PathMD™: Board Review Letter

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1. A 50 y/o male presents with an enlarged thyroid. Due to the mass effect, a complete thyroidectomy is performed. Based on the images shown, what is the best diagnosis?
   A. Nodular hyperplasia
   B. Follicular adenoma
   C. Follicular carcinoma
   D. Follicular variant of papillary carcinoma
   E. Adenomatoid nodules

Answer: C. This case represents a minimally/angioinvasive follicular carcinoma. Note the extension through the capsule, and the focus of angioinvasion within a venule adjacent to the capsule. To be considered true vessel invasion, the veins must be either in the capsule or beyond the capsule. Vein invasion within the tumor has no known prognostic significance. (Sternberg, p. 576-578)

2. What is the fatality rate for widely invasive follicular carcinoma tumors?
   A. 3%
   B. 25%
   C. 50%
   D. 75%
   E. >90%

Answer: C. Approximately 50% of the widely invasive follicular carcinoma tumors prove to be fatal. This is compared to only ~3% of the minimally invasive tumors. Tumors confined to the thyroid have a >80% survival rate at 10 years. (Sternberg, p. 577)

3. All of the following are true with respect to follicular thyroid carcinoma EXCEPT:
   A. Higher incidence in iodine deficient areas
   B. Propensity for lymphatic invasion
   C. Represent ~5% of thyroid cancers
   D. Microinvasive follicular carcinomas have an excellent prognosis
   E. Radiation exposure is a risk factor

Answer: B. Follicular thyroid carcinomas tend to disseminate via vascular channels, and rarely have lymph node metastasis. Papillary thyroid carcinoma has more of a propensity to spread via the lymphatics, usually to adjacent lymph nodes in the neck. (Sternberg, p. 576-578)

4. Where in the normal thyroid gland are C-cells found in the highest concentration?
   A. Upper pole
   B. Middle portion
   C. Lower pole
   D. Isthmus

Answer: A. C-cells are found in higher concentration in the upper lateral portion of the thyroid gland. The reason for this is that the upper poles of the thyroid gland are formed from the fourth-fifth branchial pouches, which contains the ultimobranchial body (associated with C-cells). (Sternberg, p. 557)

5. A 60 y/o man undergoes colonoscopy, and is noted to have a "nodular" mucosal surface. Multiple biopsies are performed, and representative images and stains are shown. Based on the findings, what is the best diagnosis?
   A. CLL/SLL
   B. Mantle cell lymphoma
   C. Marginal zone lymphoma
   D. Diffuse large B-cell lymphoma
   E. Atypical reactive lymphoid hyperplasia

Answer: B. This case is an example of mantle cell lymphoma involving the colon. Mantle cell lymphoma commonly involves the GI tract, and is almost always widespread at the time of diagnosis. The characteristic immunophenotype is CD20, CD5, CD43, and bcl-1/cyclin D1 positive and CD10/CD23 negative. (Odze, p. 531-533)

6. In the images for the previous case, which of the following FISH studies would most likely be positive?
   A. t(11;14)
   B. t(14;18)
   C. t(8;14)
   D. t(2;5)

Answer: A. Mantle cell lymphoma is associated with t(11;14), which joins the cyclin D1/bcl-1 gene on chromosome 11 with the IgH chain on chromosome 14. Follicular lymphoma is associated with t(14;18) BCL-2/IgH. Burkitt lymphoma is associated with t(8;14) c-myc/IgH, and anaplastic large cell lymphoma is associated with t(2;5). Sometime c-myc may be translocated with the λ gene on chromosome 22 – t(8;22) or the κ gene on chromosome 2 – t(2;8) in Burkitt lymphoma. *Test taking strategy* The lymphoma translocations should be easy points on the board exam!!

7. A 42 y/o female presents with a 10 cm pancreatic head mass. A CT guided biopsy is performed, which was noted to be very bloody during the procedure and on touch preparation. Based on the histology and special stains, what is the best diagnosis?

A. Lymphangioma
B. Epithelioid angiosarcoma
C. Acinar cell cystadenoma
D. Pancreatic pseudocyst
E. Microcystic serous cystadenoma

Answer: E. This is a classic example of a needle biopsy from a microcystic serous cystadenoma of the pancreas. These lesions are also known as glycogen-rich (note PAS positivity) or serous cystadenoma. The gross appearance is comparable to an infantile polycystic kidney. There is a prominent vascularity to these tumors, which can sometime be confusing if the cubiodal epithelium is not evident (especially on small needle core biopsies). (Rosai, p. 1075-1077)

8. In the previous case, it can be associated with all of the following EXCEPT:

A. Von Hippel-Lindau disease
B. Multiple endocrine neoplasia
C. Ductal adenocarcinoma of the pancreas
D. Diabetes

Answer: B. Microcystic serous cystadenoma can be associated with diabetes if enough islet cells are destroyed, and has been found in pancreases with ductal adenocarcinoma elsewhere. Some cases are from individuals with von Hippel-Lindau disease. MEN is associated with endocrine component of the pancreas. (Rosai, p. 1075-1077)

9. A patient is found to have an adrenal mass. The adrenal gland is removed, and representative histologic images are shown. Based on the findings, what is the best diagnosis?

A. Adrenal cortical adenoma
B. Adrenal carcinoma
C. Myelolipoma
D. Pheochromocytoma

Answer: D. This is a case of a pheochromocytoma. In other locations in the body, they are referred to as paragangliomas. These lesions express neuroendocrine markers, and have a well-defined characteristic nested pattern (“Zellballen”).

10. The patient in the above case was found to have a gastric stromal sarcoma. In light of the new findings, what is the best clinical diagnosis for this patient?

A. Von Hippel-Lindau disease
B. Multiple endocrine neoplasia
C. Carney's triad
D. Carney's syndrome
E. None of the above

Answer: D. The combination of paraganglioma and gastric stromal sarcoma inherited in an autosomal dominant pattern is characteristic of Carney's syndrome. The association of extra-adrenal paraganglioma, gastric malignant stromal tumors, and pulmonary chondroma characterizes carney's triad. It is only rarely seen in a familial setting. Paragangliomas can be associated with von Hippel-Lindau disease and multiple endocrine neoplasia, but are not associated with gastric stromal sarcomas. (Rosai, p. 1142-1145) *Test Taking Strategy*. Syndrome/complexes like chromosomal translocations are easy points on the board exam. I recommend keeping a list of well-described characteristics of rare entities to review for the board exam. I think this strategy is very effective, as exam writers look to evaluate the depth and scope of knowledge.

References:

Rosai and Ackerman's Surgical Pathology. Rosai J, 9th Ed. 2004.

Surgical Pathology of the GI Tract, Liver, Biliary Tract, and Pancreas, Odze, R. *et al*. 1st Edition. 2004. pp.

Sternberg's Diagnostic Surgical Pathology. Mills, SE, *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)

