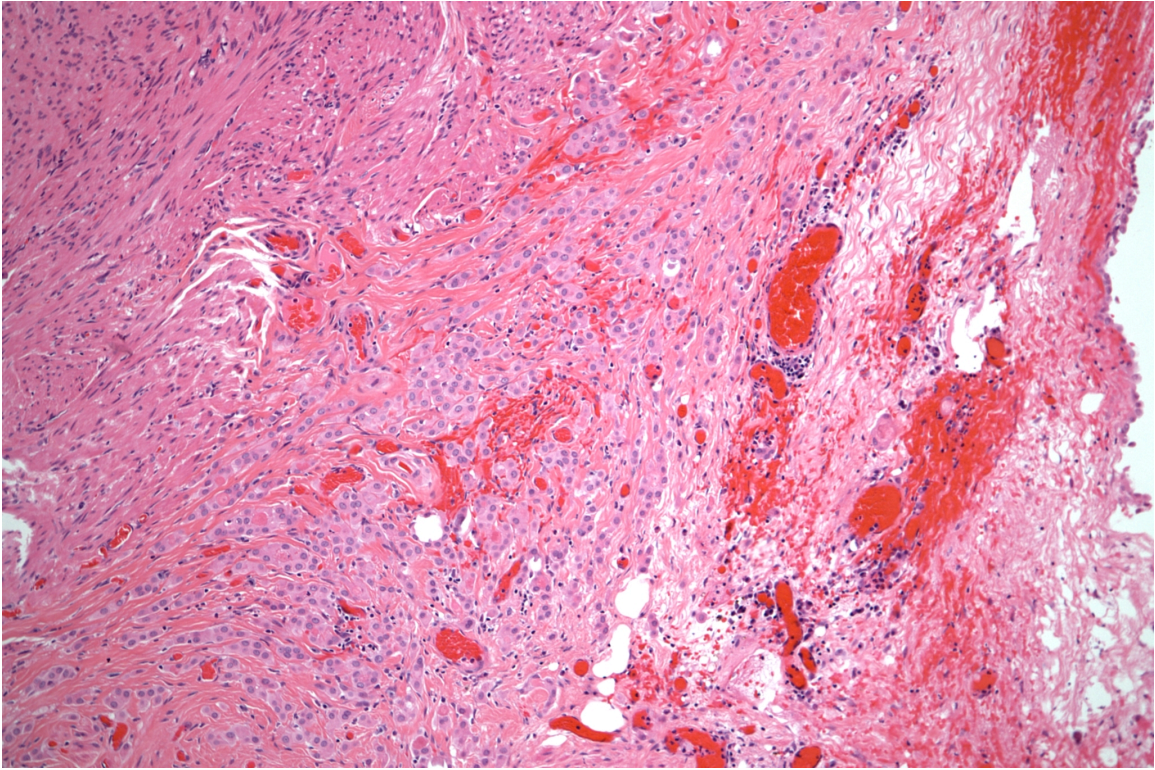
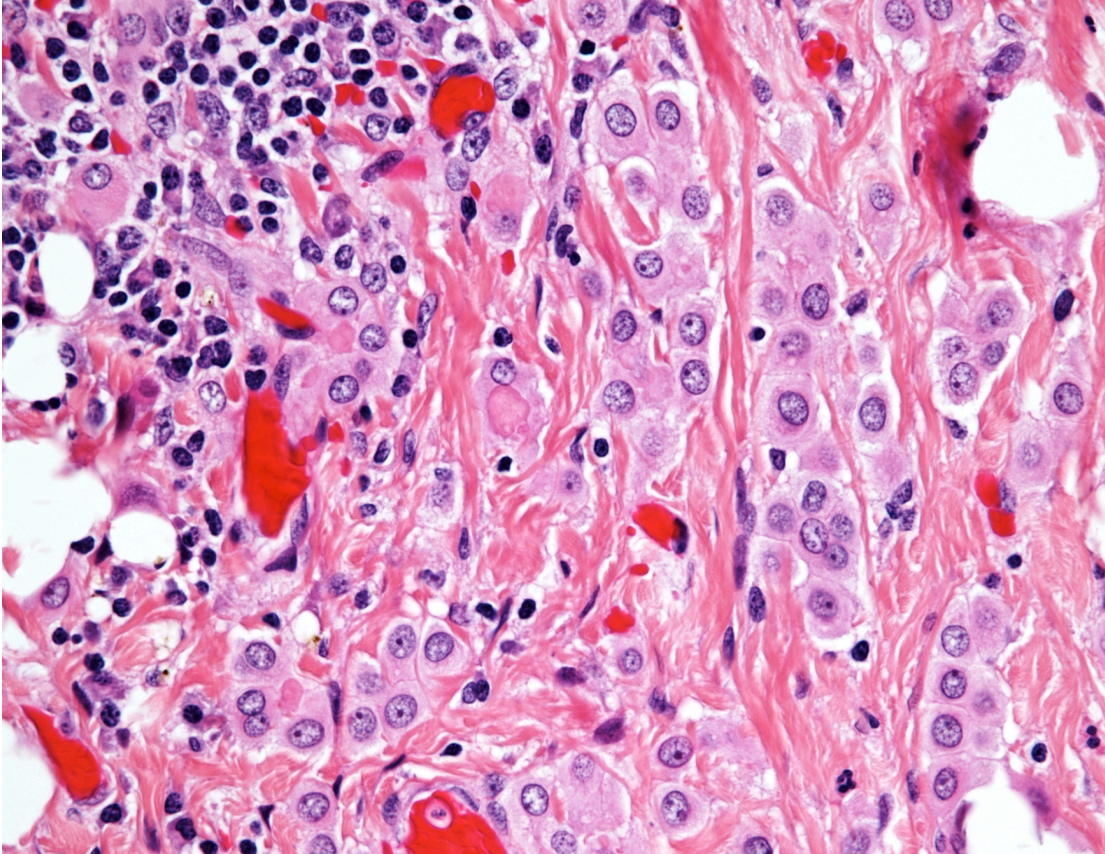
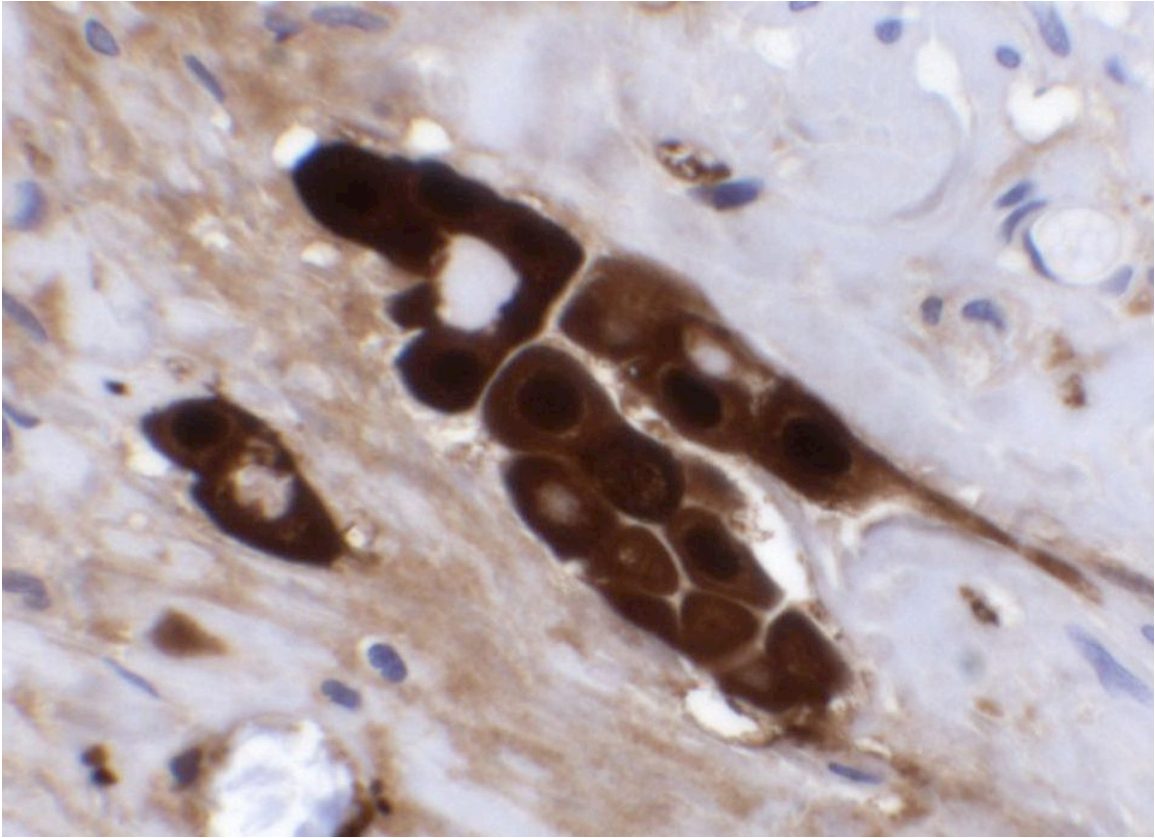


Case

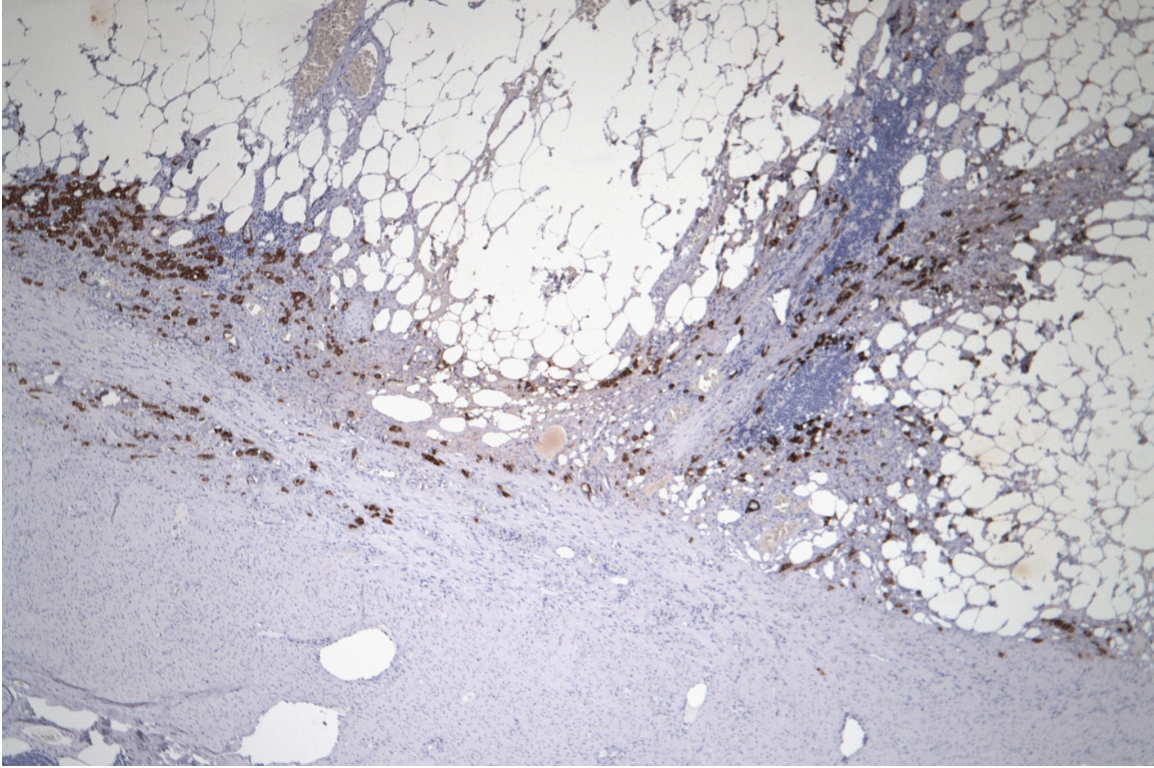
The patient is a 47-year-old male with a 3-week history of abdominal pain. A CT scan of the abdomen revealed a suggestion of wall thickening at the tip of the appendix only. An appendectomy was performed. Grossly, the appendiceal serosa was mildly erythematous. The appendix was otherwise unremarkable. Intraoperatively, no abnormalities were noted in the peritoneal cavity.







Calretinin



Cytokeratin AE1:AE3

What is your diagnosis?

- A) Neuroendocrine tumor
- B) Malignant peritoneal mesothelioma, epithelial type
- C) Metastatic carcinoma
- D) Fibrous adhesions with reactive mesothelial cell proliferation
- E) Primary peritoneal carcinoma

Answer and Discussion:

B) Malignant peritoneal mesothelioma, epithelial type

Approximately 1/10 of malignant mesotheliomas arise from the serosal lining of the peritoneum. These tumors are typically widespread at the time of diagnosis and easily identifiable grossly and radiologically as confluent nodules/plaques or diffuse serosal thickening. Occasionally, they occur as a localized mass. There are isolated reports of malignant peritoneal mesotheliomas presenting as inflammatory lesions (e.g. appendicitis, cholecystitis). In these rare cases, mesothelioma could only be seen microscopically at the time of initial evaluation. However, on follow-up, gross tumor developed.

Histologically, the morphologic spectrum is highly variable, and for this reason, mesotheliomas can mimic many different types of tumor. In general, they are divided into epithelial (most common), sarcomatous, and biphasic subtypes. By immunohistochemistry, epithelial mesotheliomas are always positive for pancytokeratin, and variably positive for any or all of CK5/6, calretinin, WT-1, D2-40, and thrombomodulin, although no marker is absolutely sensitive or specific. In this case, the tumor cells were positive for pancytokeratin, calretinin, WT-1, and D2-40, while negative for synaptophysin and chromogranin. This staining pattern argues against neuroendocrine tumor and metastatic carcinoma (choices A and C). In women, metastatic ovarian and primary peritoneal serous carcinomas (choice E) are often considered in the differential diagnosis of peritoneal mesothelioma. WT-1 is expressed in approximately 90% of serous carcinomas and the other mesothelial markers listed above can be positive in a subset of these tumors. Therefore, other immunostains must be employed. PAX8, estrogen receptor, and the carcinoma markers MOC-31 and Ber-EP4 are all typically positive in serous carcinoma and negative in mesothelioma.

Once mesothelial origin is established, it may be difficult to distinguish between malignant mesothelioma and a benign mesothelial proliferation (choice D). The presence or absence of cytologic atypia is not helpful, as malignant mesothelioma may be deceptively bland. True stromal invasion, which is best identified as invasion of fat (as in this case) is the best criterion for mesothelial malignancy. Cytokeratin immunostain can aid in appreciating the distribution of cells and separating an invasive process from entrapment of benign mesothelial cells within inflammatory tissue.

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