

Slide Seminar 5, HEPATOBILIARY AND PANCREATIC PATHOLOGY

CASE 8

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Clinical history

A 60-year-old woman symptomatically presented with abdominal discomfort. Abdominal US revealed a large cystic mass, measuring 120 mm in diameter, in the liver. Radiologically, the tumour showed a multiloculated cystic appearance with septation. Solid mural nodules were also identified. CA19-9 and CEA were within normal range. The tumour was surgically resected.

Macroscopic features

The cystic lesion was multilocular and surrounded with fibrous cyst wall. Slicing revealed some mural small cysts. Some papillary mural nodules were also identified. Luminal surface was slightly rough and focally haemorrhagic.

Histological features

Most parts of the cyst are lined by a minimally atypical biliary-type epithelium with occasional mucin-containing cells. The epithelium was supported by fibrous connective tissue in which ovarian-like stroma consisting of cellular spindle cells was identifiable. Mural nodules consisted of dysplastic epithelium arranged in tubule-papillary architecture. Dysplastic cells showed enlarged nuclei, an increased nuclear / cytoplasmic ration, and distorted cellular polarity. Mitotic figures were also identified. Invasive growth was not seen. On immunostaining spindle cells in ovarian-like stroma were positive for estrogen receptor.

Diagnosis

Mucinous cystic neoplasm with high-grade dysplasia of the liver

Discussion

Mucinous cystic neoplasms (MCNs) of the hepatobiliary system, also called biliary cystic neoplasms, are rare tumours usually developing within the liver.¹ The most recent WHO

classification defined the hepatic MCNs as a cyst-forming epithelial neoplasm, usually showing no communication with the bile ducts, composed of cuboidal to columnar, variably mucin-producing epithelium, associated with ovarian-type subepithelial stroma.² Most patients with MCN are female and range widely in age. Most hepatic MCNs were located in the left lobe, in particular 54% in segment IV. Multilocular cysts with septation or a cyst-in-cyst appearance are distinctive. Based on the most recent study which analyzed 29 patients with hepatic MCN,³ 26 patients (90%) are histologically benign and only 2 (7%) were borderline malignant, and 1 (3%) was a carcinoma in situ. Invasive cancer was not recorded in that study. Although local recurrence rarely occurs, at times years after surgery, the prognosis is excellent.

MCNs need to be distinguished from other cystic liver lesions including simple cysts, cystic hamartomas, and other cystic tumours including intraductal papillary neoplasm of the bile duct (IPNB).^{4,5} The patients with MCN are significantly younger than the patients with IPNB. MCNs are significantly larger than IPNBs (median diameter: 110 mm vs. 50 mm, $p=0.008$). In contrast to IPNBs which are usually histologically malignant, 90% of MCNs (90%) are adenomas. Benign MCNs have the pure biliary immunophenotype CK7+/CK20-/MUC2-/MUC5AC-/MUC6-, whereas gastrointestinal markers (CK20, MUC2, MUC5AC, and MUC6) are more frequently expressed in IPNBs and borderline or malignant cases of MCNs.

Hepatic and pancreatic MCNs share most clinicopathological features. However, there are several pathological differences, the most significant one regarding the proportion of malignant cases. Based on the largest study on pancreatic MCNs,⁶ out of 163 patients with pancreatic MCNs, 118 (72%) were adenomas, 17 borderline neoplasms (10.5%), 9 carcinomas in situ (5.5%), and 19 were invasive carcinomas (12%). There are also some differences between hepatic and pancreatic MCNs regarding the immunophenotype of the lining epithelium.^{7,8}

References

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