

Case #1 A 35 y/o HIV patient presents with an acute abdomen. At surgery the ileocecal area was perforated. Representative histologic findings are presented in the images. Based on the history and findings, the best diagnosis is:

B. CMV colitis

Answer: B. This is an example of **CMV colitis**. It most commonly occurs in individuals with an immunodeficiency, and often presents with bloody or watery diarrhea. Histologically, the classic appearance is with “owl’s eye” inclusions, which can be either intranuclear or intracytoplasmic. (Odze, p. 31-35) When CMV infects the endothelial cells, they can swell and cause ischemia. This can lead to necrosis and perforation. HSV is usually found in the rectum and perianal skin. Inclusions are not usually seen in the colon. (Sternberg, 4<sup>th</sup> Ed., p. 1497-98)

Case #2 A 45 y/o female presents to the ER with right upper quadrant pain after eating fatty meals. Ultrasound showed cholelithiasis, and a cholecystectomy was scheduled. Upon pathologic examination, a mass was found in the fundus of the gallbladder. Based on the histologic findings, the best diagnosis is:

A. Adenomyoma

Answer: A. **Gallbladder adenomyoma**. This entity represents exaggerated diverticula (usually in the fundus), which is surrounded by smooth muscle hyperplasia. The important part of this diagnosis is recognizing the entity, and not to confuse it with an adenocarcinoma. In the diffuse form of this entity it is referred to as adenomyomatosis. (Sternberg, 4<sup>th</sup> Ed., p. 1790)

Case #3 A 19 y/o female presents with embarrassing diarrhea and flatulence over the past month. The symptoms began after a recent mission trip with her church to Romania. EGD and colonoscopy were negative for any gross disease. Duodenal biopsy images are shown. Based on the history and histologic findings, which of the following stains would be most helpful in highlighting the pathology:

D. Trichrome

Answer: D. Trichrome. This case illustrates *Giardia lamblia* infection, which often presents with abdominal pain, diarrhea, and/or flatulence. It is usually recognized in tissue sections, but a trichrome stain with an iron hematoxylin counter stain makes the organisms more conspicuous. The organisms will usually group as a “school of fish” and mostly have a “falling leaf” appearance (profile image of the trophozoite). A few of the organisms may be *en face*, which reveals the classic “pear shape.” Always remember to look above the intestinal surface in duodenal biopsies. The pathology isn’t always in the tissue. (Sternberg, 4<sup>th</sup> Ed., p. 1483-84)

Case #4 A 30 y/o male presents with an elevated bilirubin. ERCP showed a “string of beads” appearance, and the liver biopsy showed changes represented by the histologic images showed. Based on the findings, the most likely diagnosis is:

B. Primary Sclerosing Cholangitis

Answer: B. **Primary Sclerosing Cholangitis (PSC)**. PSC is usually associated with inflammatory bowel disease (Ulcerative Colitis – 70% of cases), and is more common in men. It consists of destruction of the extrahepatic biliary tree over time. There is a classic “string of beads” appearance on ERCP imaging which results from focal constrictions in the bile duct. Histologically, the liver usually shows feature of chronic large bile duct obstruction. Concentric periductal fibrosis (shown in this case) is a rare and non-specific finding. In the end, the pathologist can only suggest the diagnosis. The clinician must correlate the patient’s findings with radiographic evidence to reach a diagnosis. (Sternberg, 4<sup>th</sup> Ed., p. 1674-75)

Case #5 A 40 y/o male presented with mildly elevated AST and ALT ( 3 ½ times normal) lab values. Studies for hepatitis A, B, and C were negative. ANA titer was 1:80, but no history of autoimmune disease was present. The patient was significantly overweight, and noted to have a fasting glucose level of 157. A liver biopsy was performed. Based on the history and histologic findings (trichrome stain not shown shows increased pericellular fibrosis), the best diagnosis is:

B. Non-alcoholic steatohepatitis

Answer: B. **Steatohepatitis** is defined by macrosteatosis and evidence of hepatocellular injury (hepatocyte ballooning – with or without Mallory hyaline). Acinar inflammation (usually mild and mixed) is usually present, but many believe not required for the diagnosis. Often non-alcoholic steatohepatitis patients will have a low grade ANA-titer. (LD Ferrell, 2005 CAP)

Case #6 A 45 y/o patient was found to have a mesenteric mass. Histologic images are shown. Closed examination found the spindle cell proliferation to infiltrate the outer intestinal wall and adipose tissue. Foam and inflammatory cells were scarce. S-100, CD117, and CD34 were negative. Based on these findings, the best answer is:

D. Fibromatosis

Answer: D. This is an example of **fibromatosis**. All of the possible answers are very plausible given the histologic findings, and the immunohistochemistry is very important to separate these entities. GIST should be CD117 positive and are usually CD34 positive. S-100 should be positive in a schwannoma. A smooth muscle tumor is positive for both desmin and smooth muscle actin. (Sternberg, 4<sup>th</sup> Ed., p. 1555) IMT is usually well circumscribed and most often is in children. IMT and IRF both have a more prominent inflammatory component compared to fibromatosis (intra-abdominal desmoid). (Sternberg, 4<sup>th</sup> Ed., p. 1524-25)

Case #7 A 55 y/o male undergoes a colon resection for an unrelated mass and is found to have a lesion within the wall of the colon. Based on the histological images, the best diagnosis is:

C. Ganglioneuroma

Answer: C. **Ganglioneuromas** are characterized by spindle cells and fibrillar matrix with ganglion cells present in the lamina propria. S-100 is positive. Usually, these lesions may appear like a tubular adenoma or hyperplastic polyp to the clinician. (Sternberg, 4<sup>th</sup> Ed., p. 1549-50)

Case #8 A 75 y/o male undergoes a colon resection for a mass. Representative images of the lesion are shown. The cells of interest are Cytokeratin, cyclin D1, CD5, chromogranin, synaptophysin, and S-100 negative, and are positive for CD45 and CD20. Based on these findings, the best answer is;

E. Diffuse large B-cell lymphoma

Answer: E. **DLBCL of the rectum.** Melanoma would be S-100 positive. CD45 (LCA) puts this lesion into the hematopoietic category. Mantle cell lymphoma can resemble DLBCL when there is a pleomorphic blastoid variant, but it will usually express CD43, CD5, and cyclin D1. Therefore, DLBCL is the best answer for this case.

Question #1 Mesenteric fibromatosis is associated with which of the following:

- A. Gardner Syndrome
- B. Weber-Christian Disease
- C. Familial Adenomatous Polyposis
- D. Both A and C are correct

Answer: D. Mesenteric fibromatosis (Desmoid) is associated with Gardner's syndrome and Familial Adenomatous Polyposis. Weber-Christian Disease can have findings identical to Idiopathic Retractable Mesenteritis. (Sternberg, 4<sup>th</sup> Ed., p. 1523-25)

Question #2 Which of the following is most commonly associated with primary sclerosing cholangitis?

B. Ulcerative colitis

Answer: B. Ulcerative colitis (UC) is found to be associated with 70% of cases of PSC. It is somewhat counter intuitive to think of UC being associated with a disease that is not in the colon, and is characterized by "Skip" lesions on ERCP. One would think Crohn's disease would be more commonly associated. And while it can be, it is by far not the most common (UC). (Sternberg, 4<sup>th</sup> Ed., p. 1674-75)

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

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<sup>1</sup> 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)