



DEPARTMENT OF PATHOLOGY

Short Report in Pathology

Organ system: Ampullary

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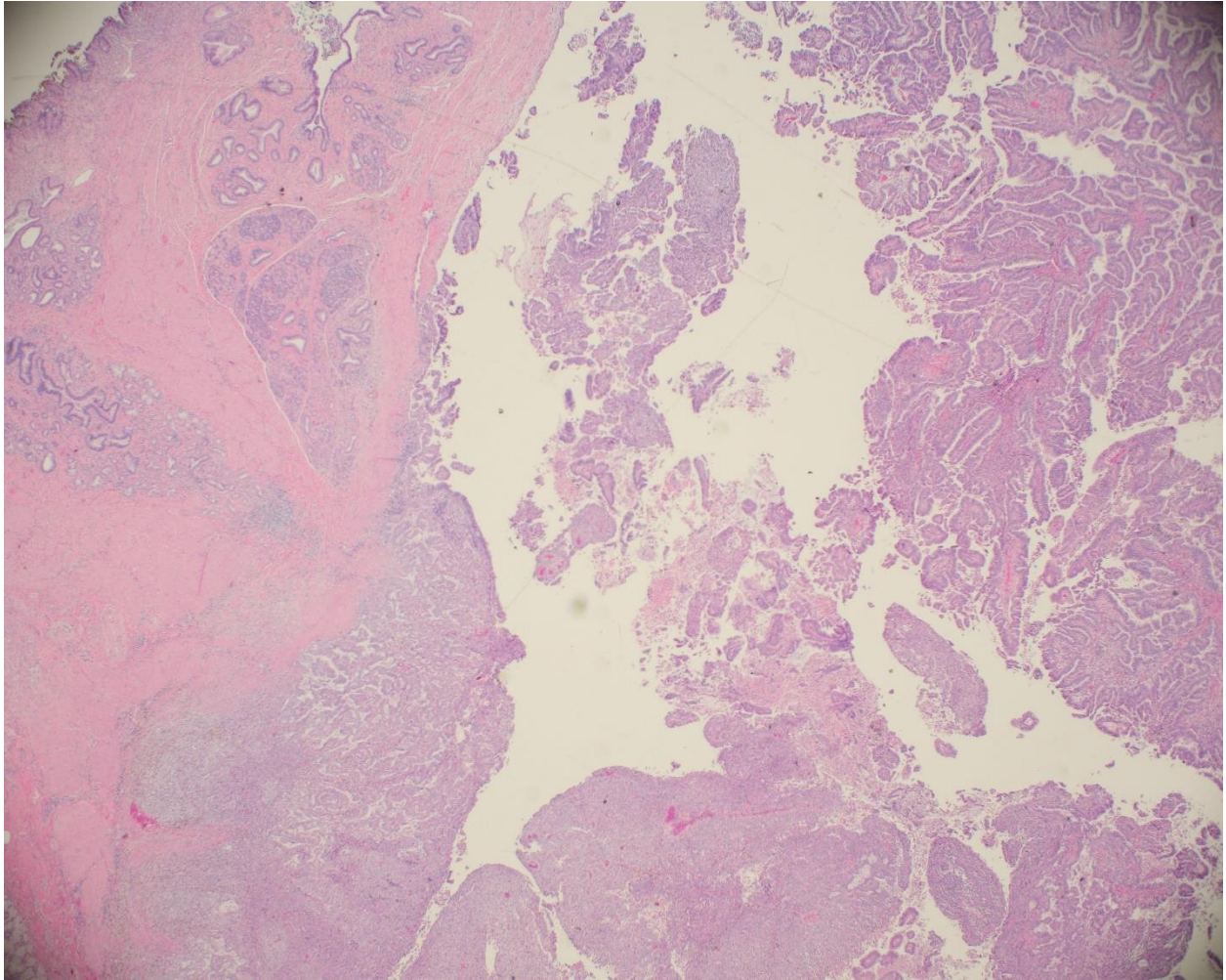
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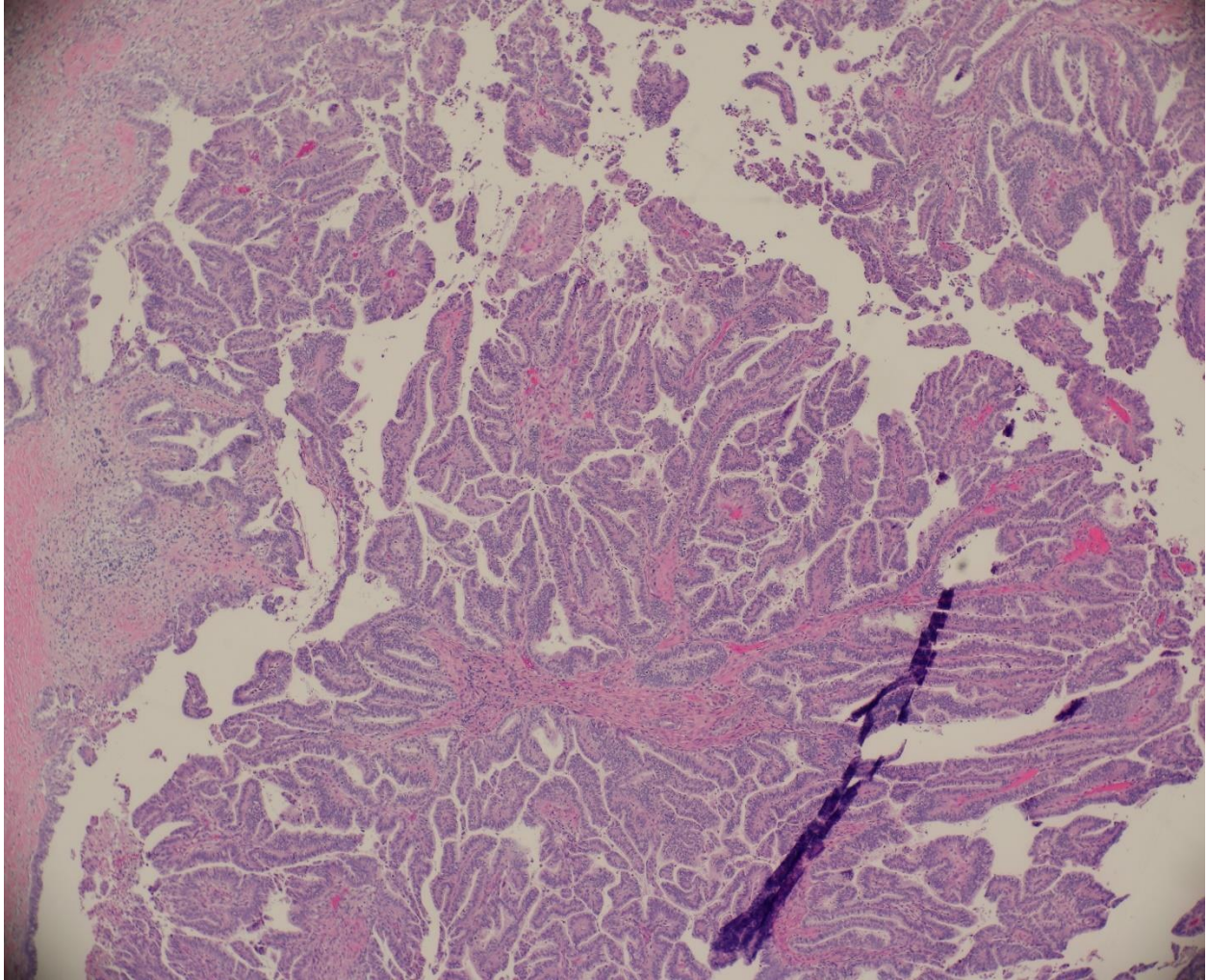
History:

A 57-year-old man presented with abdominal pain and unintended weight loss for the last 6 months. Recently, his family members also noted that he had jaundice in his eyes. He has no significant family history. An MRI of the abdomen with MRCP showed filling defects in the distal common bile duct (CBD). Esophagogastroduodenoscopy (EGD) examination showed a very prominent major ampullar area with focal ulceration. A Whipple procedure (pancreaticoduodenectomy) was done.

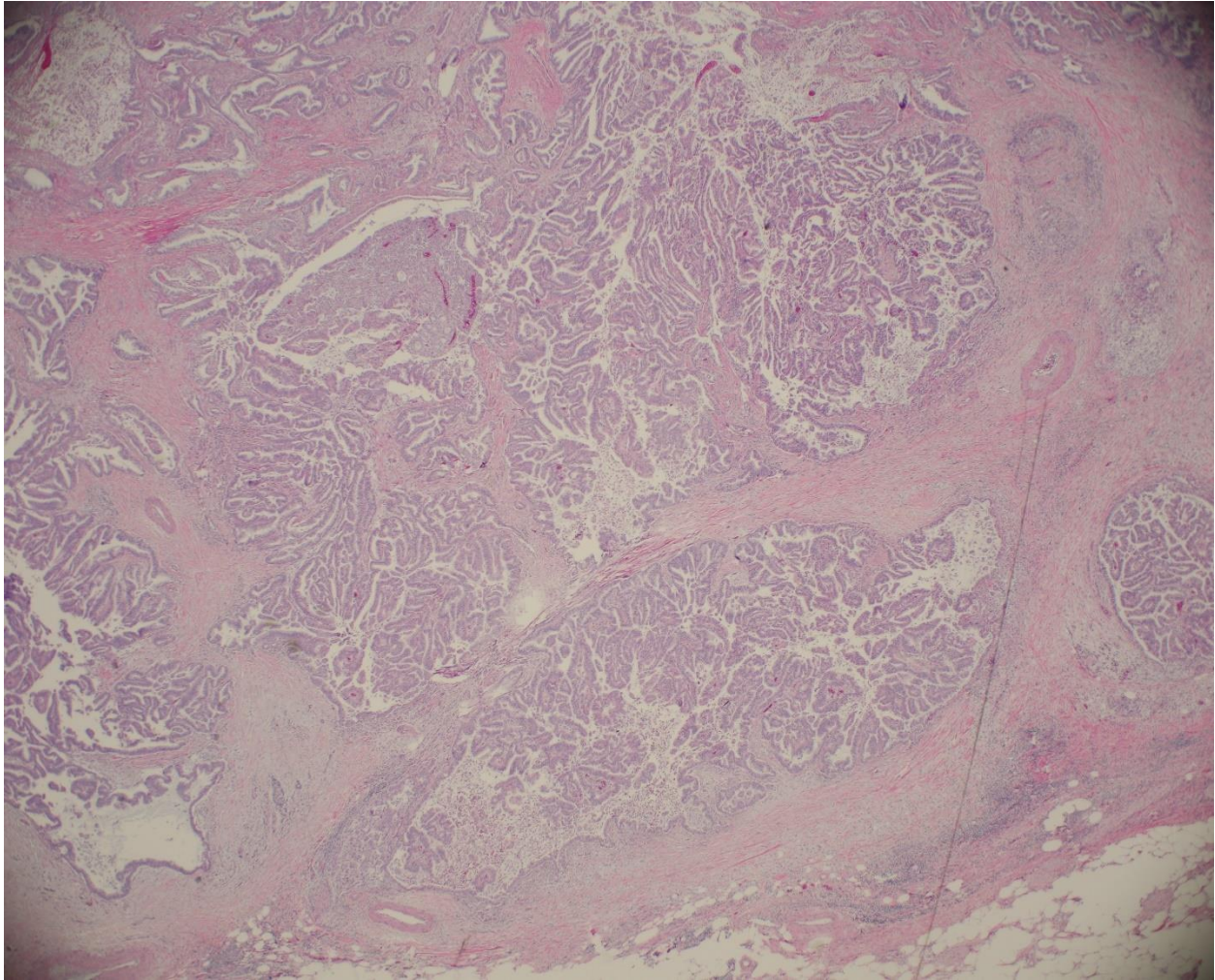
Microscopic Images:



Low magnification image showing an exophytic tumor in the distal ampullary channel in the major ampulla area causing dilatation and obstruction. The tumor shows predominantly papillary architecture with focal tubular growth pattern. The lining epithelium shows predominantly pancreatobiliary phenotype. (H&E, 10X)



Medium magnification image. The tumor shows predominantly papillary growth pattern and is lined predominantly by pancreatobiliary epithelium with low- and high-grade dysplasia. (H&E, 20X)



At the base of the tumor, there are focal areas with infiltrative growth and desmoplastic stroma, suggestive of focal invasion. The adenocarcinoma invades beyond the sphincter of Oddi and into the submucosae (H&E, 10X)

Immunohistochemistry:

Neoplastic epithelium co-express CK7 and CK20.

Immunostaining for MUC2 and CDX2 is positive in the areas with intestinal differentiation.

MUC1, MUC5AC, and MUC6 are focally positive in the areas with gastric and pancreatobiliary differentiation.

Differential diagnoses:

1. Intraductal papillary mucinous neoplasm (IPMN)
2. Intraductal tubular papillary neoplasm (ITPN)
3. Duodenal/periampullary adenoma

Diagnosis:

Invasive ampullary adenocarcinoma arising from intraampullary papillary-tubular neoplasm (IAPN).

Discussion:

Intra-ampullary papillary-tubular neoplasm (IAPN) is a relatively rare tumor that originates in the ampulla of Vater, where the common bile duct (CBD) and the main pancreatic duct (PD) merge and enter the duodenum. IAPN is considered a distinct entity among the various types of benign and malignant tumors that can occur in this complex region. IAPN primarily affects middle-aged to elderly patients and occurs predominantly in men. While the exact cause of IAPN remains unclear, there are certain risk factors including chronic inflammation, biliary stones, and certain genetic abnormalities. Clinical presentation of IAPN may vary, but common symptoms include abdominal pain, jaundice, weight loss, and changes in bowel habits.

Grossly, IAPN typically shows elevation or ulceration of ampullary region, often with a patulous papilla orifice and nodular/granular mucosa. Usually there is no overt mucinous secretion as seen in its pancreatic counterpart IPMN. On section, there usually is an exophytic tumor causing dilatation in the ampullary channel causing dilatation. Microscopically, IAPN typically demonstrates mixed papillary or tubular growth pattern, and exhibit a mixture of low- and high-grade dysplasia. The majority of IAPNs show mixed (intestinal, gastric, pancreatobiliary) differentiation. Approximately 75% of IAPNs are associated with invasive carcinoma at the time of diagnosis, but the invasive component is usually less than 1 cm in diameter.

Differentiating IAPNs from other benign or malignant tumors occurring in the ampullary region (such as duodenal adenoma, pancreatic ductal adenocarcinoma, duodenal adenocarcinoma etc.) has important therapeutic implications. It requires careful gross and histologic examination. A non-intestinal phenotype is helpful in distinguishing these lesions from duodenal adenomas because the duodenal adenoma shows mostly intestinal differentiation.

Noninvasive IAPNs have an excellent prognosis, but long-term follow-up is still highly recommended. Invasive adenocarcinoma arising from IAPN is associated with better prognosis than conventional (invasive) ampullary carcinoma not arising from IAPN, which is most likely due to early diagnosis.

References:

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2. Ohike N, Kim GE, Tajiri T, Krasinskas A, Basturk O, Coban I, Bandyopadhyay S, Morohoshi T, Goodman M, Kooby DA, Sarmiento JM, Adsay NV. Intra-ampullary papillary-tubular neoplasm (IAPN): characterization of tumoral intraepithelial neoplasia occurring within the ampulla: a clinicopathologic analysis of 82 cases. *Am J Surg Pathol*. 2010 Dec;34(12):1731-48. doi: 10.1097/PAS.0b013e3181f8ff05.