surrounding the vessels, and EBV by in situ hybridization, which highlights scattered cells. Numerous cells are CD2, CD3, and CD5 positive but have markedly decreased CD7 expression. CD30 highlights rare scattered cells. Based on these findings in the patient's clinical history, what is the best diagnosis?

A. Adult T-cell leukemia/lymphoma
B. Lymph node involvement by Sezary syndrome
C. Mixed cellularity Hodgkin lymphoma
D. Angioimmunoblastic T-cell lymphoma
E. Anaplastic large cell lymphoma

Answer: D. This case illustrates a classic example of angioimmunoblastic T-cell lymphoma (AILT) arising in a female with a history of hypergammaglobulinemia. The images for this case highlights the polymorphous appearance of a lymph node with diffuse effacement of the architecture. Note the high endothelial venules. There are also many cells that resemble Hodgkin cells. All these features in combination with the findings of rare scattered EBV positive cells in the highlighting of Kerry venular follicular dendritic cells is classic for AILT. (WHO, pages 225-226)

Case #2 The patient is a 40-year-old man with a history of a large anterior mediastinal mass and multiple axillary lymph nodes concerning for lymphoma. Representative images of the histologic sections from the lymph node are shown. Based on the morphology in the immunophenotypic findings shown, what is the best diagnosis?

A. Nodular lymphocyte predominant Hodgkin lymphoma
B. Anaplastic large cell lymphoma
C. Classical Hodgkin lymphoma -- lymphocyte depleted
D. Classical Hodgkin lymphoma -- syncytial variant
E. Classical Hodgkin lymphoma -- nodular sclerosing subtype

Answer: D. The answers for this questioner not entirely fair. This case represents classical Hodgkin lymphoma, nodular sclerosing in subtype, but it represents the syncytial variant. The syncytial variant classically has areas of necrosis and sheets of Hodgkin cells. Therefore one can argue correctly that both D. and E. are correct answers. The lymphocyte depleted variant of Hodgkin lymphoma can only be diagnosed if the nodular strokes in subtype is excluded. (WHO, pages 244-253)

Case #3 This patient has a 40-year-old Hispanic male with no known past medical history presents to the hospital with fever, malaise, and general fatigue. During the medical workup for CD4 count was found to be less than 10. A bone marrow biopsy was also performed, and Representative images and special stains are shown. Based on the findings in this case, what is the best diagnosis?

A. Plasma cell dyscrasia
B. Cryptococcal infection
C. Pneumocystis carinii infection
D. Blastomycoses infection
E. Histoplasmosis infection

Answer: E. This case represents histoplasmosis in a patient with newly diagnosed HIV in a CD4 count <10. It is not uncommon to see a plasmacytosis in the bone marrow patient with HIV. The positivity the GMS stain identifies a disorder is being fungal. The organisms are very small approximately 4 µm with narrow-based budding. Both cryptococcal and histoplasmosis infections can have narrow-based buds, but Cryptococcus usually has a prominent capsule. Blastomycoses is characterized by broad-based buds. Additional information not given is that the patient had a very high urine histoplasmosis antigen, which is a very good screening tool.

Case #4 The patient is a 70-year-old male with a history of an Ig-G kappa SPEP. Laboratory studies showed a prolonged PT and aPTT. A bone marrow biopsy was performed for the monoclonal gammopathy, and representative images are shown. Based on the findings, what is the best explanation for the patient's problems?

A. Multiple myeloma
B. AA amyloidosis
C. AL amyloidosis
D. HIV infection
E. Monoclonal gammopathy of undetermined significance

Answer: C. AL amyloidosis is associated with plasma cell dyscrasias. The amyloid circulating in the blood can passively buying to factor X, resulting in an acquired inhibitor. *Test Taking Strategy.* This is a classic association and popular exam question. It is also popular to get to and answers against each other such as B. and C. in this case.

Question #1 In case #4 is the most likely etiology of the prolonged PT and aPTT?

A. Thrombin deficiency
B. Dysfibrinogenemia
C. Factor X deficiency
D. Factor XI deficiency
E. Factor II deficiency

Answer: C. An acquired factor X inhibitor is characteristic of some cases of amyloidosis. Factor II and thrombin are the same thing. Factor XI is part of the intrinsic pathway would not cause prolongation PT.

Case #5 A 45 y/o female presents with a "pancreatic mass" on CT scan. An ulcerated mass was found in the second portion of the duodenum. Multiple biopsies were taken, and representative sections and special stains are shown. Based on the findings, what is the best diagnosis?

A. Undifferentiated carcinoma
B. Classical Hodgkin Lymphoma
C. Diffuse Large B cell Lymphoma
D. Anaplastic Large Cell Lymphoma
E. None of the above

Answer: D. This case represents anaplastic large cell lymphoma (ALCL) with an unusual presentation in the small bowel. In this case there is CD3 positivity, which is actually one of the least common markers of T-cell lineage found in ALCL. Most importantly, CD30 is positive and CD20 is negative. Cytotoxic markers (not shown for this case) are also helpful (TIA-1, Granzyme, and Perforin), especially if the ALK-1 is negative.

Case #6 A 60 y/o old male with a significant past medical history presents with worsening pancytopenia. A bone marrow biopsy was performed and representative sections, including special stains are shown. Based on the findings, what is the best diagnosis?

A. Hairy Cell Leukemia
B. Diffuse Large B-Cell Lymphoma
C. T-Cell Leukemia/Lymphoma Unspecified
D. AML-M4
E. None of the above

Answer: A. This case represents marrow involvement by hairy cell leukemia (HCL). It is characterized by CD20 and TRAP positivity (also DBA44 – limited availability of the antibody now). By flow cytometry it has the characteristic immunophenotype of CD11c, CD25, and CD103 positivity.

Case #7 A 48 y/o female presents with bilateral pulmonary nodules and hilar lymphadenopathy. A wedge resection is performed and representative images are shown. Based on the morphology and special stains, what is the best diagnosis?

A. Multi-centric Tuberculosis
B. Anaplastic Large Cell Lymphoma
C. Classical Hodgkin Lymphoma
D. Diffuse Large B-Cell Lymphoma
E. None of the above

Answer: C. This is an example of nodular sclerosing Hodgkin lymphoma involving the lung parenchyma. Classical Hodgkin lymphoma is characteristically positive for CD30 and CD15 and negative for CD45. In contrast nodular lymphocyte predominate Hodgkin lymphoma is CD45 and CD20 positive and negative for CD15 and CD30.

Question #2 If properly stored tissue can often be held up to 48 hours before being processed, and 72 hours before being analyzed. Which of the following specimens should be processed and analyzed within 12 to 24 hours?

A. Whole blood, acute leukemia
B. FNA, follicular lymphoma
C. Solid tissue, Hodgkin lymphoma
D. Solid tissue, Burkitt lymphoma
E. None of the above

Answer: D. Burkitt's lymphoma has a high turnover-proliferation rate. Specimens with a high turnover rate should not be delayed in processing. {Reichard, K., *Practical Approach to Bone Marrow Examination;* 2005 CAP Meeting}

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)

