



DEPARTMENT OF PATHOLOGY

Case of the Week

Gastrointestinal & Liver Pathology: Low grade mucinous adenoma of appendix with peritoneal spread

Prepared by: Sam Sirotnikov (Medical student) and Brendan Belovoarac (Resident).
Michael Bannan (Attending)

January 2, 2018

History

The patient is a 70 year-old female with history of ascites who presented with worsening abdominal distension and vague abdominal pain. CT findings revealed large volume ascites and a 3.7 x 4.5 cm possible appendiceal mucocele. The patient then underwent diagnostic laparoscopy with biopsy of an abdominal wall pelvic nodule and a perihepatic nodule, along with abdominal paracentesis.

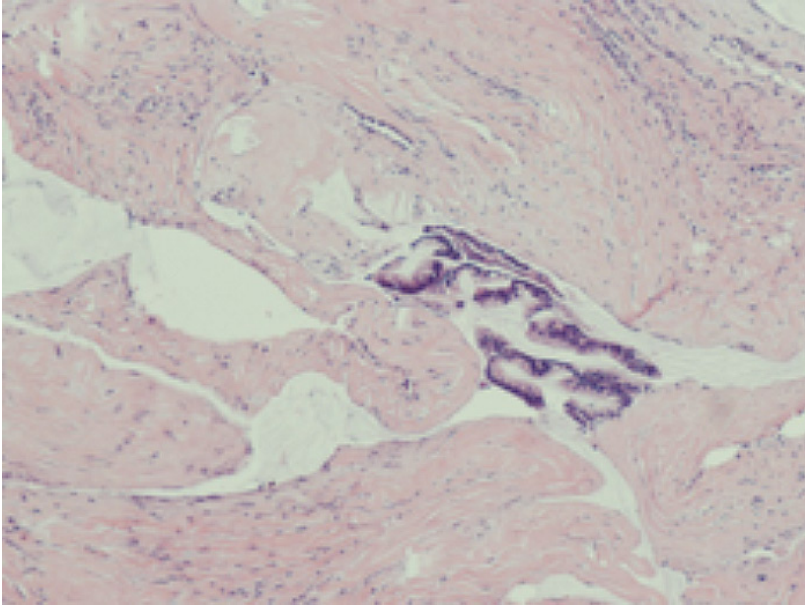


Figure 1: Scant glandular structures (tumor cells) in peritoneal biopsy (Hematoxylin and eosin; 40 x)

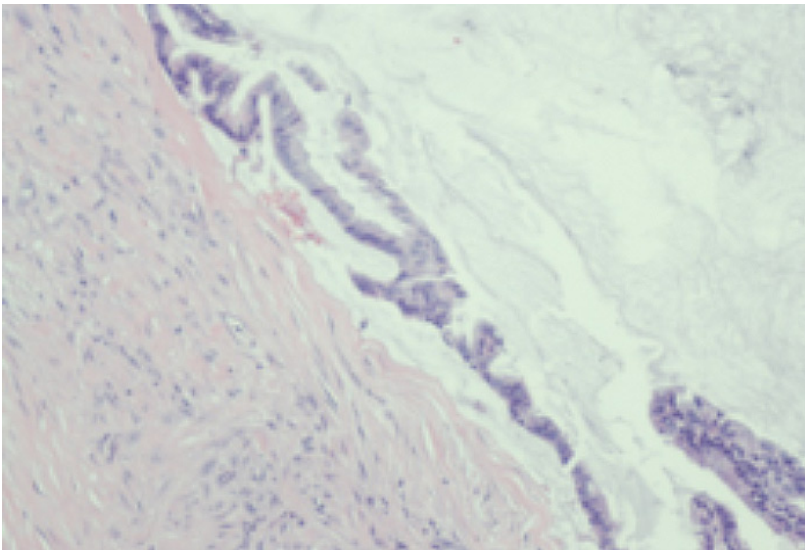


Figure 2: Abundant mucin associated with well differentiated neoplastic glands (tumor cells) in peritoneal biopsy (Hematoxylin and eosin; 100 x)

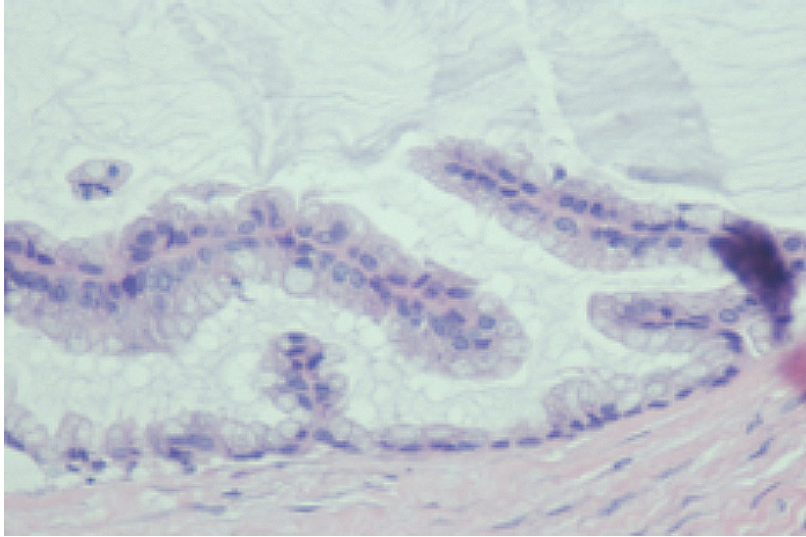


Figure 2: Well differentiated appearance of neoplastic glands with cells closely resembling benign intestinal epithelium (Hematoxylin and eosin; 200 x)

Diagnosis

Adenocarcinoma, low grade, mucin-producing (of appendix origin) with peritoneal spread (pseudomyxoma peritonei, clinical)

Discussion

Pseudomyxoma peritonei (PMP) is a clinical neoplastic presentation or condition resulting from mucinous tumor implants which produce large amounts of intra-abdominal mucinous material. The incidence is about one per million per year. Most cases arise from primary appendiceal mucinous neoplasms (low grade appendiceal mucinous neoplasms which produce a mucin filled appendix, clinically termed appendiceal mucocele whereas pathologically termed low grade appendiceal neoplasm). The tumor may less commonly be of ovarian origin, from mucinous cystadenoma or cystadenocarcinoma or rarely mucinous neoplasm arising in mature cystic teratoma. Other less common sites of origin include the colon, stomach, gallbladder, lung, breast and pancreas.

PMP or peritoneal spread of a low grade mucin producing neoplasm follows rupture of the neoplasm at site of origin (appendix) with spread of mucin-producing neoplastic epithelial cells. The cells redistribute within the peritoneal cavity and produce large amounts of viscous mucus which may lead to radiologic and clinical findings such “scalloping” of the liver and “omental caking.” Eventually, massive accumulation of mucus can lead to bowel obstruction.

The pathologic entity underlying pseudomyxoma peritonei is the spread of highly differentiated neoplastic glandular cells which produce abundant pools of mucin.

The neoplastic process underlying PMP (reference 2) is classified into three categories of adenocarcinoma: low grade, high grade, and high grade with signet ring cells. Low grade adenocarcinoma under prior classification system was termed disseminated mucinous adenomucinosis and high grade adenocarcinoma was previously termed peritoneal mucinous carcinomatosis.

Immunohistochemistry may be used to determine site of origin specifically to differentiate ovarian from appendiceal origin. SATB2, CK20, CDX2 and MUC2 will stain positively in cases of appendiceal origin, whereas PAX8 and ER may stain positively in cases of ovarian origin.

Treatment of pseudomyxoma peritonei ranges from repeated surgical debulking to more aggressive treatment involving complete cytoreduction and heated intraperitoneal chemotherapy (mitomycin and fluorouracil). Prognostic indicators include grade of the neoplasm and preoperative tumor markers (CEA, CA125 and CA19.9).

References

1. Rosai J, Ackerman LV. Rosai and Ackerman's Surgical Pathology. Elsevier. 2011.
2. Carr NJ, Cecil TD, Sugarbaker PH,. A Consensus for Classification and Pathologic Reporting of Pseudomyxoma Peritonei and Associated Appendiceal Neoplasia: The Results of the Peritoneal Surface Oncology Group International Modified Delphi Process. *Am J Surg Pathol.* 2016; 40(1):14-26.
3. Buell-Gutbrod R, Gwin K. Pathologic diagnosis, origin, and natural history of pseudomyxoma peritonei. *Am Soc Clin Oncol Educ Book.* 2013:221-5.
4. Fallis SA, Moran BJ. Management of pseudomyxoma peritonei. *J BUON.* 2015; 20(Suppl 1):S47-55.
5. Strickland S, Parra-Herran C. Immunohistochemical characterization of appendiceal mucinous neoplasms and the value of special AT-rich sequence-binding protein 2 in their distinction from primary ovarian mucinous tumours. *Histopathology.* 2016;68(7):977-87.