Case 1: 35-year-old male with clinical signs of portal hypertension and imaging suggestive of cirrhosis
Answer: ***Congenital hepatic fibrosis***

Q1: Dx?
Q2: What is the larger family of disease?

Autosomal recessive disorder in the fibropolycystic diseases family (like polycystic kidney disease) impacting liver and kidneys.

Malformation of ductal plate (embryologic precursor to biliary system)→ don't remodel the way they are supposed to → large, dilated, irregularly-shaped ducts.

Usually present with symptoms of portal hypertension, but can have biliary symptoms.
Treatment: Largely supportive

Case 2: 60-year-old man with jaundice
Answer: ***Clonorchis sinensis infection***

Q3: Dx?
Q4: How acquired?
Q5: Most feared complication?

Helminths. A type of liver fluke endemic mostly to southeast Asia and acquired through eating raw or undercooked fish/crayfish.

Occlude bile duct→ dilated ducts with wall thickening→ Signs of biliary obstruction (jaundice, fever, RUQ pain) → can cause cholangiocarcinoma long-term due to chronic inflammation

Case 3: 45-year-old woman with a liver mass
Answer: ***Angiomyolipoma (PEComa)***

Let ask for stains!!!!!!

Q6: Dx?
Q7: How do these behave?

Benign tumors, just like in the kidney! Think of this if you see fat.

Variable admixture of fat, smooth muscle, and thick-walled blood vessels. Associated with tuberous sclerosus. Usu. Asymptomatic. Stain with HMB45 and CathepsinK. MelanA+/-

Case 4: 55-year-old man with pancreatic and kidney lesions
Answer: ***Clear cell well-differentiated neuroendocrine tumor (and Serous cystadenoma, like has VHL).***

Stains?

Q8: Dx for large tumor?
Q9: What clinical syndrome is likely at play?

Clear cell well-differentiated neuroendocrine tumors behave like other GI WD-NET. Mainly important to recognize so 1) Don't diagnose as something else, and 2) associated with Von-Hippel Lindau syndrome.

Serous cystadenoma: Benign. Often identified incidentally. Composed of bland, uniform, cuboidal cells with clear, glycogen-rich cytoplasm. Cysts lined by a single layer of cells, with well-defined cell borders. Small, round nuclei. Glycogen→ stains with PAS (and digested by diastase).
Characteristic multilocular, sponge-like appearance with a central scar (think of a cut orange!)
Also, associated with von Hippel-Lindau syndrome (VHL) (can get multiple).

Case 5: 60-year-old man with weight loss, liver mass found on imaging.
Answer: ***Combined hepatocellular carcinoma-Cholangiocarcinoma***

Stains?
Q10: Dx?
Q11: Outcome/Treatment?

A single tumor with morphologically distinct areas of HCC (Arginase and Hepar +) and Cholangiocarcinoma (CK7+).

Treated and prognosis similar to cholangiocarcinoma (Worse than HCC, No transplantation).