

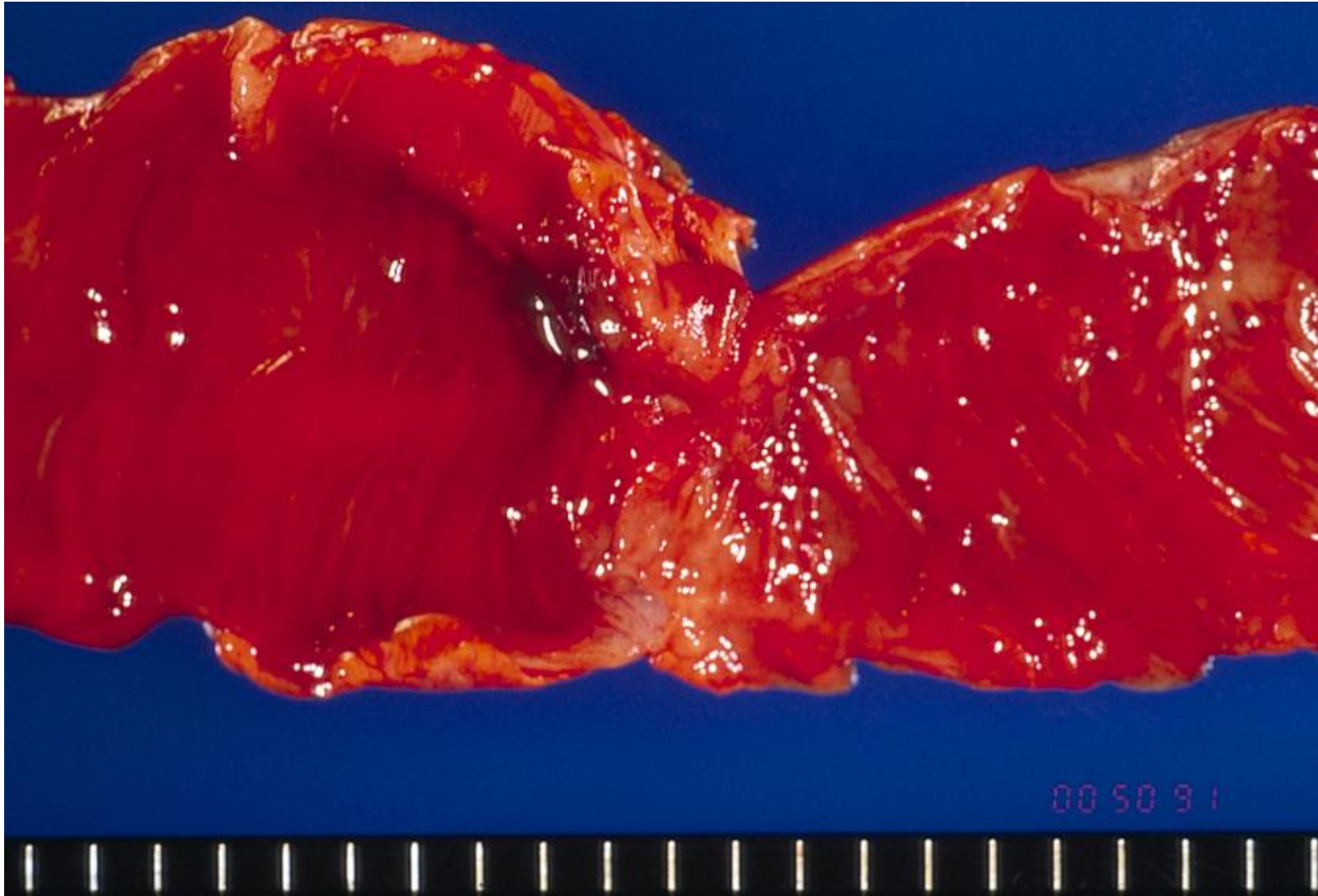
Carcinoid syndrome due to ileal carcinoid tumor

Carcinoid syndrome is a paraneoplastic syndrome caused by liver-metastasizing serotonin-secreting neuroendocrine tumor (formerly known as carcinoid tumor). In most cases, the primary site of the neuroendocrine tumor is the mid-gut zone (ileum and vermiform appendix). It should be noted that the carcinoid tumors of mid-gut origin have a higher potential of metastasis. Liver metastasis allows the elevation of serum serotonin (5-HT) levels, causing symptoms such as flushing and diarrhea. Less frequently, heart failure, vomiting, bronchoconstriction and retroperitoneal fibrosis-related hydronephrosis are associated. In the heart, right-sided tricuspid and pulmonic valves reveal fibrotic thickening to provoke murmur as carcinoid heart disease. Of note is that palpation of the enlarged liver with metastatic lesions may induce flushing episodes, as was so in the present case.

Ref.: Gade AK, et al. Carcinoid syndrome: a review. Cureus 2020; 12(3): e7186. doi: 10.7759/cureus.7186



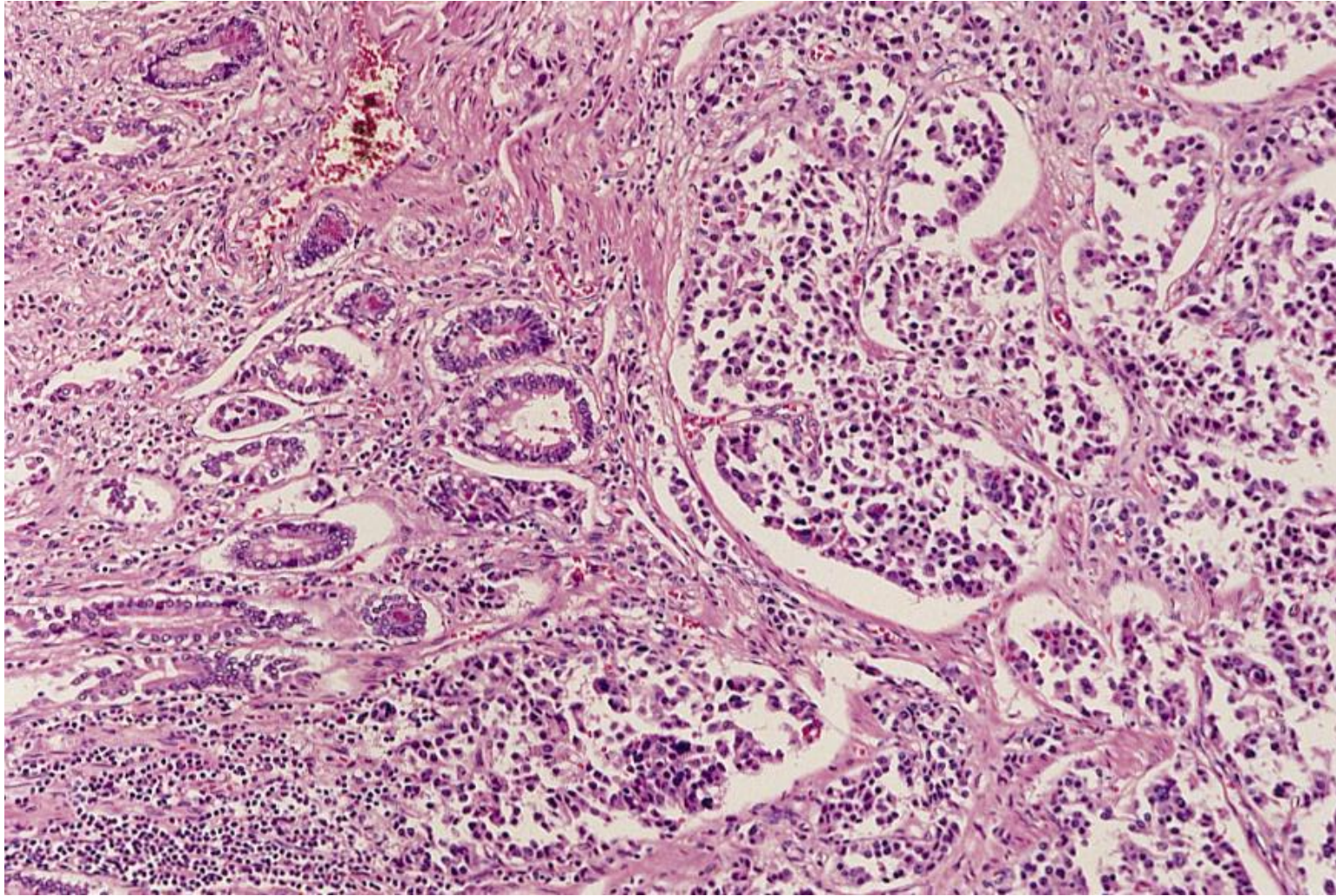
Gross appearance of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. A stenotic tumorous lesion is seen.



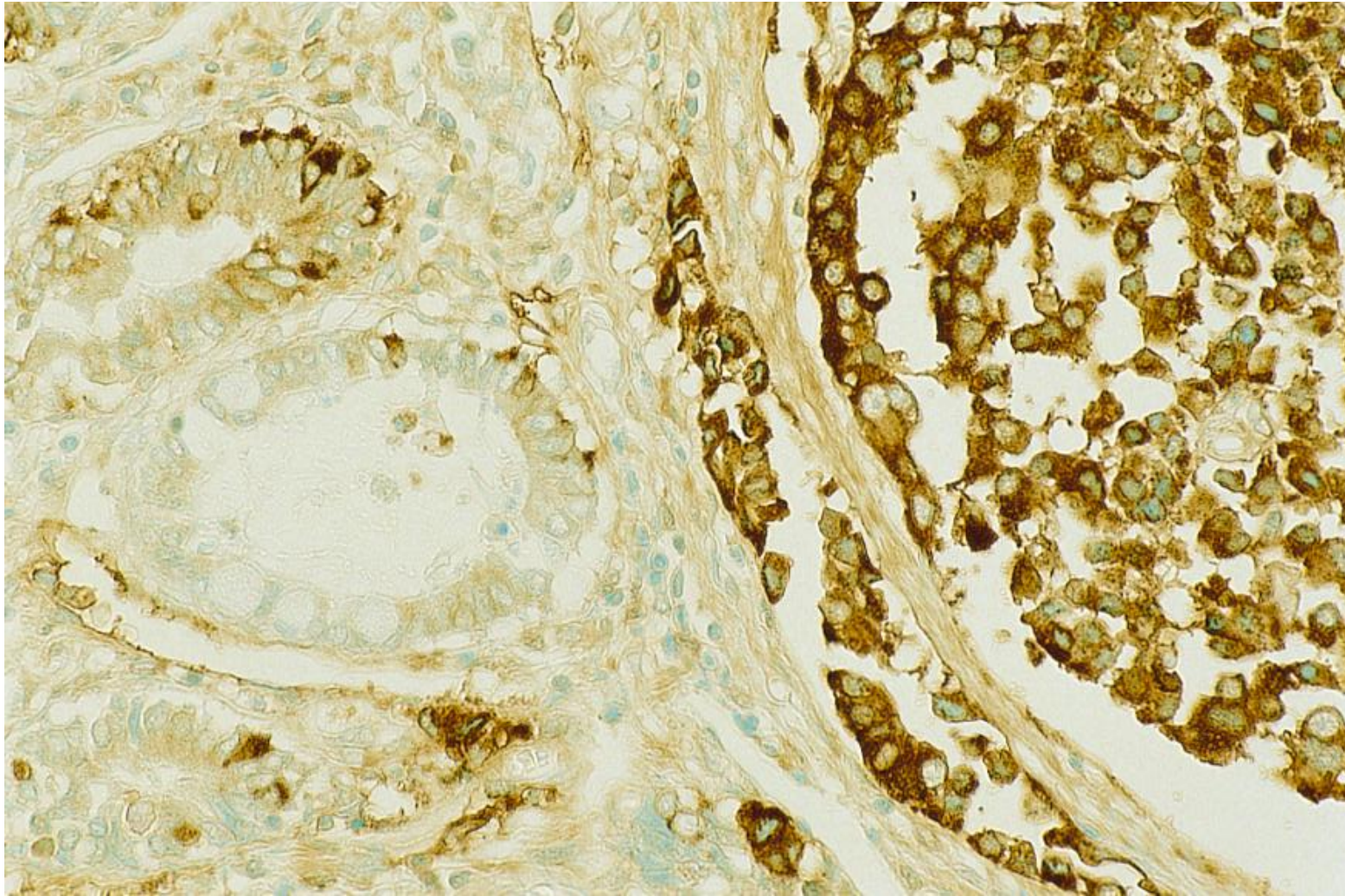
Gross appearance of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. A stenotic tumorous lesion with shallow ulceration is seen.



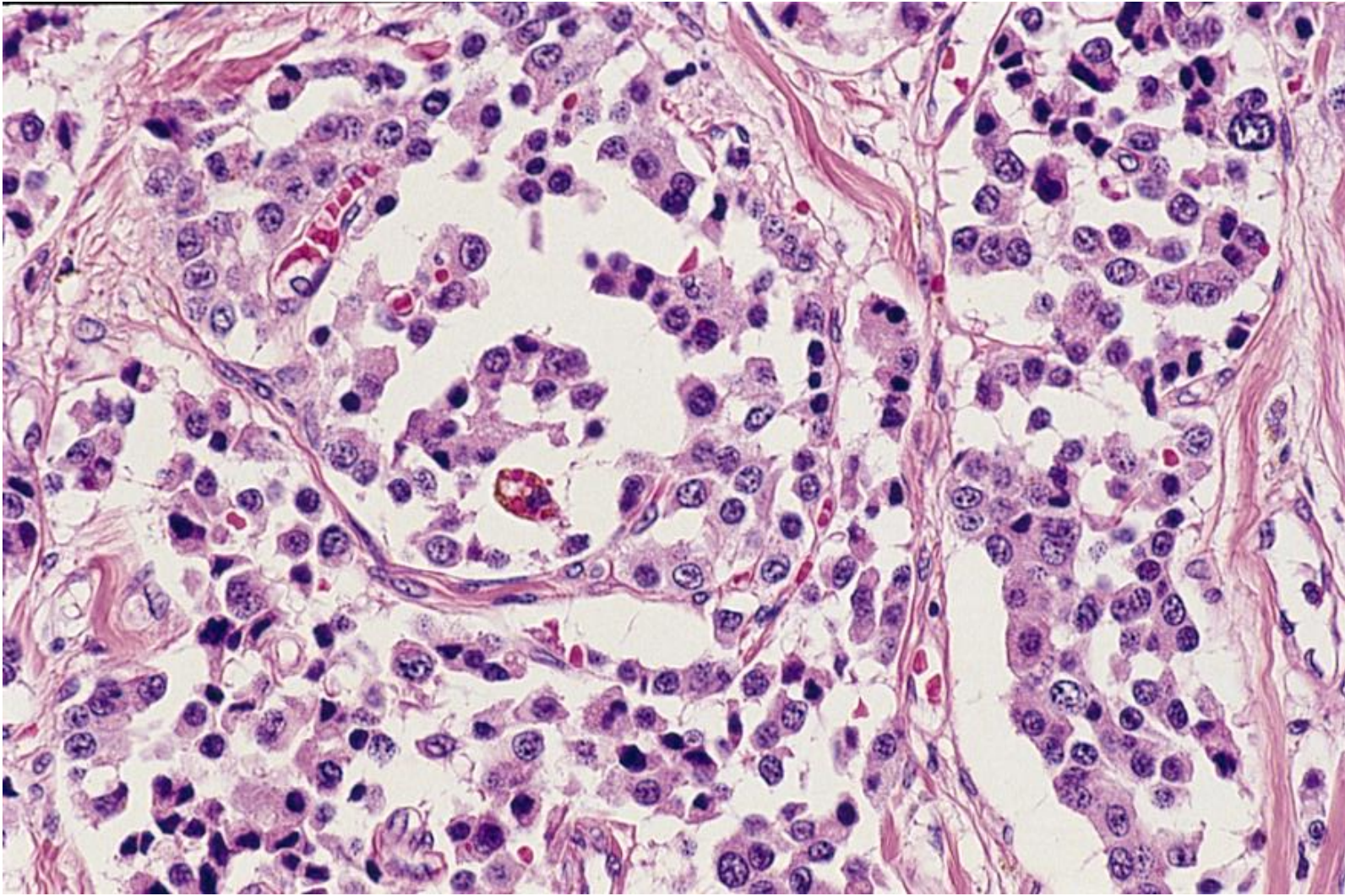
Gross appearance of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. A cut surface after formalin fixation reveals transmural infiltration of solid tumor with shallow ulceration. The tumor size is 15 mm.



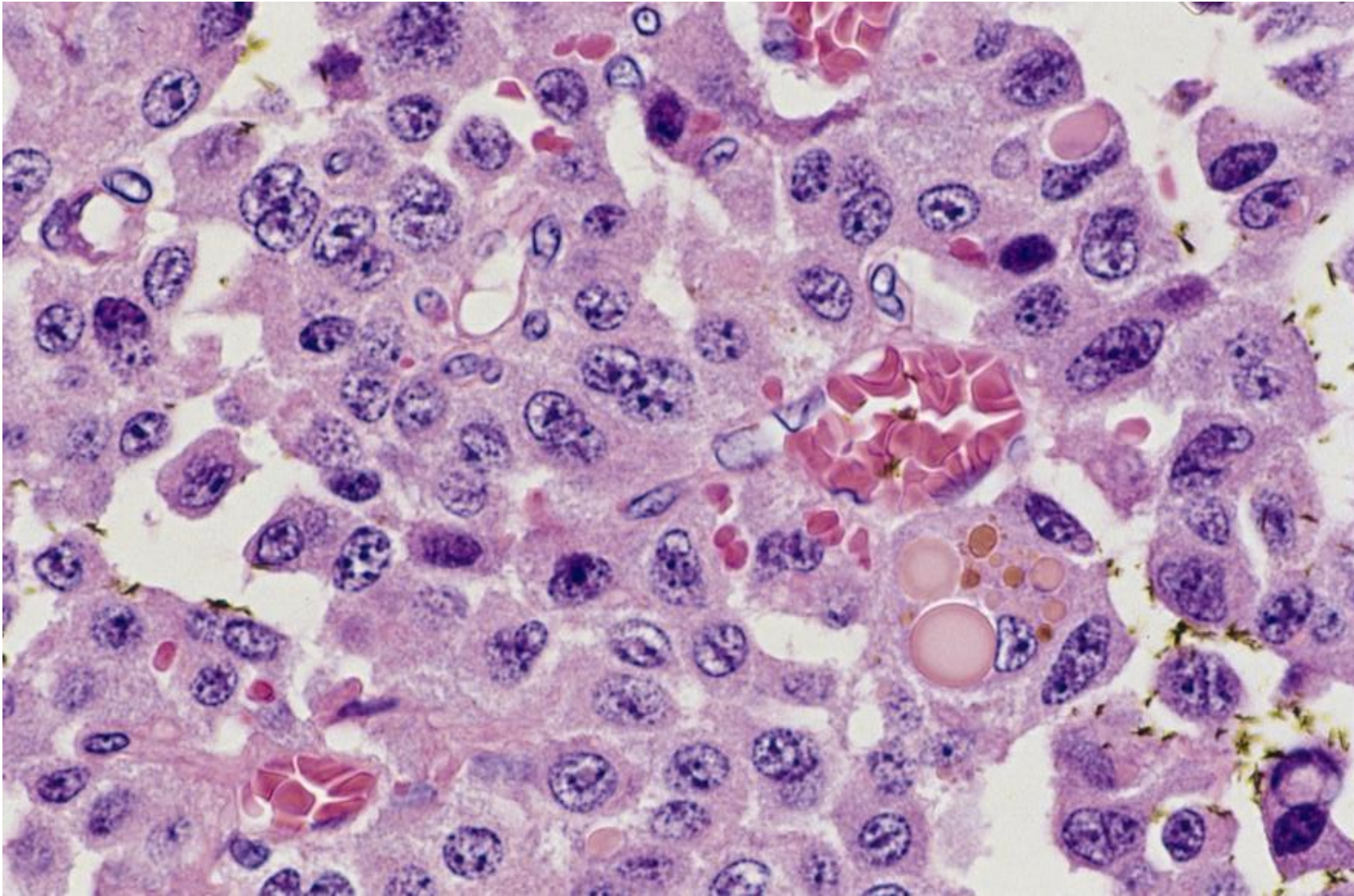
Microscopic appearance of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. Submucosal invasion of small nest-forming tumor cells is seen. The mucosal involvement is focally observed. H&E-1



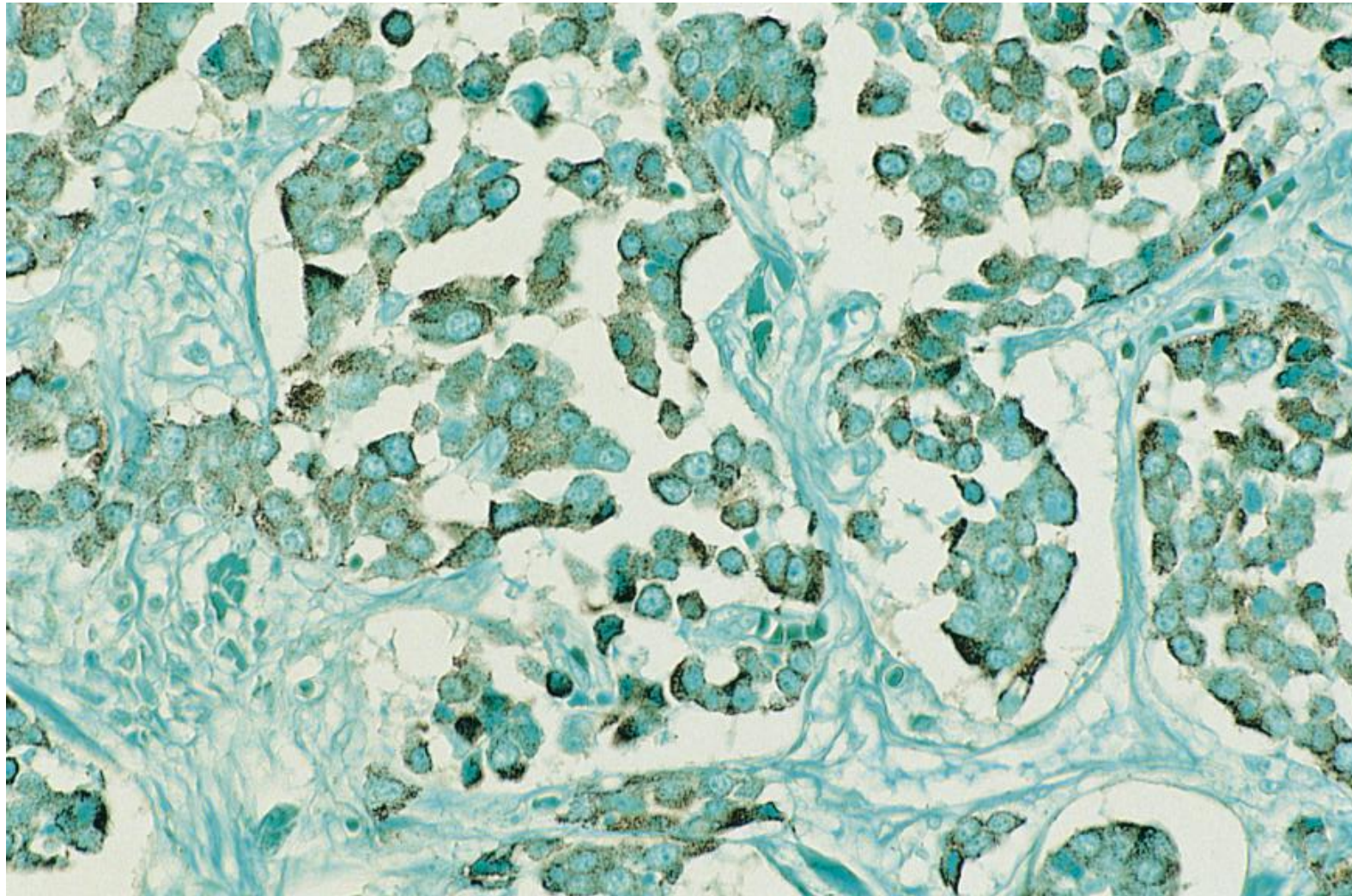
Serotonin production in ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. Submucosally invasive tumor cells are diffusely and strongly immunoreactive for serotonin. Immunostaining for 5-HT



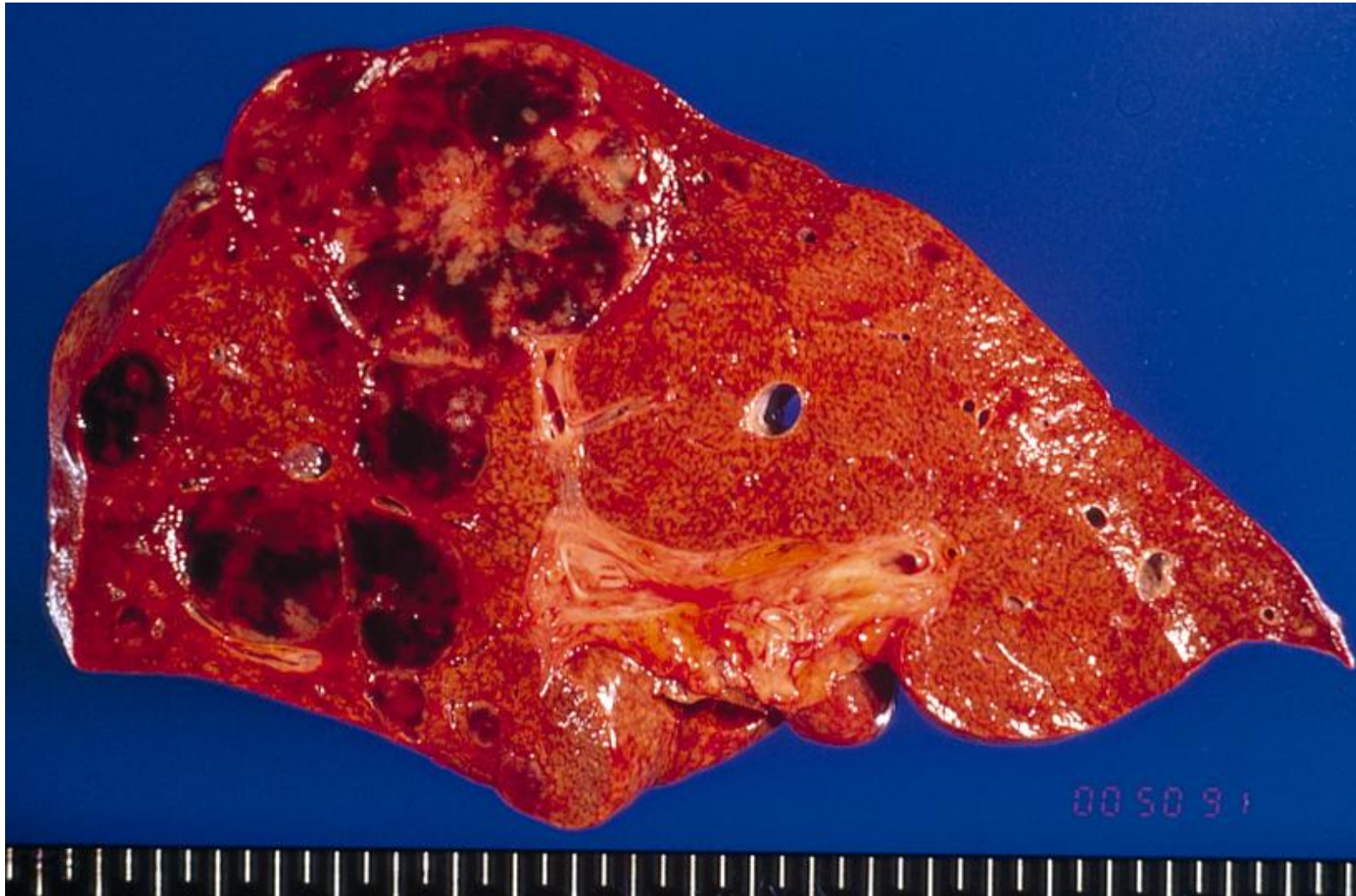
Microscopic appearance of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. Nuclear pleomorphism and hyperchromasia are discerned. H&E-2



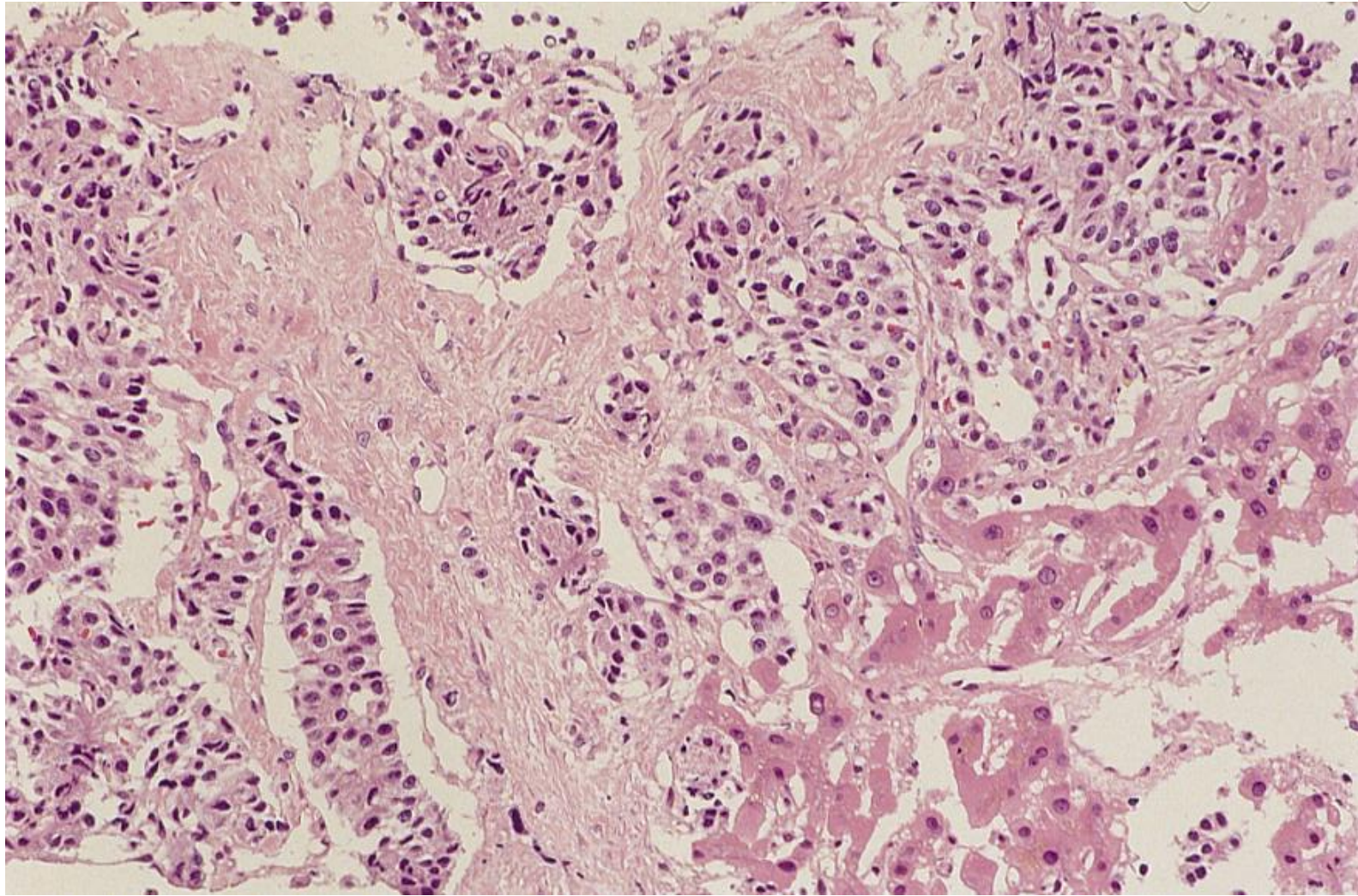
High-powered microscopic appearance of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. The tumor cells show mild nuclear pleomorphism and hyperchromasia. Formation of hyaline droplets is focally associated. H&E-3



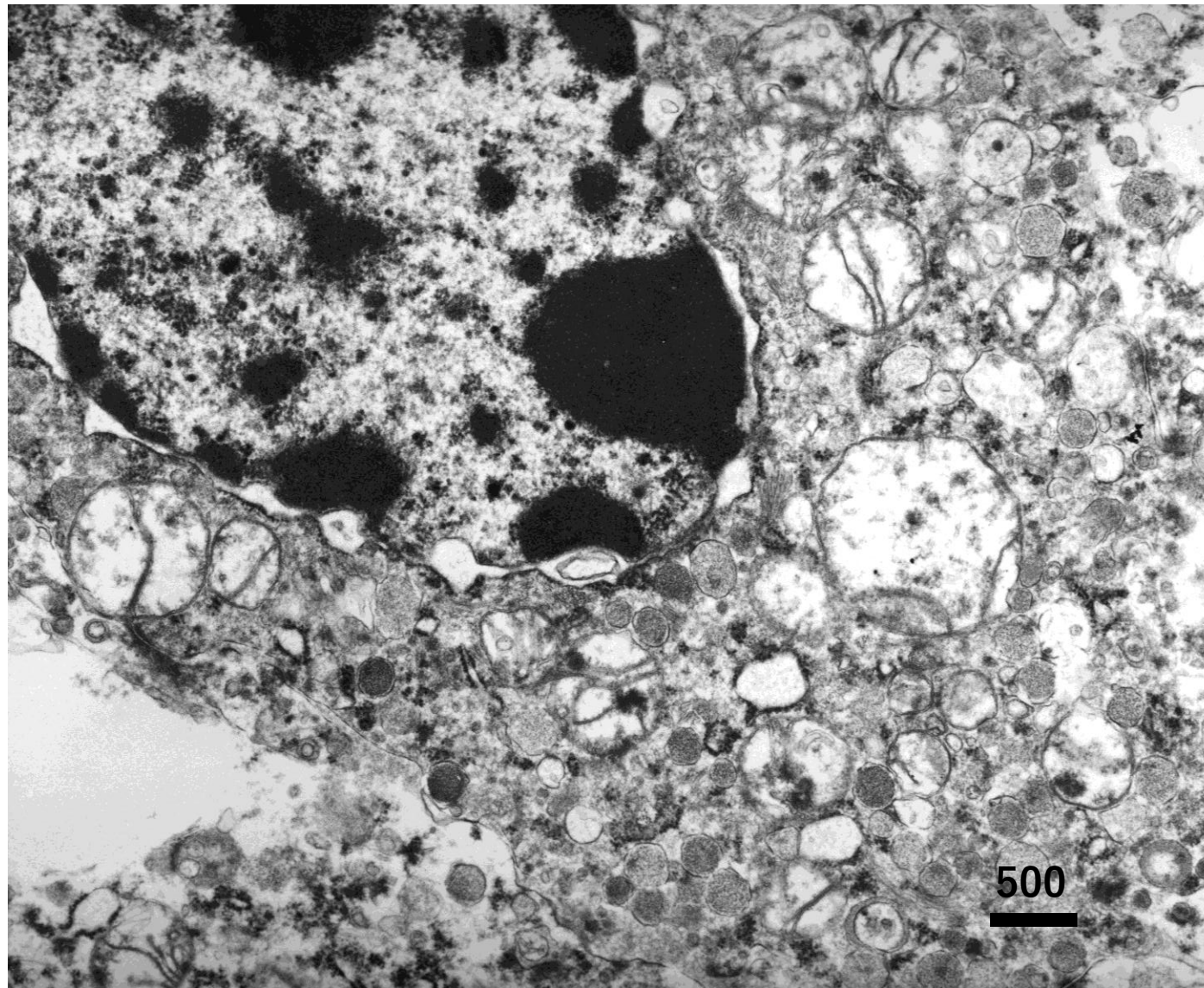
Microscopic appearance of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. The tumor cells possess argyrophilic cytoplasmic granules. Grimelius silver staining



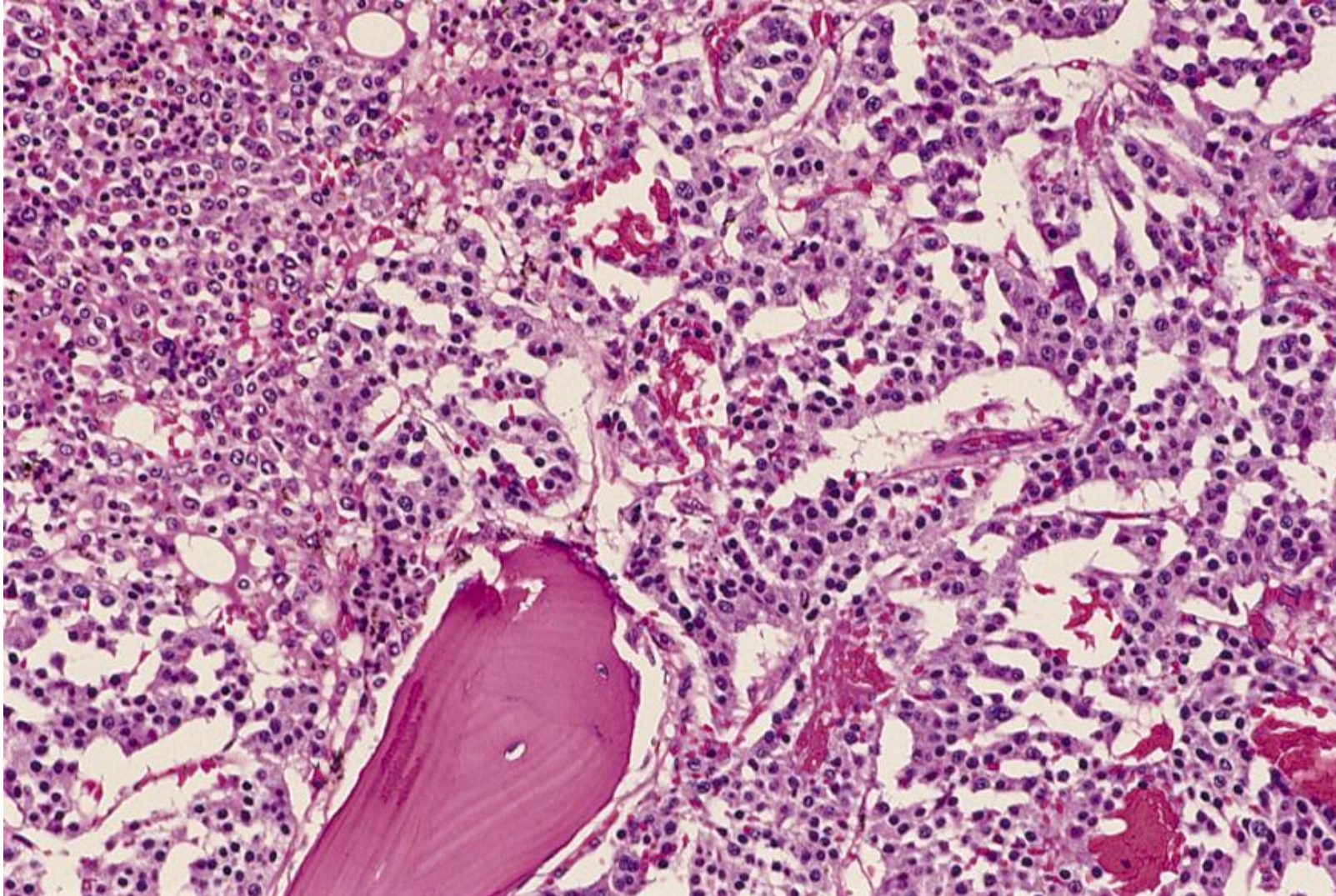
Gross appearance of liver metastasis of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. Large hemorrhagic metastatic lesions are seen in the liver. Palpation of the liver induced flushing syndrome in this case.



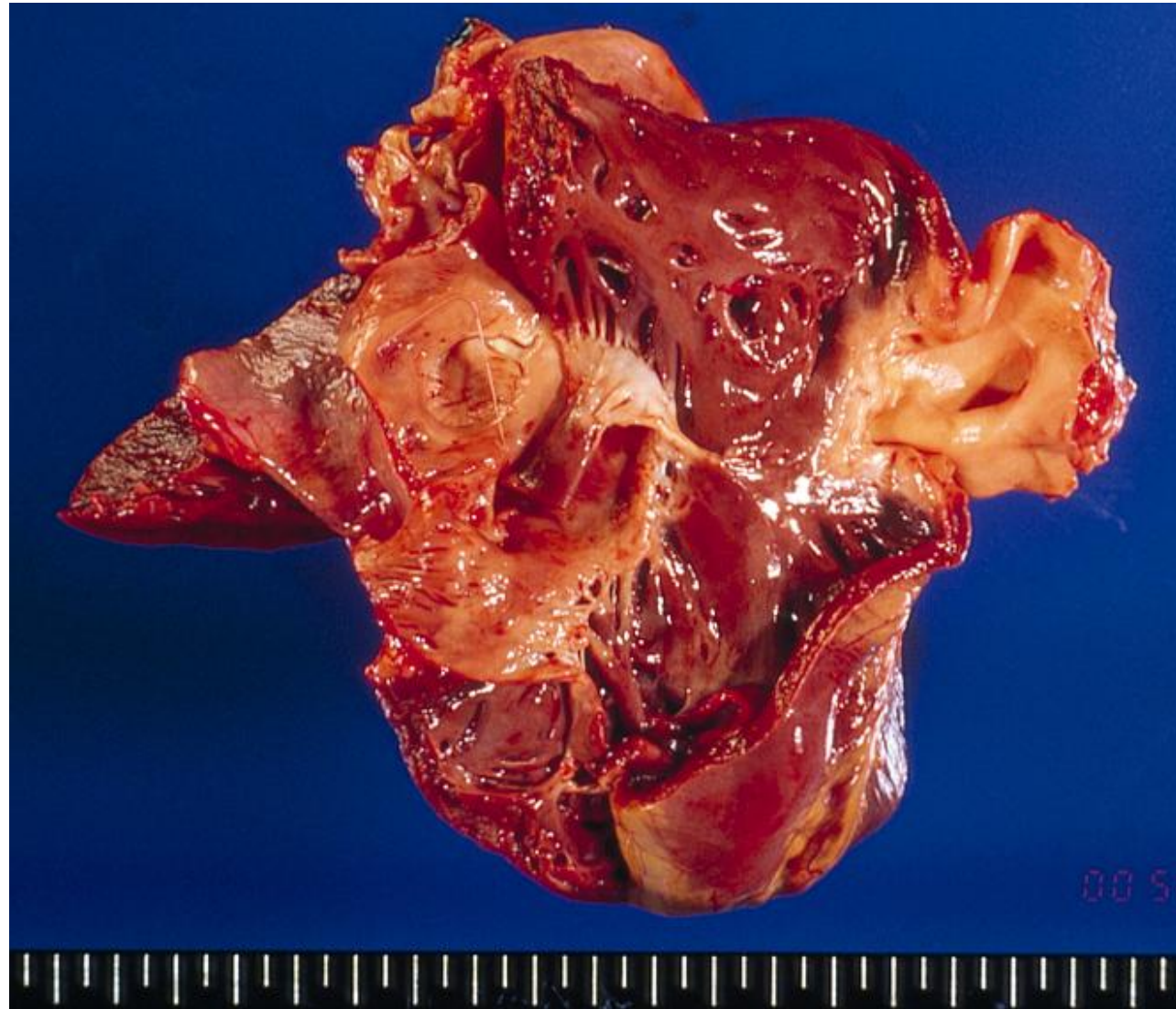
Microscopic appearance of liver metastasis of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. The tumor cells reveal trabecular or ribbon-like arrangements. H&E-4



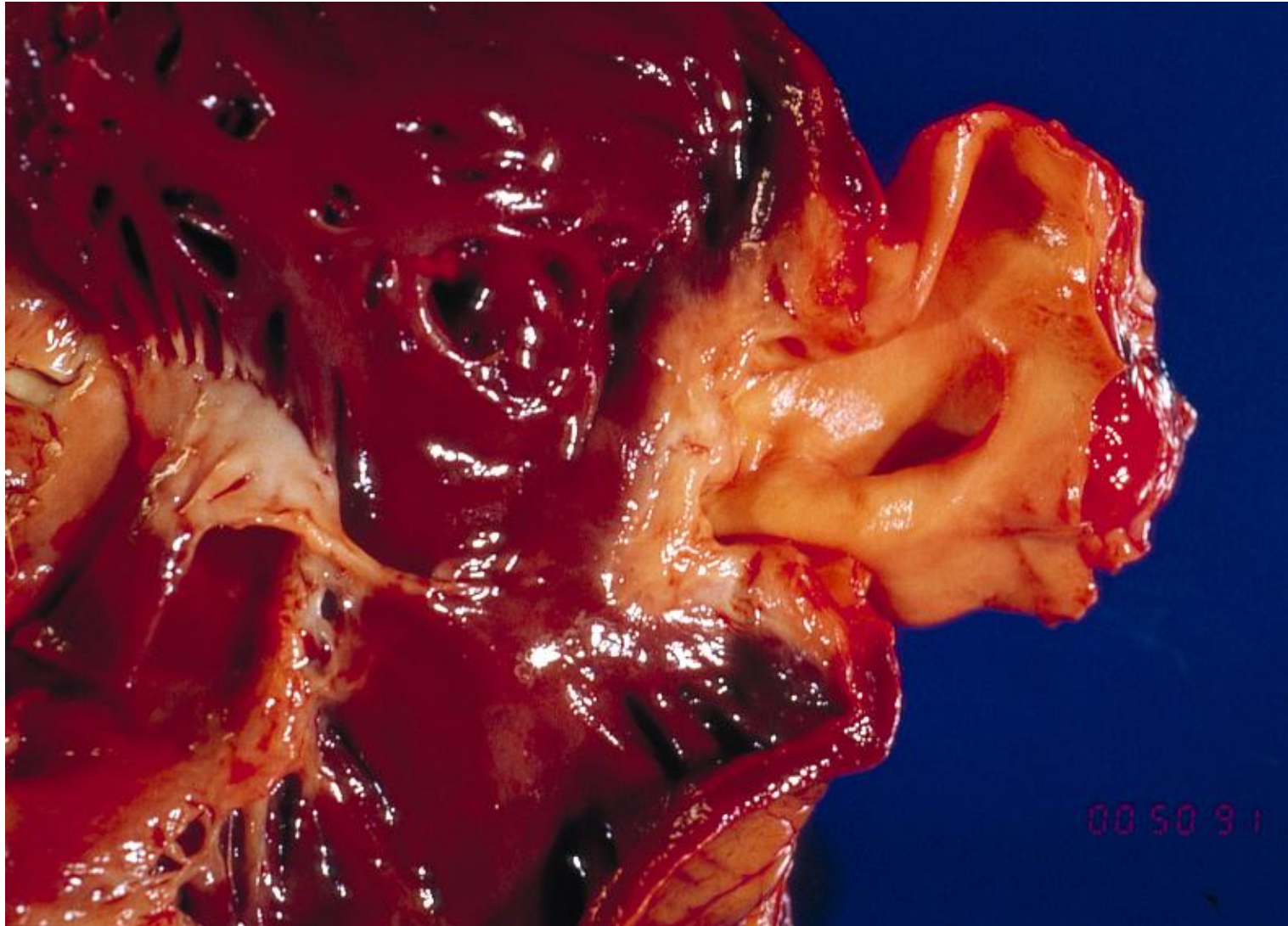
Ultrastructural appearance of liver metastasis of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. The tumor cell contains round-shaped neuroendocrine granules measuring around 300 nm in diameter. Irregular elongated-shaped (serotonin-type) granules are not observed. EM



Microscopic appearance of bone metastasis of ileal neuroendocrine tumor (carcinoid tumor) causing carcinoid syndrome seen in a 70-year-old male patient. The tumor cells reveal trabecular or ribbon-like arrangements. H&E-5



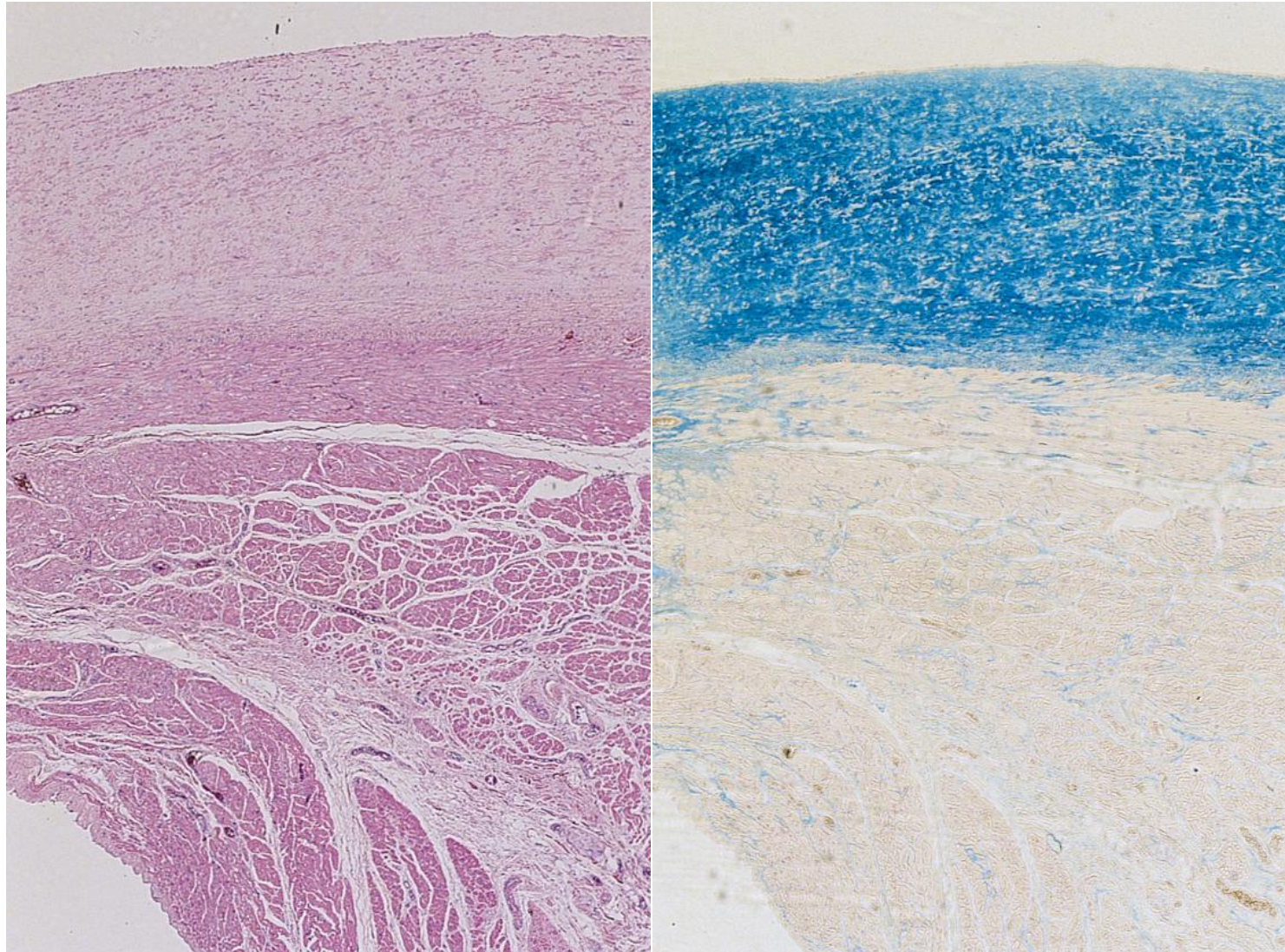
Carcinoid heart disease. Thickening of tricuspid valve and pulmonic valve is discerned. Persistently high venous levels of serotonin have provoked deposition of myxofibrous material of the right-sided heart valves.



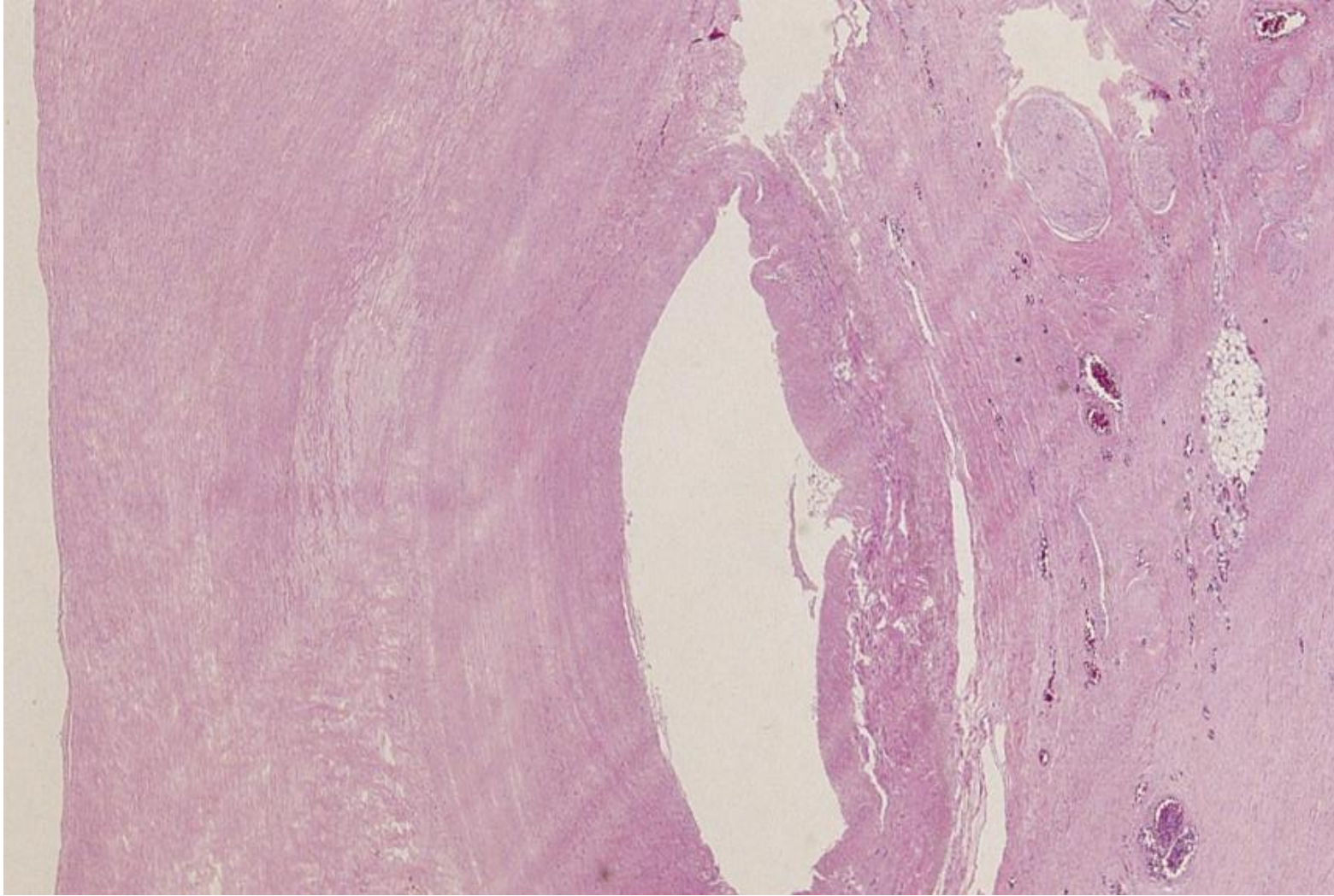
Carcinoid heart disease. Thickening of tricuspid valve and pulmonic valve is discerned. Persistently high venous levels of serotonin have provoked deposition of myxofibrous material of the right-sided heart valves.



Carcinoid heart disease. Myxofibrous thickening is also discerned in the mitral valve in this case.



Carcinoid heart disease. Myxofibrous thickening of the intima of the endocardium is microscopically discerned. Deposition of acid mucosubstance is demonstrated. Left: H&E, right: colloidal iron staining



Retroperitoneal fibrosis. Deposition of myxoid substances is seen in the retroperitoneal space, provoking ureteric stenosis and hydronephrosis. Stenosis of inferior mesenteric artery lead to ischemic necrosis of the sigmoid colon. H&E