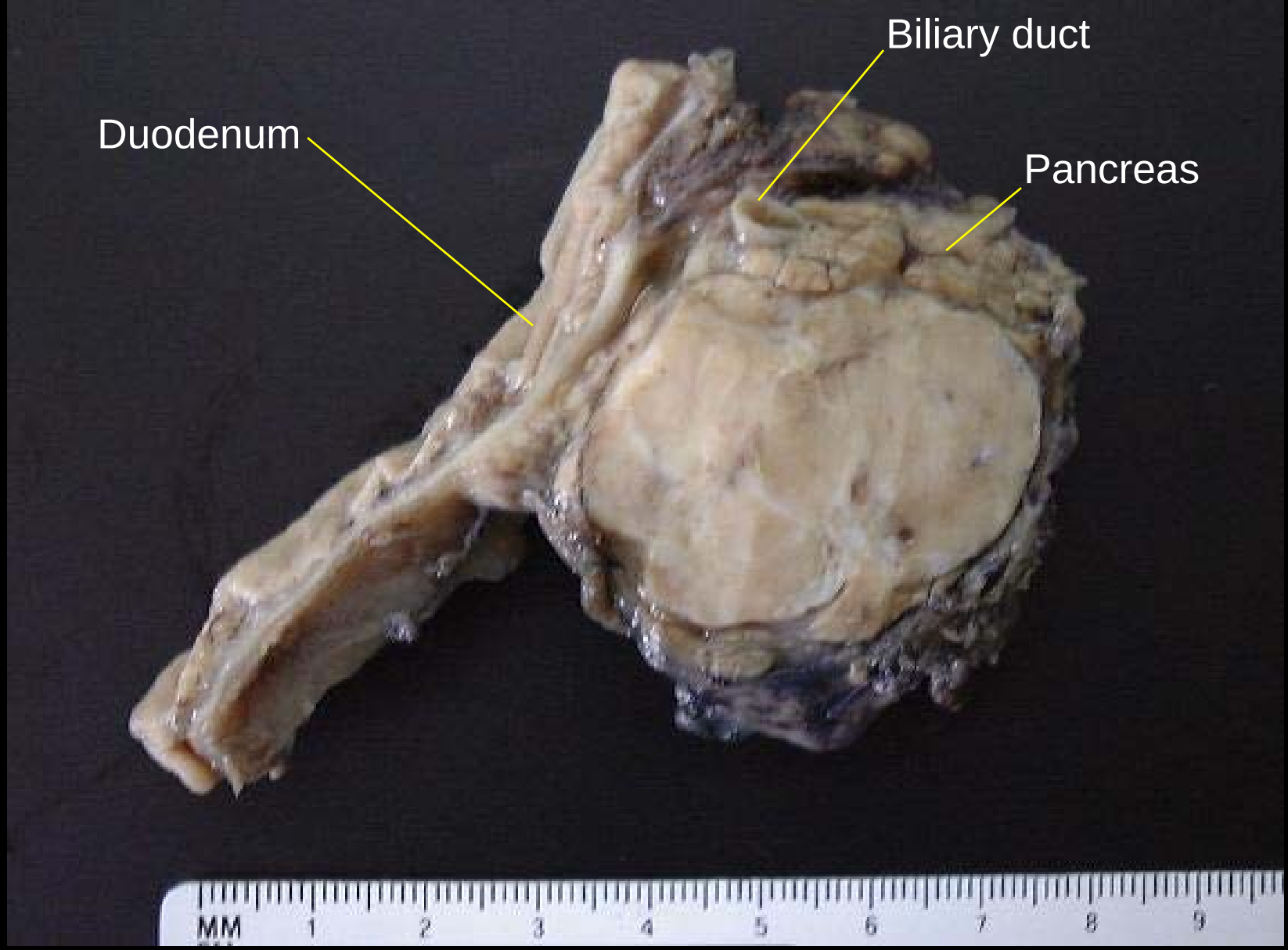
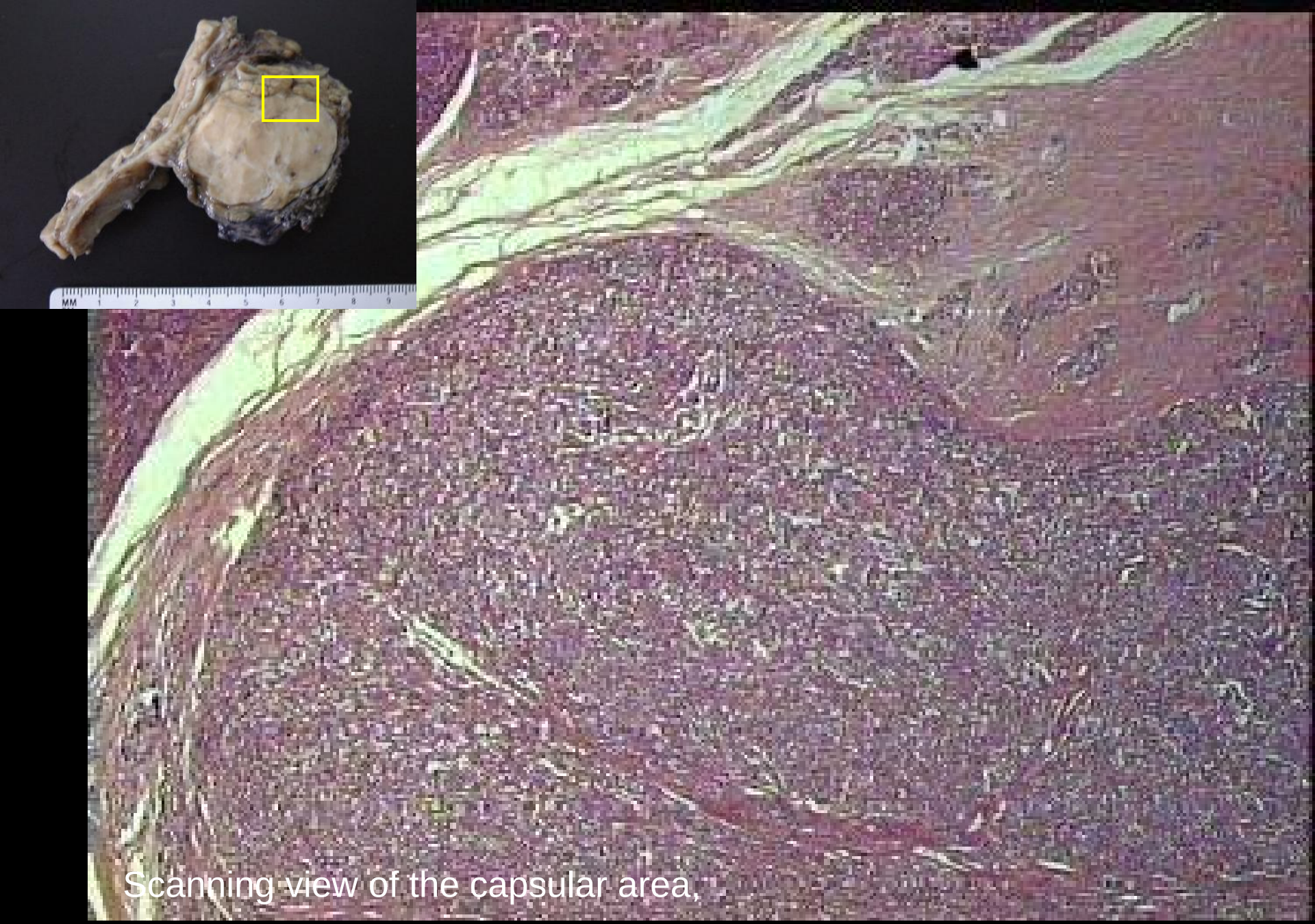
HDF Case 992741

- 34 yo female, with a familial history of MEN1 (father and sister).
- Previous surgery 19 years ago for hyperparathyroidism , and 9 years ago for a prolactinoma.
- Recently she complains from severe asthenia, anorexia with weight loss (8 kg. in 2 months), diarrhea, flush.
- Abdominal MRI reveals a tumor of 5 cm. of the head of the pancreas.
- Whipple's procedure performed.



Specimen opened, showing a well circumscribed mass embedded in the head of the pancreas.



Scanning view of the capsular area.

MAIN MENU

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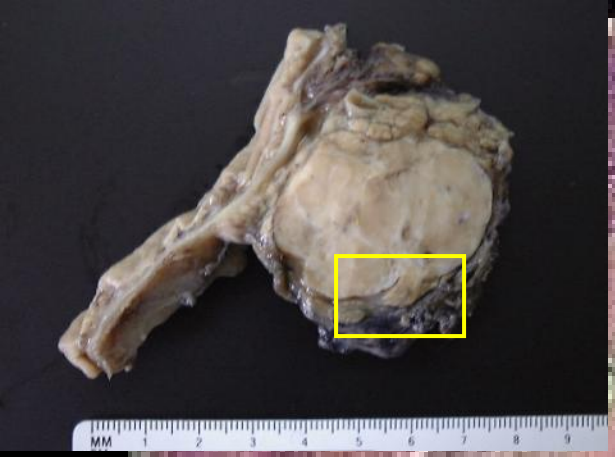

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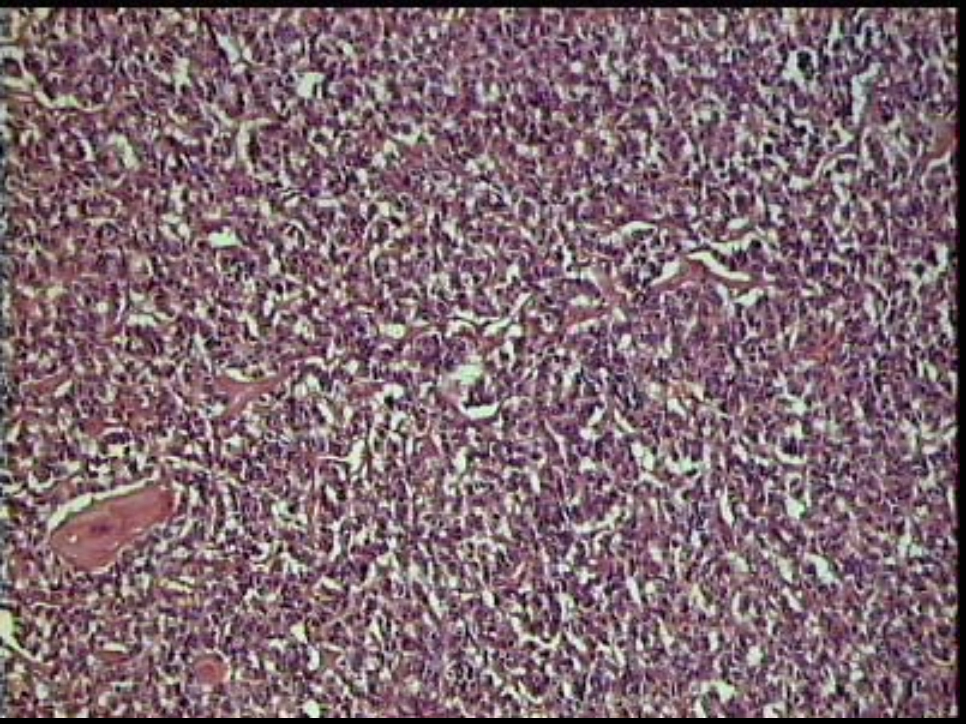

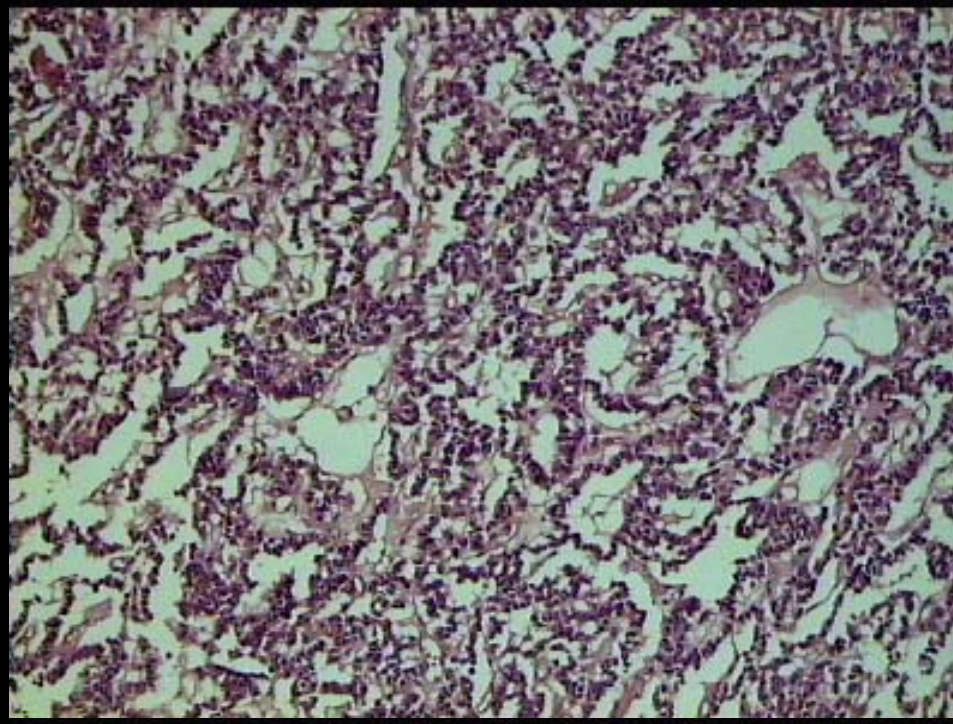

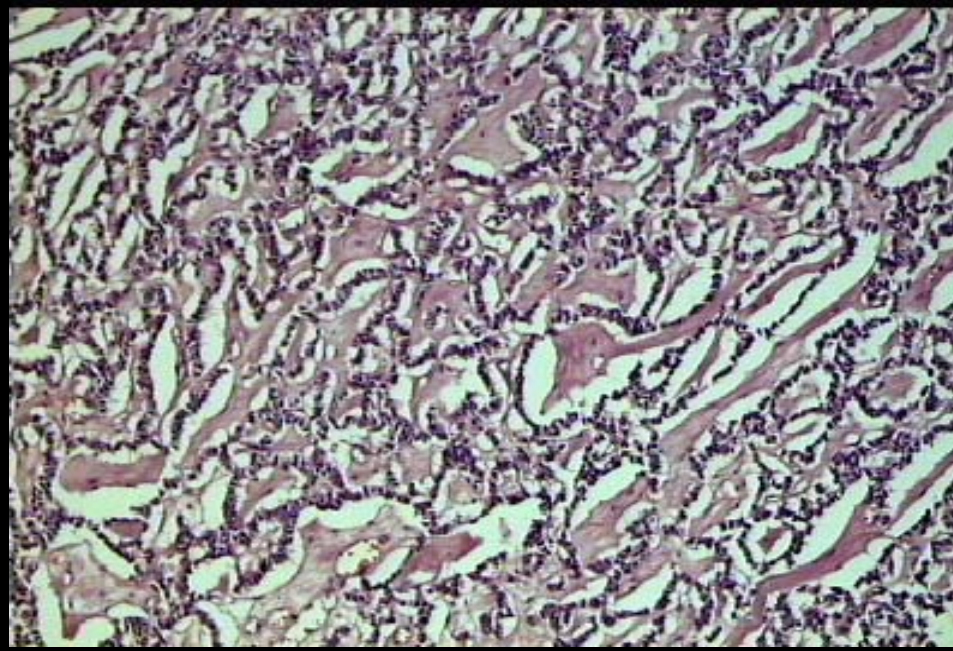

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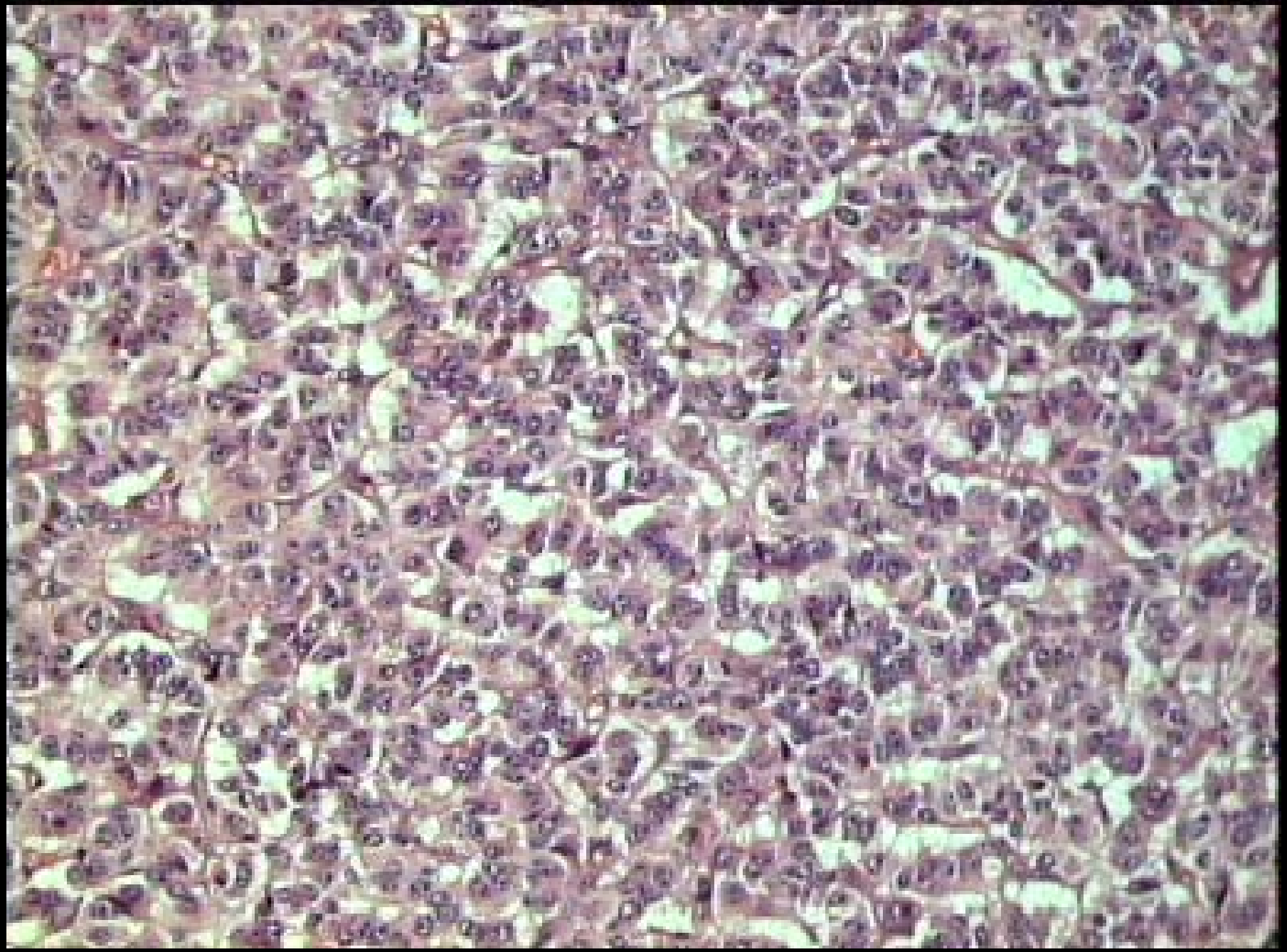

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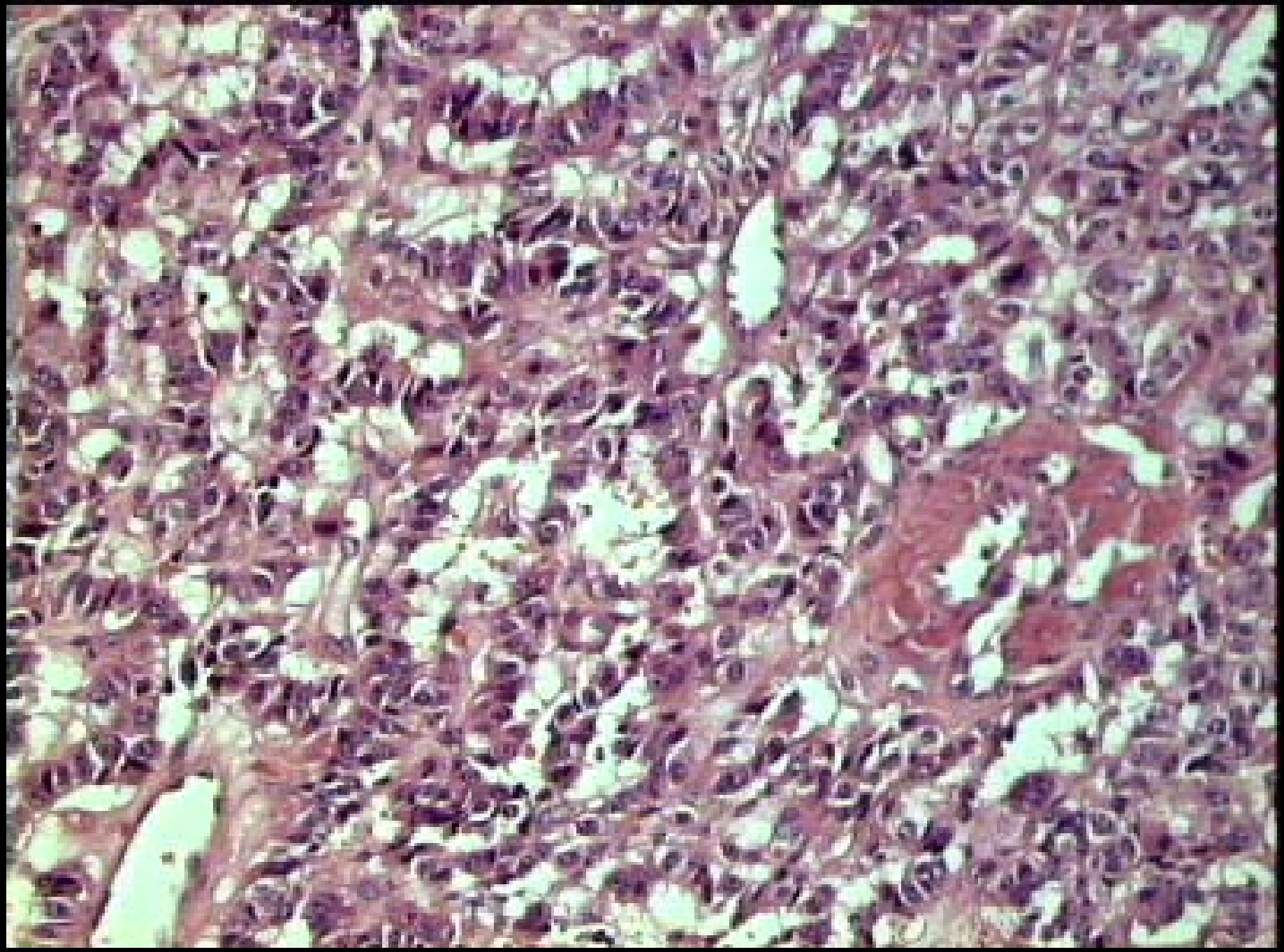

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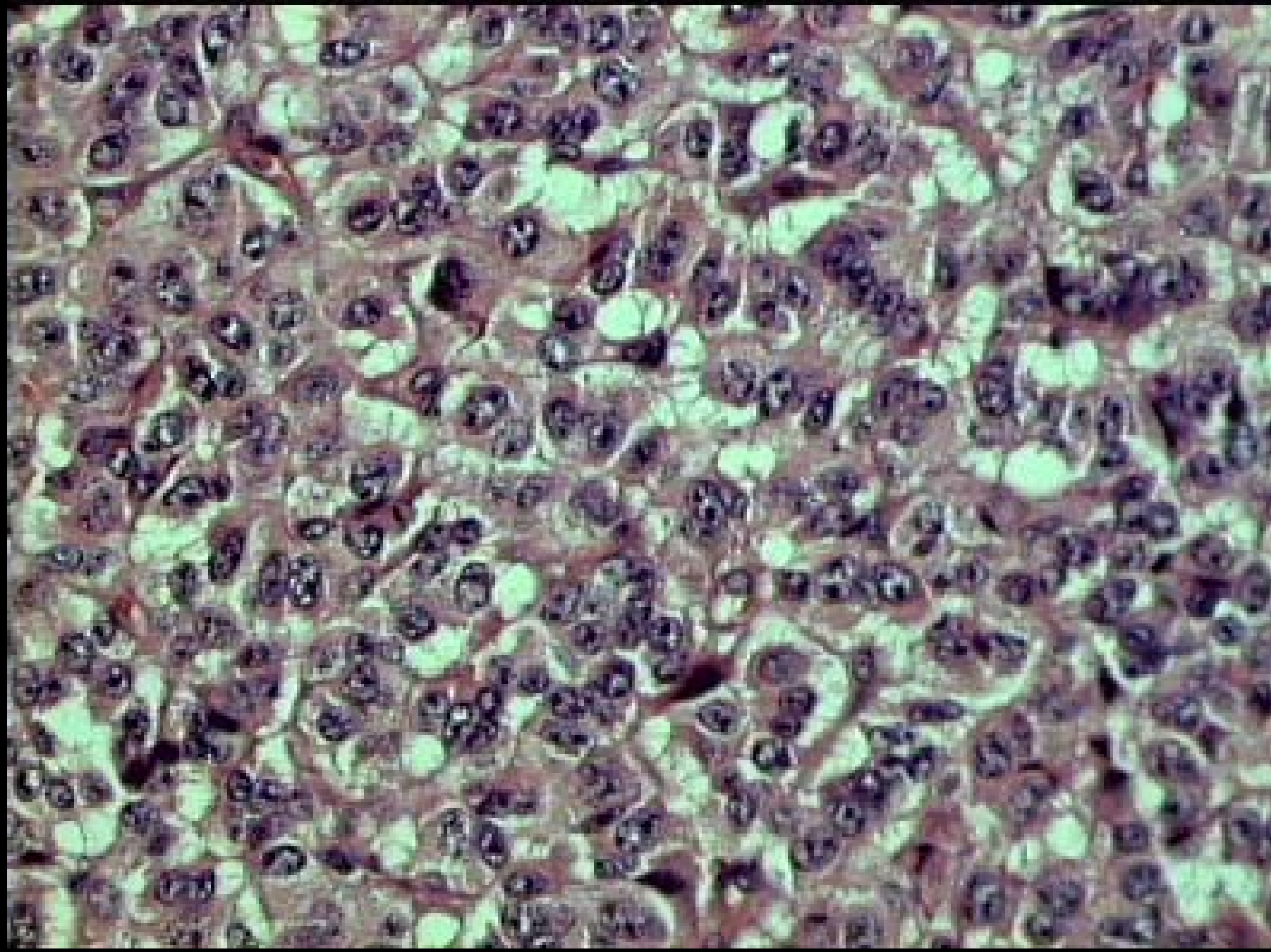

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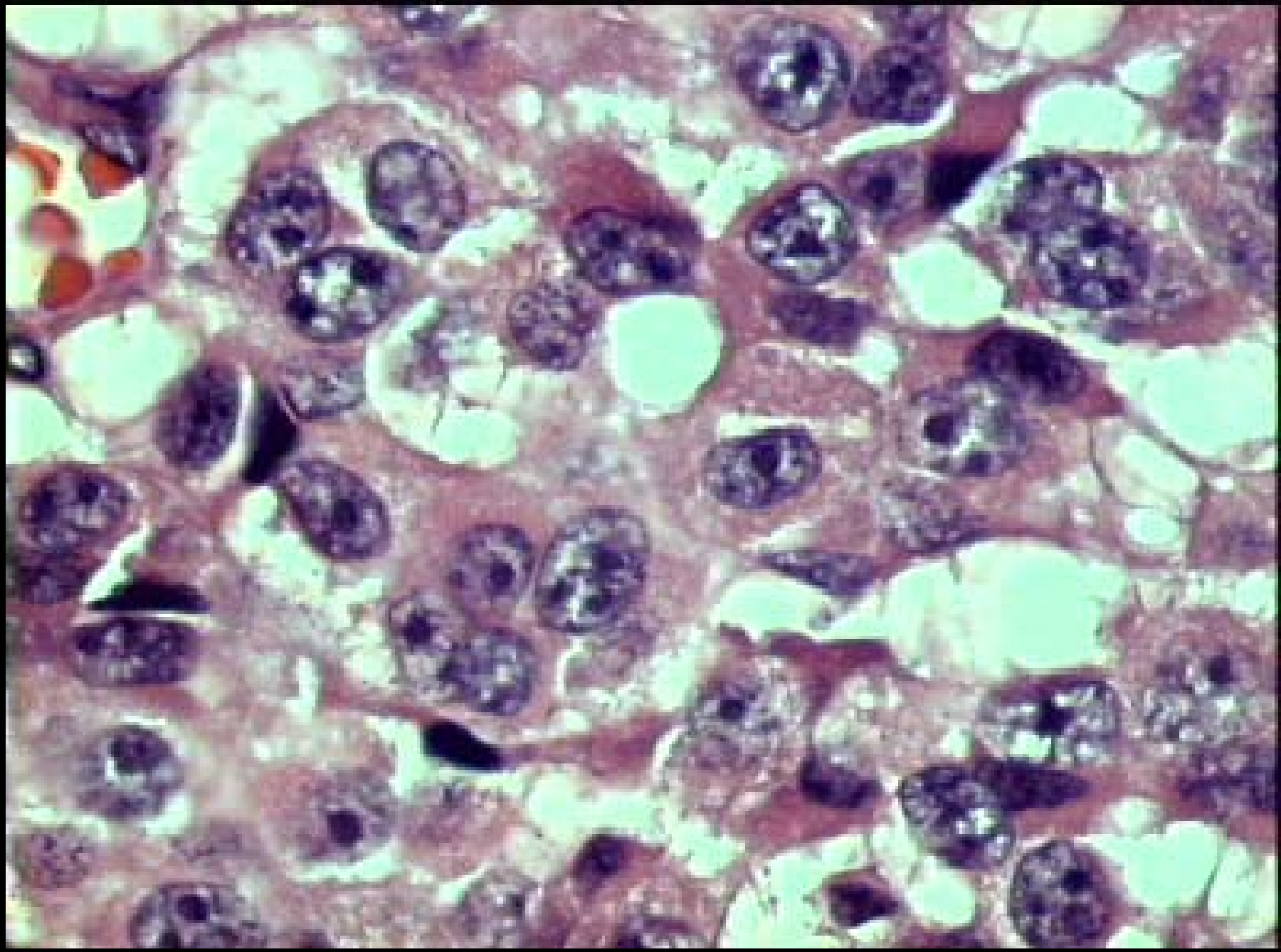

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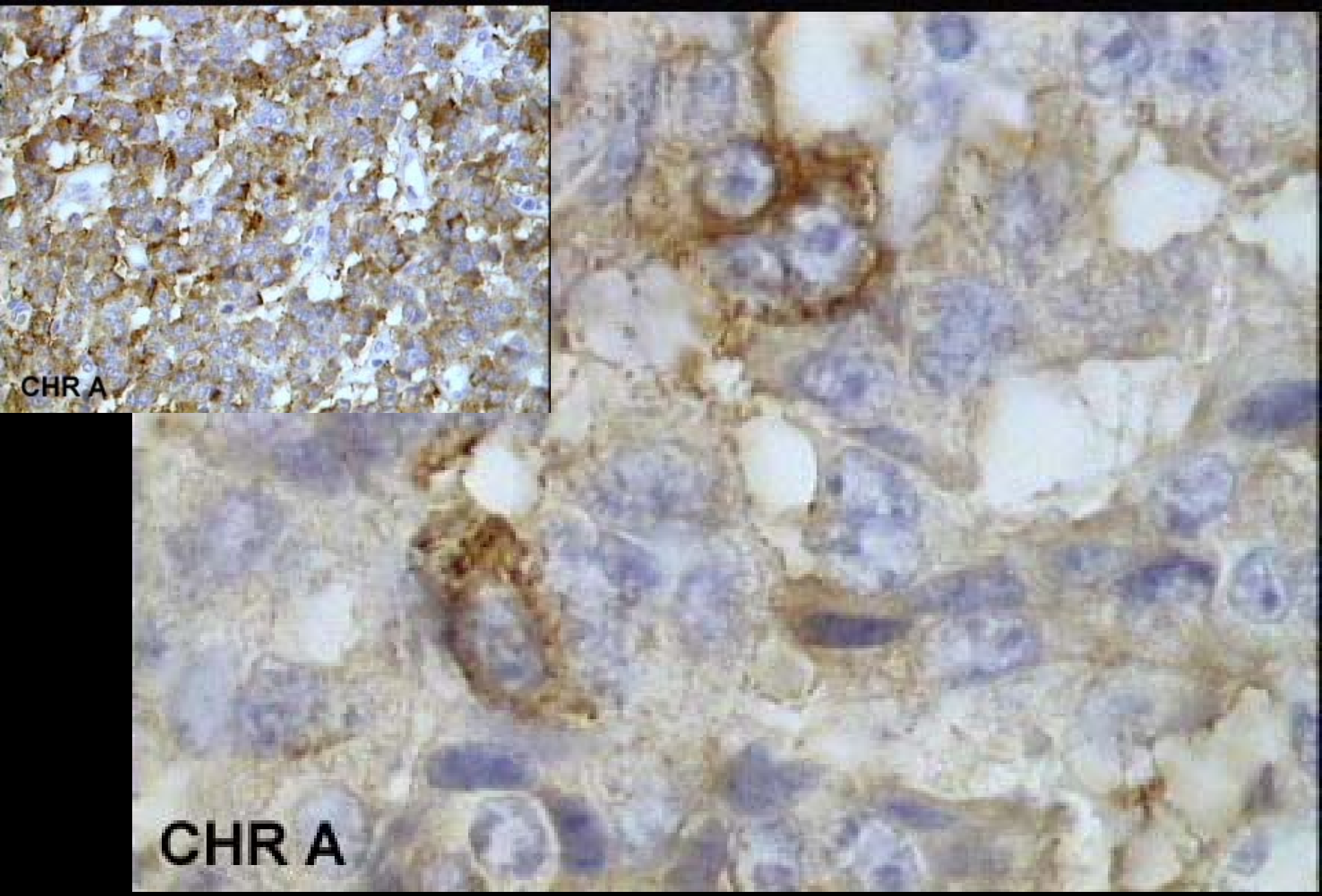
CHR A
CHR A

Multiple endocrine neoplasia

- Multiple endocrine neoplasia (MEN) syndromes are inherited in an autosomal dominant fashion and are characterized by hyperplastic or neoplastic proliferation of more than one endocrine gland. Three distinct types have been described, although they sometimes seem to overlap.
- MEN type I (Werner's syndrome) is characterized by involvement of the pituitary gland (adenomas), pancreas, and parathyroid glands (chief cell hyperplasia). The pancreatic abnormalities are represented by G-cell tumors in 50% of the cases, beta cell tumors in 30%, VIP cell tumors in 12%, and alpha cell tumors in less than 5%.
- Thus the main clinical manifestations are primary hyperparathyroidism, the Zollinger-Ellison syndrome, and acromegaly or hypopituitarism. Other abnormalities, although less constant, involve the adrenal cortex and thyroid gland (in the form of nodular hyperplasia or adenomas), carcinoid tumors of various locations but mainly of foregut derivatives (lung, thymus, and gastrointestinal tract), multiple soft tissue lipomas, and Menetrier's disease of the stomach. We (Rosai et al) have seen cases of MEN type I with thymic carcinoid tumors and others associated with multiple carcinoid tumors of the gastric fundus. The MEN I gene has been located to a small region in chromosome I Iq 13