Case #81

NAME Educational Activities Committee
Case provided by:

Dr. Thomas J. Auen (PGY2) and Dr. Ana Yuil-Valdes (University of Nebraska Medical Center).
1. This cystic liver lesion was discovered during the autopsy of a 43-year-old man. On sectioning the lesion contained green, granular material. Based on the histology, which of the following is the most likely diagnosis?

- Polycystic liver disease
- Simple biliary cyst
- Mucinous cystic neoplasm
- Biliary cystadenocarcinoma
- Ciliated hepatic foregut cyst
Answer...
E. Ciliated hepatic foregut cyst — (CORRECT ANSWER, 33.20 % of responses)

The Masson Trichrome stain highlights blue elastic connective tissue layers, while allowing visualization of entrapped smooth muscle bands in the cystic wall. Overlying these subepithelial components, there is a ciliated layer of pseudostratified epithelium, though denuded through autolysis in the postmortem state. These histological findings are most suggestive of a ciliated hepatic foregut cyst\textsuperscript{6,7}.
A ciliated hepatic foregut cyst is a rare cystic lesion that arises from the embryonic foregut. The classic histologic pattern is comprised of 4 distinct layers: An inner ciliated epithelial lining, smooth muscle, loose connective tissue and a fibrous capsule.

Most reported ciliated hepatic foregut cysts are localized to the center of the liver, particularly in segment IV, though other sites have been noted. They tend to be 4 cm in dimension or smaller, as was the case with the illustrated lesion.
A. **Polycystic liver disease** *(1.61 % of responses)*

Polycystic liver disease is characterized by multiple unilocular cysts of variable size filled with straw-colored fluid, causing a massively enlarged liver. On histology, the cysts will be lined by cuboidal to flat biliary epithelium\(^1\). Another common finding in polycystic liver disease is the presence of von Meyenburg complexes, which were not seen in our case.

B. **Simple biliary cyst** *(45.88 % of responses)*

Simple biliary cysts are well-circumscribed, and typically unilocular, with a fluid filled cavity and an inner surface that is smooth. Histologically, the epithelium will be a cuboidal or columnar derivative of biliary tissue, and the cyst wall can contain fibrotic and hyalinized tissues along with other types of metaplasia including intestinal or squamous\(^2\).
C. Mucinous cystic neoplasm (11.87 % of responses)

Mucinous cystic neoplasms (MCNs) of the liver are typically multilocular and have a smooth lining. They can either be lined by columnar, cuboidal, or flattened biliary epithelium or by mucinous epithelial cells overlying an ovarian-type stroma with densely packed, spindle shaped cells. MCNs may contain serous, mucinous or gelatinous material, and do not communicate with the biliary tract.

D. Biliary Cystadenocarcinoma (7.44 % of responses)

Biliary cystadenocarcinoma is identified by a proliferating malignant epithelium (often multilayered), with frequent mitotic figures, loss of polarity and nuclear pleiomorphism. The presence of mesenchymal and/or ovarian stroma is often implicated in cystadenocarcinoma prognosis, with better outcomes in those with mesenchymal associations. There is overlap with biliary cystadenomas, the benign variant of similar histology, which demonstrates three distinct layers. Specifically, a mucin producing epithelium, a layer of undifferentiated mesenchymal cells, and a dense layer of collagenous connective tissue.
REFERENCES


