

# PathMD™: Board Review Letter

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Hematopathology - Part 2

Volume 1, Number 35

Case #1 A 50 y/o male presents with nasal obstruction. A biopsy is performed and representative sections are shown. The cells of interest are positive for CD56 and EBV by in situ hybridization (EBER). Based on these findings, what is the best diagnosis?

Answer: D. Extranodal NK/T-cell lymphoma is characterized by an angiocentric infiltrate that is EBV and CD56 positive and is associated with extensive necrosis. Wegner's granulomatosis can be a difficult differential diagnosis, especially if necrosis is the predominate morphology without the characteristic perivascular infiltrate of NK cell lymphoma. (WHO, p. 204-207)

Case #2 A cervical lymph node is biopsied from a 25 y/o man. Representative histology from the lymph node is shown. Based on the findings, what is the expected immunohistochemistry profile?

Answer: D. This is a case of nodular sclerosing classical Hodgkin's lymphoma. Classical Hodgkin's lymphoma has an immunophenotype of CD15 +, CD 30 +, CD45 =, and is usually CD20 =. Nodular lymphocyte predominate Hodgkin's lymphoma (NLPHL) has an immunophenotype of CD20 + and CD45 +, but is negative for CD15 and CD30. Interestingly, NLPHL cells are typically rimmed by T-cells that are often positive for CD56. (WHO, p. 239-252)

Case #3 A 40 y/o man presents with isolated inguinal lymphadenopathy. An excisional biopsy is performed. Representative sections and special stains are shown. Based on the findings, what is the best diagnosis?

Answer: B. This is an example of reactive follicular hyperplasia. This is one of the few situations where bcl-2 immunohistochemistry is helpful in the differential diagnosis of lymphoma. Bcl-2 is normally negative in reactive germinal center (bcl-2 is an anti-apoptotic protein), but will be positive in follicular lymphoma, which is the usual differential diagnosis of reactive lymphadenopathy.

Case #4 A soft tissue mass is found in the breast of a 45 y/o female that was thought to probably represent breast carcinoma. Representative images are shown. The histiocytic cells are positive for S-100 and negative for CD1a. Based on the findings, what is the best diagnosis?

Answer: D. Rosai-Dorfman disease is characterized by histiocytes with emperipolesis, are S-100 positive, and have conspicuous plasma cells that are usually perivascular in location. Rosai-Dorfman disease is not limited to lymph nodes and has been described in most tissues.

Case #5 Based on the morphologic findings of the peripheral blood smear for this question, which of the following is the best diagnosis?

Answer: B. This case illustrates hemoglobin SC disease. It is characterized by target cells and variably shaped erythrocytes with condensed hemoglobin. The image for this case highlights the target cells and a single cell in the center with variably condensed hemoglobin.

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**Case #6** A 50-year-old male presents with mild splenomegaly and circulating atypical lymphocytes. Flow cytometry identifies a CD20 positive, CD5 and CD10 negative kappa restricted lymphoid population. Based on the morphologic and flow cytometry findings, which of the following is the best diagnosis?

Answer: B. This is an example of splenic marginal zone lymphoma with circulating villous lymphocytes. The image for this case illustrates the polar projections associated with the circulating villous lymphocytes. CLL and mantle cell lymphoma are CD5 positive lymphomas, and follicular lymphoma is a CD10 positive lymphoma.

**Question #1** Which of the following characteristics is the most important defining feature of AML-M0?

Answer: D. AML-M0 is an undifferentiated AML, which can only be differentiated from an ALL by flow cytometry. It is characterized by >90% myeloblasts of the non-erythroid cells, no Auer rods, and less than 3% positivity of myeloperoxidase (MPO) in the myeloblasts. It is the 3% MPO that separates a M0 from the rest of the AML-FAB categories. (Naeim, p. 198)

**Question #2** Which of the following characteristics is the most important defining feature of AML-M1?

Answer: A. AML-M1 is the FAB classification of AMLs without maturation. Meaning that <10% of the non-erythroid cells exhibit maturation beyond the blast stage. There may be rare Auer rods, and >= 3% positivity of myeloperoxidase (MPO) in the myeloblasts. (Naeim, p. 198)

**Question #3** Which of the following characteristics is the most important defining feature of AML-M2?

Answer: A. AML-M2 is characterized by maturation of at least 10% of non-erythroid cells. This is in contrast to AML-M1, which shows maturation in < than 10% of non-erythroid cells. (Naeim, p. 198)

**Question #4** AML-M3 is characterized by which of the following?

Answer: C. AML-M3 is also known as acute promyelocytic leukemia. It is defined by the t(15;17), and is responsive to therapy with all-trans retinoic acid. This is an important diagnosis to make because it has a high long-term cure rate, but can be very deadly in the acute stage due to DIC. DIC can be induced by degranulation of the leukemic cells.

References:

*Tumors of Haematopoietic and Lymphoid Tissues.* Jaffe, ES, et al. World Health Organization Classification of Tumours. 2001.

*Pathology of the Bone Marrow.* Naeim, F. Second Edition. 1998.

**Notes for question set:<sup>1</sup>**

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