Case #81

NAME Educational Activities Committee

Case provided by:

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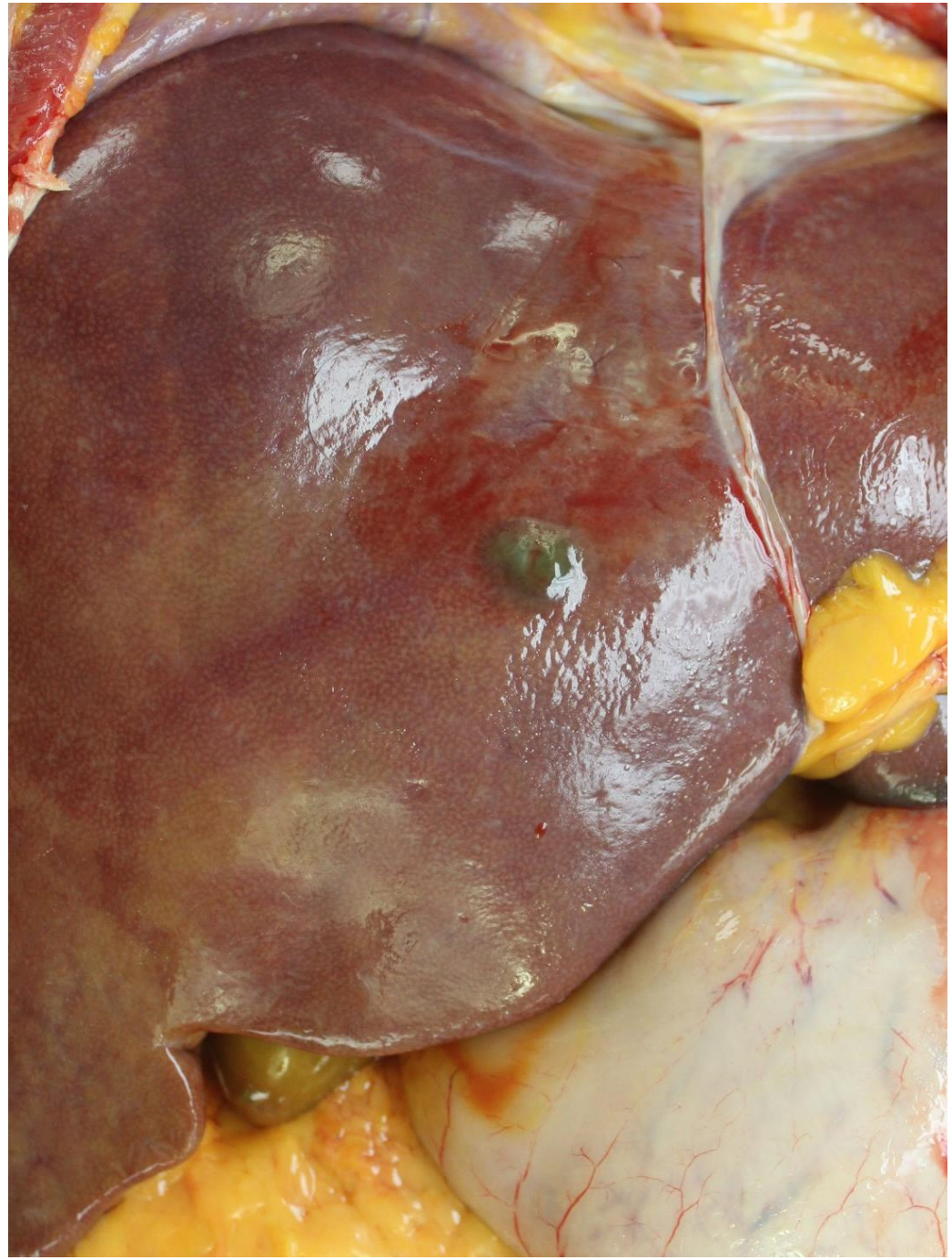

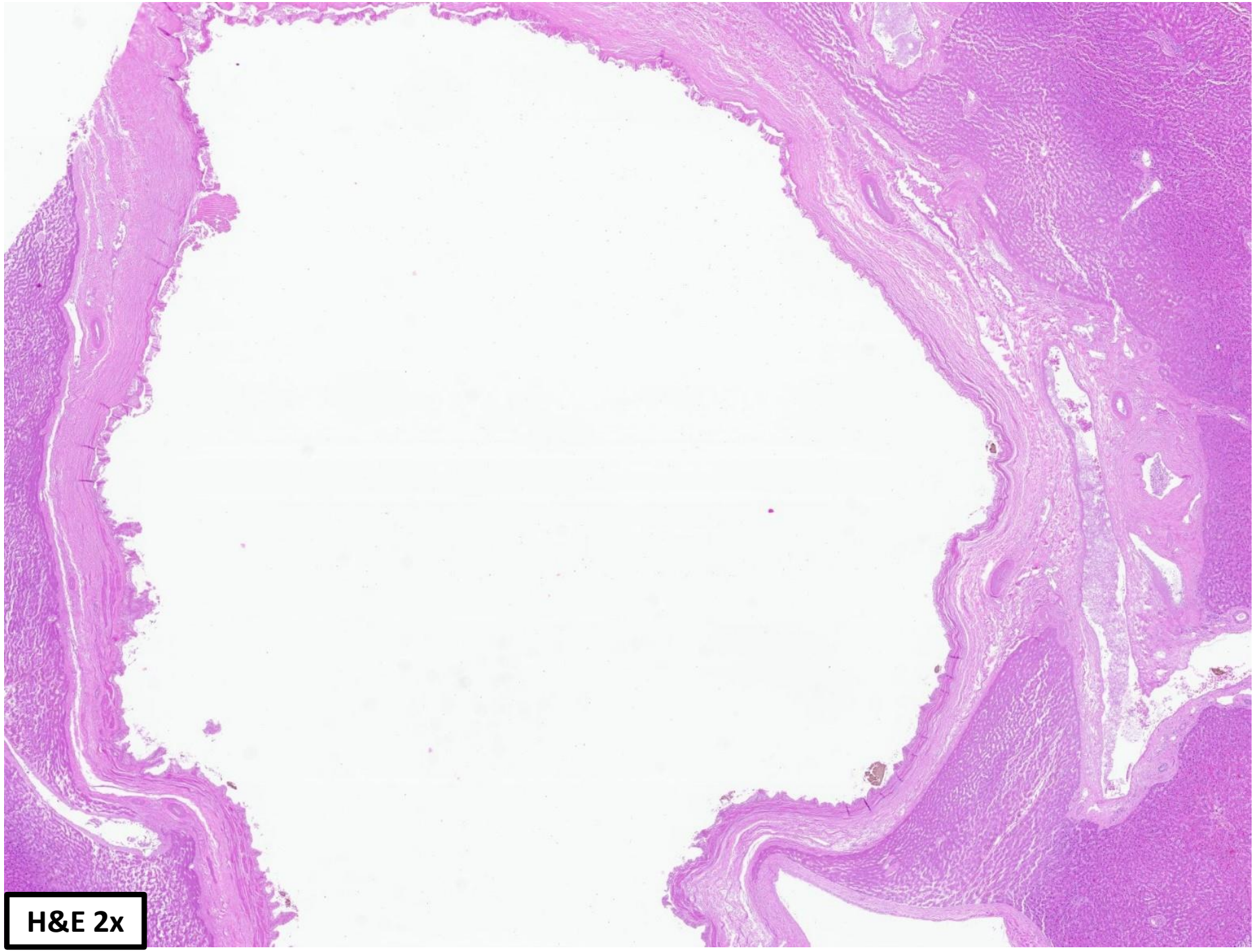
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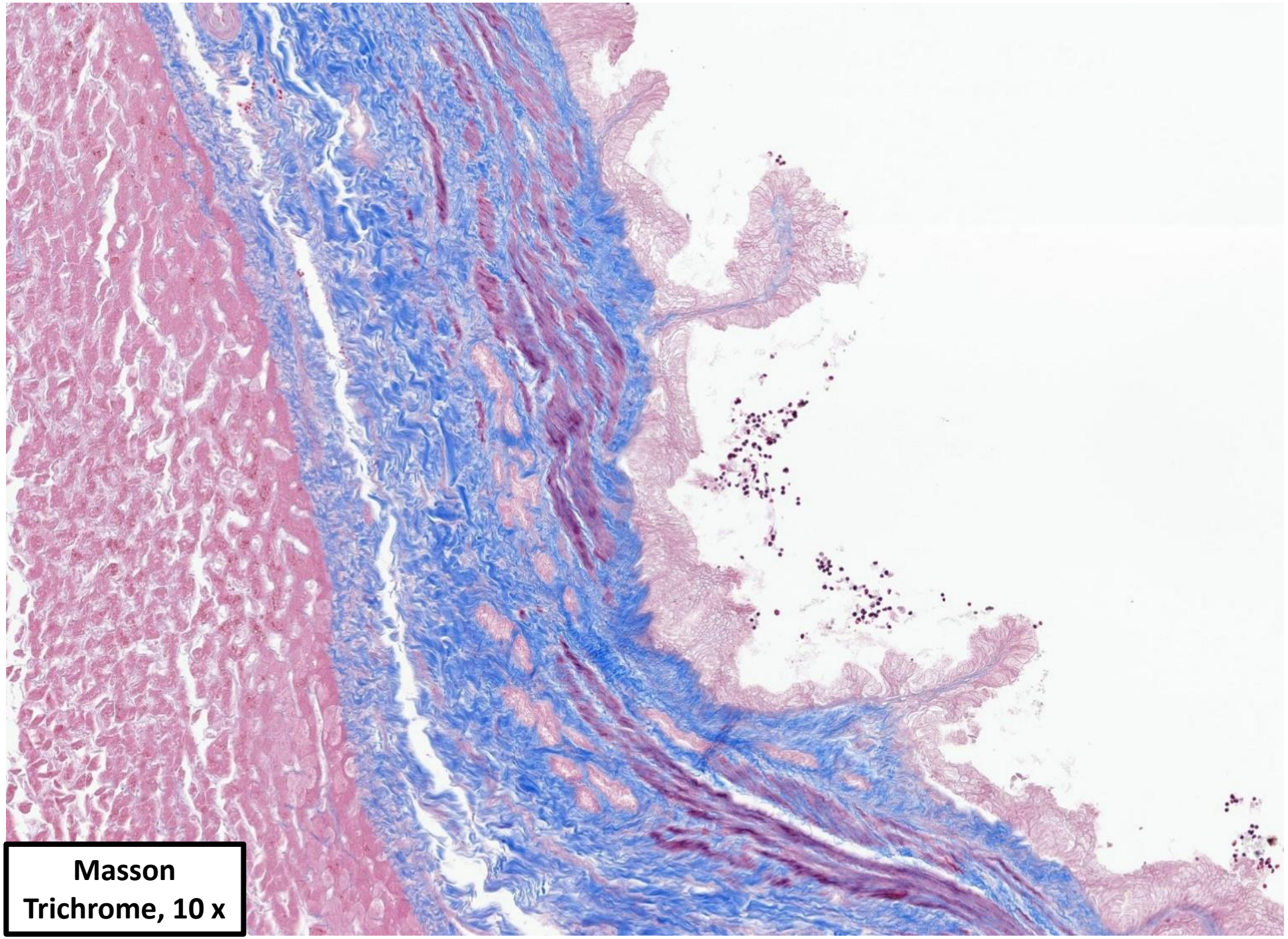
Masson
Trichrome, 10 x

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[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[8].

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[3]. 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[4]. 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[5].

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