

HDF Case 981678

- 35 yo female, treated for seizures (epilepsy?), presented 2 years ago loss of consciousness with hypoglycemia. A fasting blood glucose test, revealed levels below 50 mg%. MRI done then was normal.
- Recently an abdominal CT discloses the presence of a hypervascular lesion in the head of the pancreas.
- Surgical resection of the superficial nodule was performed.



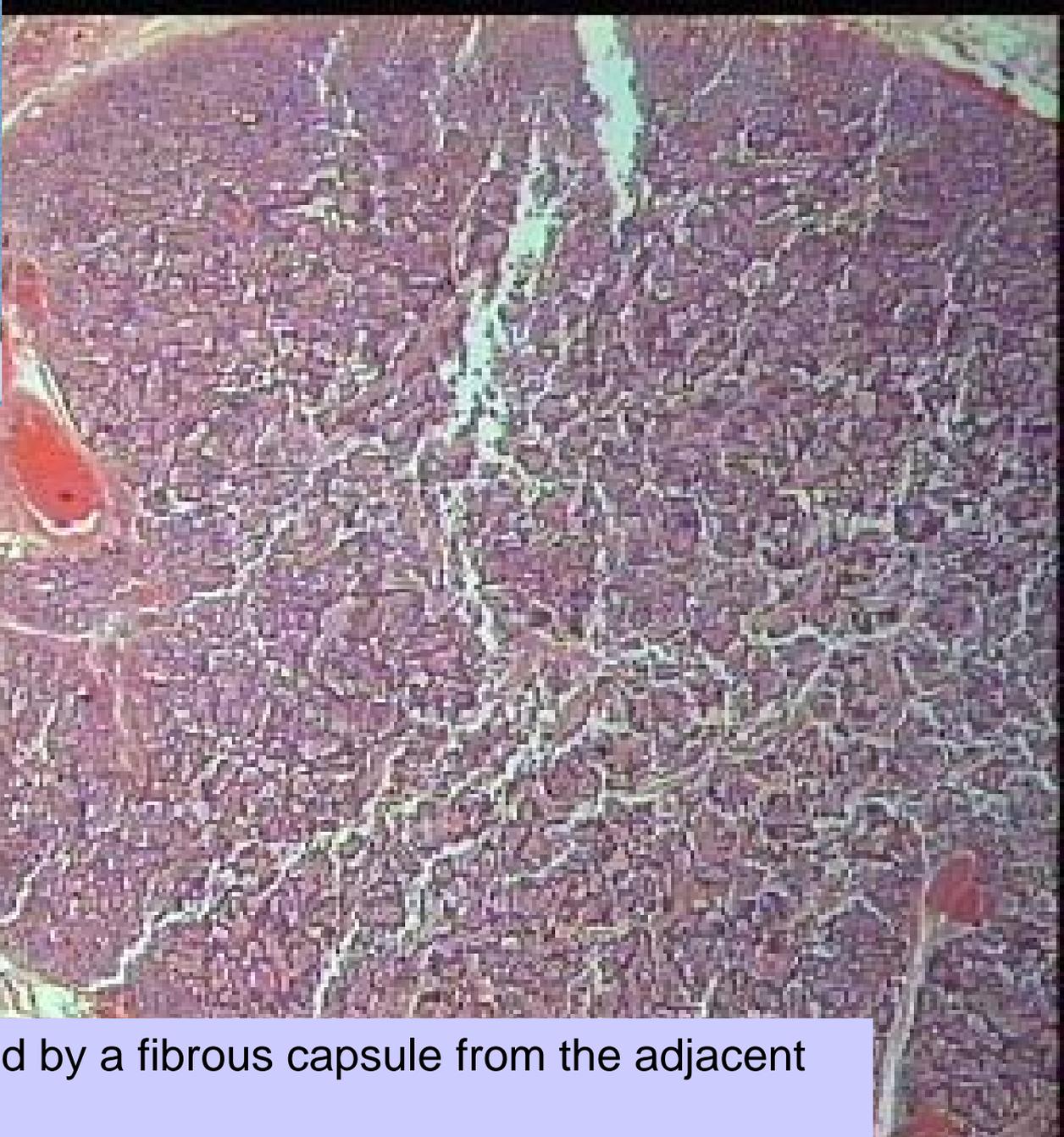
The specimen submitted for frozen section was a well circumscribed, soft, nodule, tan in colour having the gross appearance, of a lymph node.



MAIN MENU



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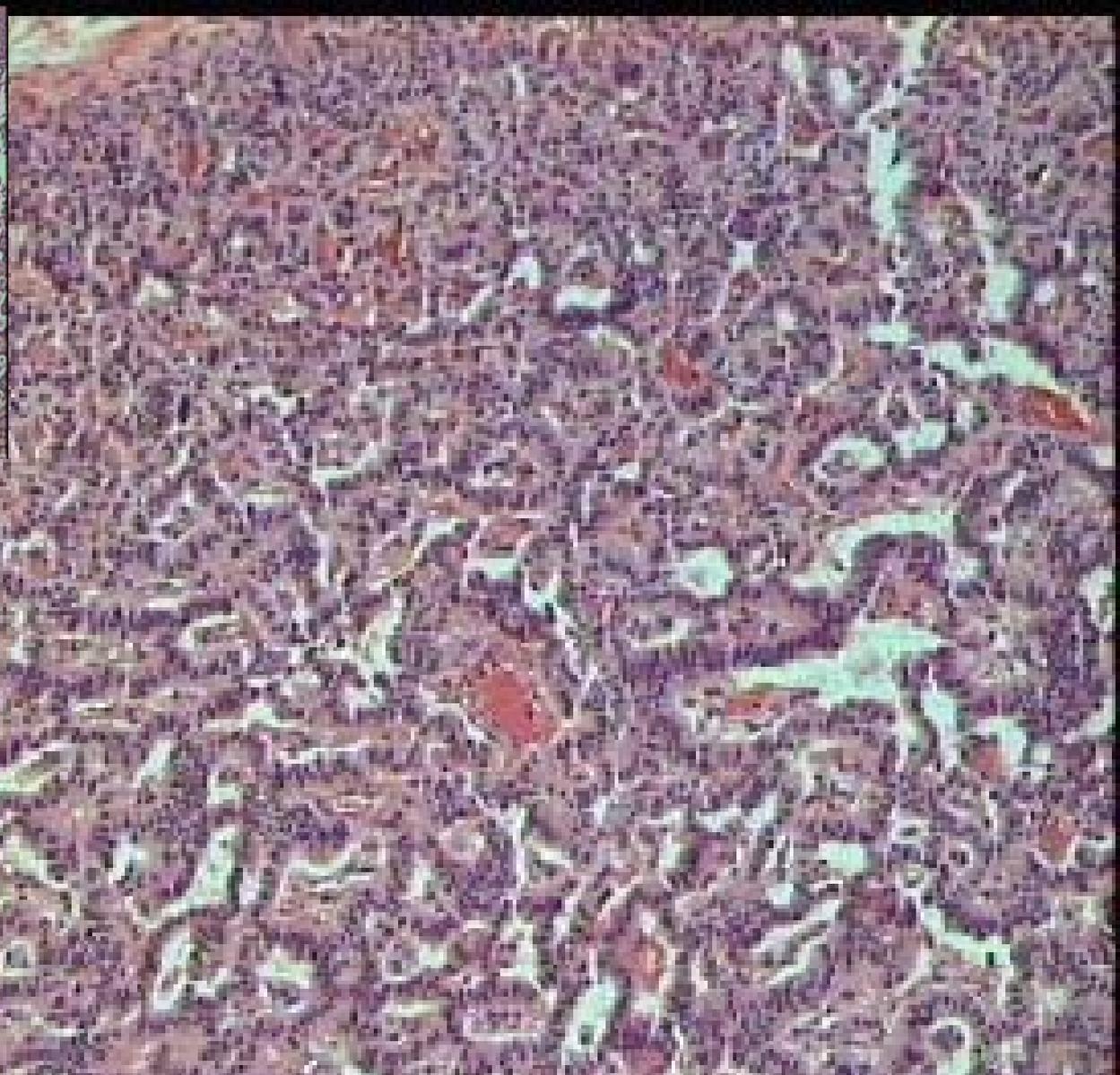


The nodule is separated by a fibrous capsule from the adjacent pancreatic tissue.

MAIN MENU



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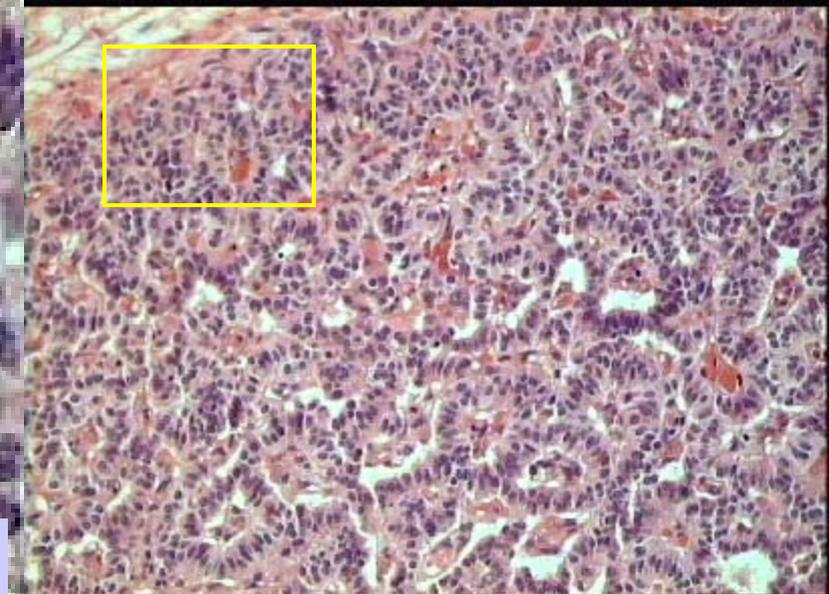
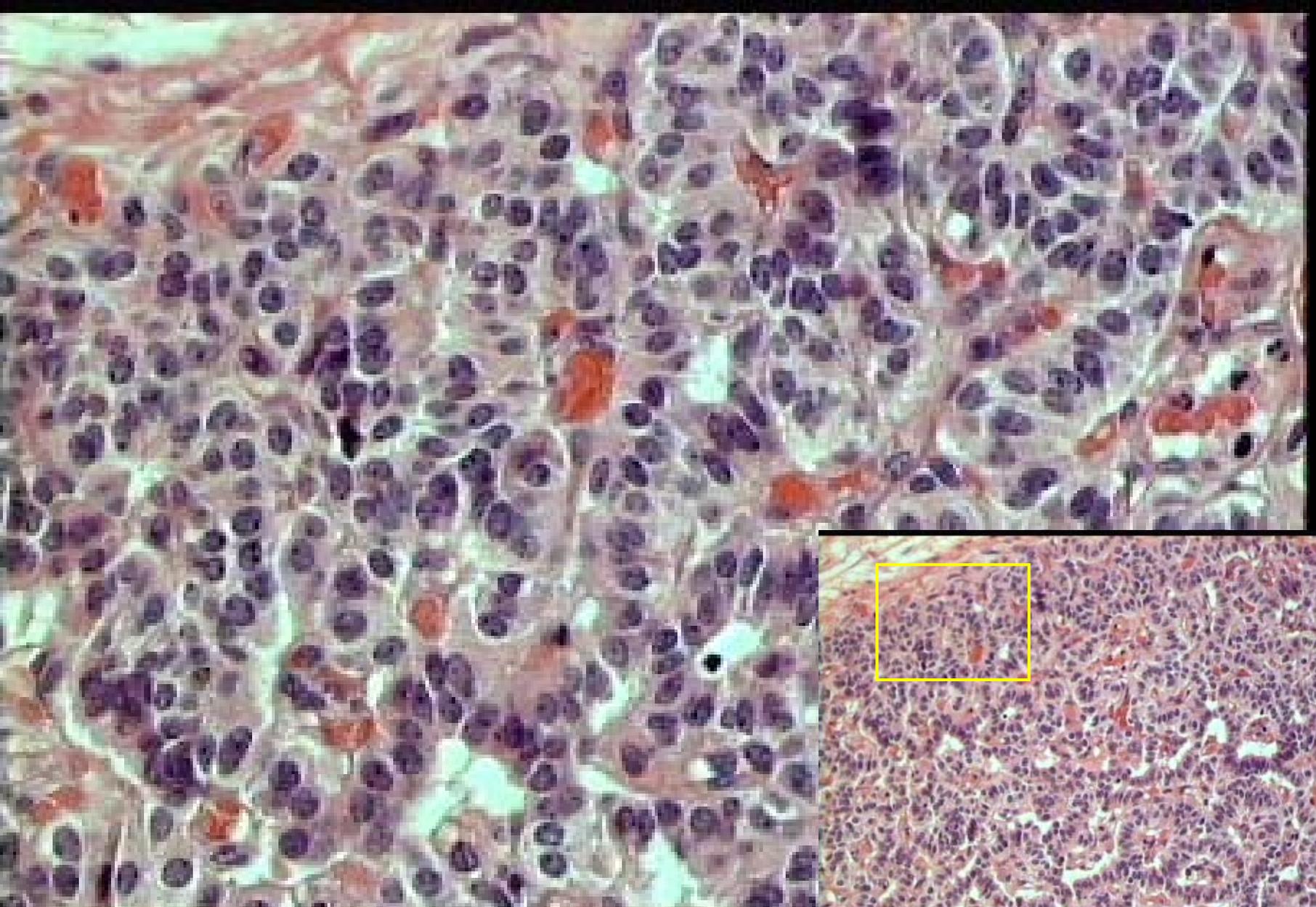


At low power, peripheral compact area surrounds a central portion showing a thin trabecular anastomosing cord pattern.

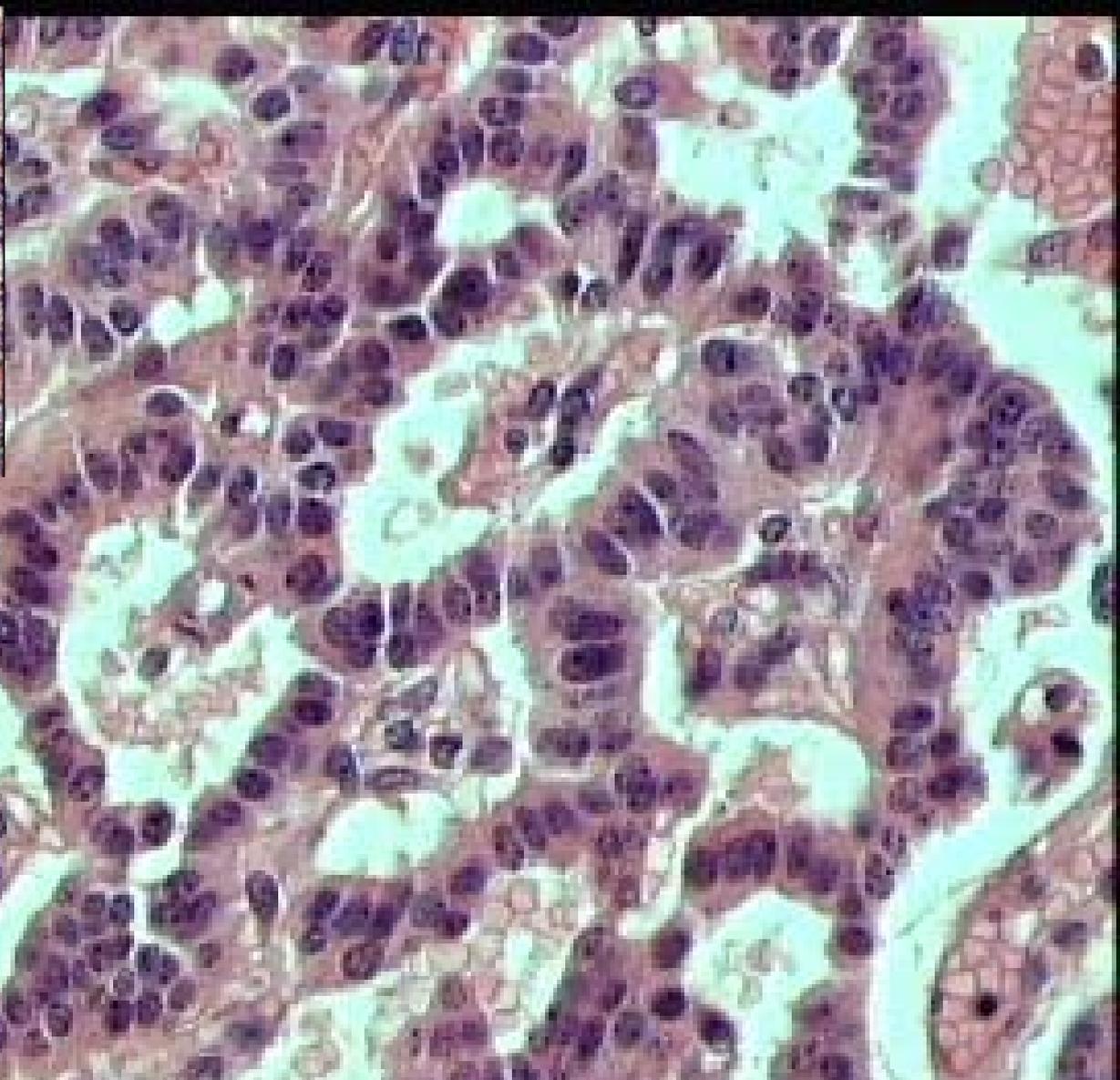
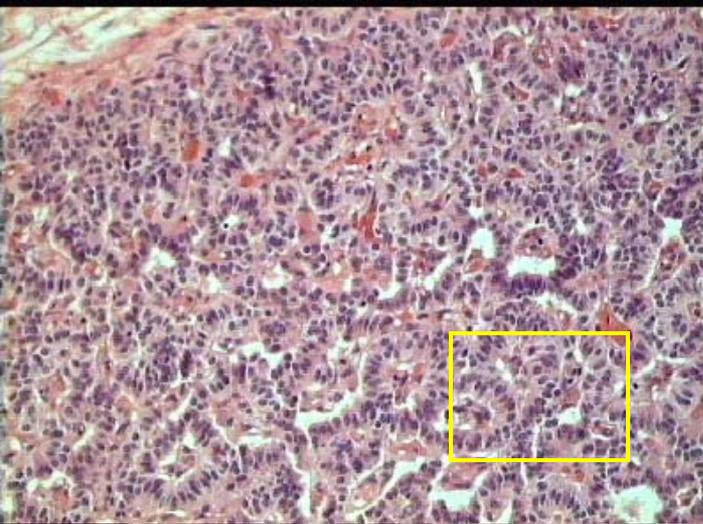
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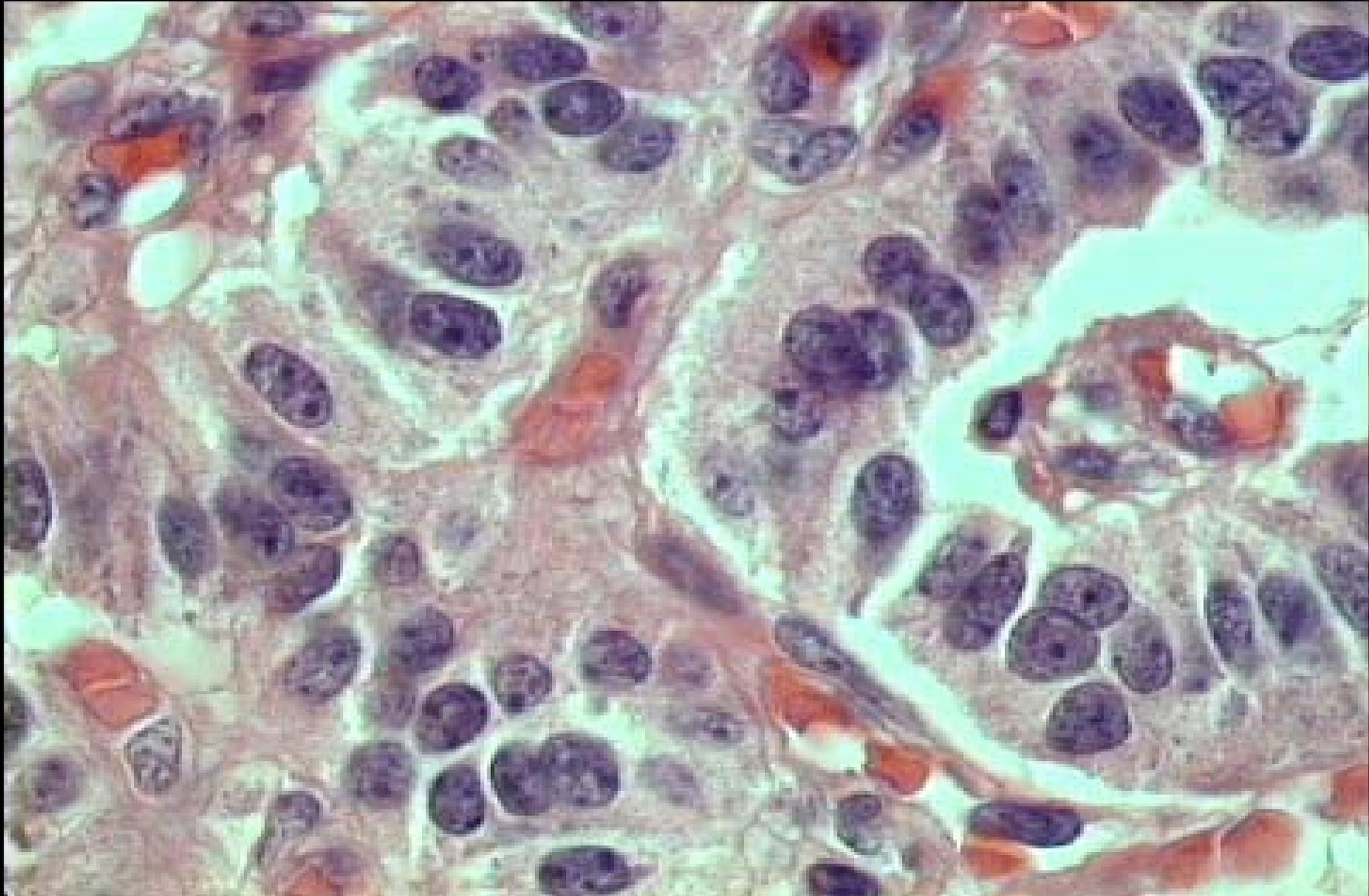
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Close up on the peripheral subcapsular area



Close up on the central area; Anastomosing cords and ribbons of cuboidal cells. Fixation artifact discloses clear spaces around a rich capillary mesh.

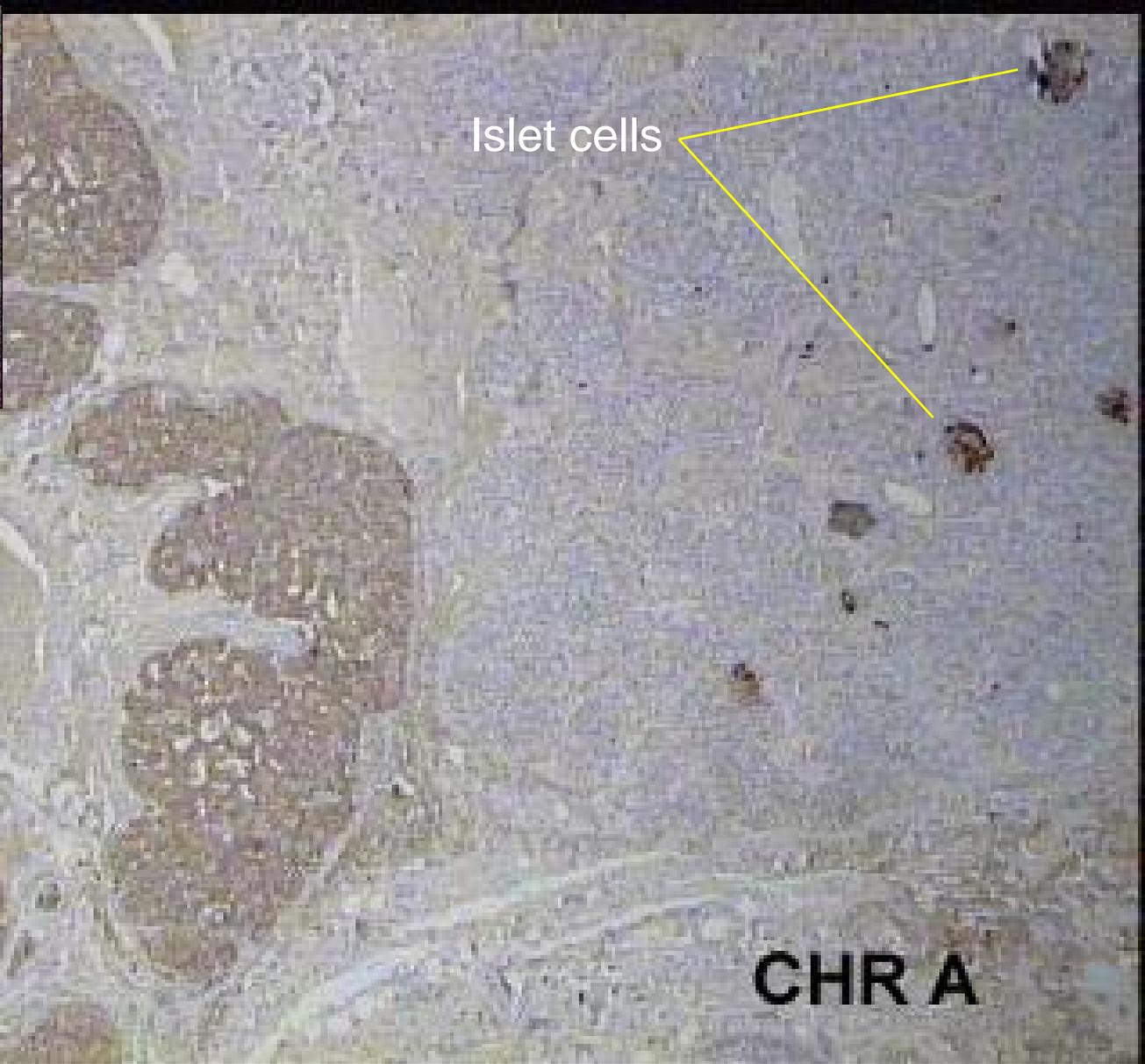


In all areas of the nodule, there is absence of atypia or mitotic figures.

MAIN MENU



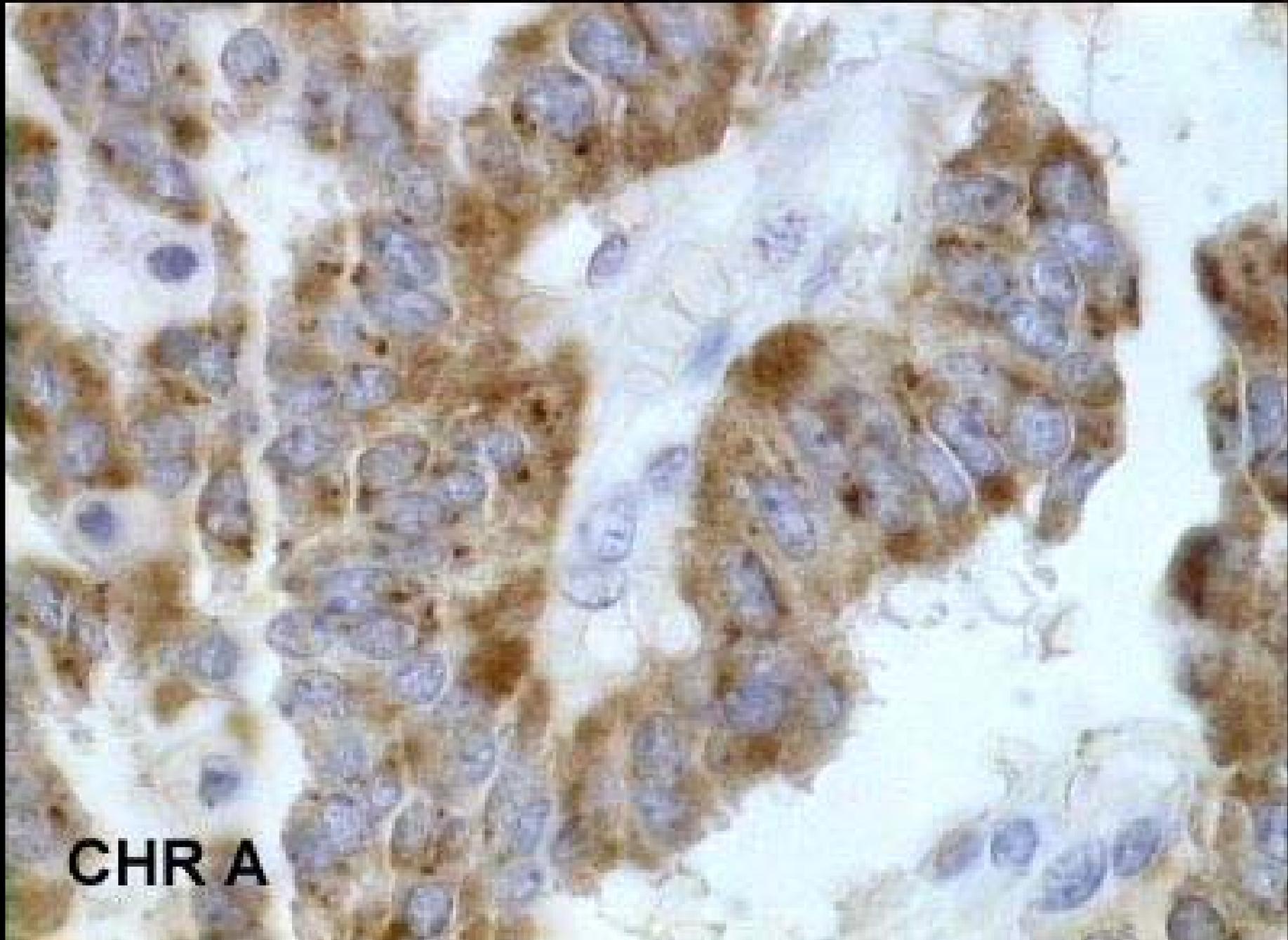
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Islet cells

CHR A

Panendocrine marker, scanning view, with a positive internal control (islet cells)



CHR A

MAIN MENU



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Diagnosis:

- Islet cell tumor, consistent in the clinical setting with an **insulinoma** (beta-cell tumor).
- Peripancreatic lymphnodes submitted, were negative.



Beta cell tumors (insulinomas)

- Insulinomas constitute the **more common and better known variety** of functioning islet cell tumors. Most occur in adults, and the incidence is equal in both sexes. Only a minority of the patients (less than 10%) are affected by MEN I. The **Whipple triad** thought to be characteristic of this tumor consists of (1) mental confusion, weakness, fatigue, and convulsions; (2) fasting blood glucose levels below 50 mg%; and (3) relief of symptoms by the administration of glucose. **Intravenous tolbutamide** administration and determinations of circulating insulin levels are useful tests for the diagnosis of insulinomas. Circulating proinsulin-like material can be detected in the serum. **Celiac arteriography** is extremely useful in the localization of these neoplasms, which may otherwise prove quite elusive at surgical exploration. The reported success rate of angiographic localization is between 60% and 75%. A newly developed and highly sensitive technique is that of **endoscopic ultrasonography**, which in one series was often successful in locating the tumor in cases in which angiography and CT had failed.

- Over 90% of beta cell tumors are **solitary**. Practically all of them are located within the substance of the pancreas; only 2% of the cases are seen in adjacent areas, such as the duodenal wall. **About 70% of them measure 1.5 cm or less**, sometimes as little as 3 or 4 mm. Microscopically, the tumor may grow in a solid or gyriform pattern; glands are usually absent.
- Immunohistochemically, there is reactivity for **insulin**, although this is usually of a lesser degree than in the normal beta cells. Beta cell tumors show a **lesser degree of reactivity for chromogranin** than other islet cell neoplasms.
- Only **7% to 10% of beta cell tumors are malignant** if the criteria of infiltration and/or metastases are adhered to. In general, the malignant variety is associated with a shorter history and more pronounced hypoglycemia. Neither conventional morphology nor DNA ploidy patterns are good predictors of behavior.

- The treatment of beta cell tumors is **surgical**. If the diagnosis of hyperinsulinism is reasonably certain and surgical exploration fails to reveal a neoplasm, a **subtotal pancreatectomy is justified**. Of thirty-three patients reported by a study in whom this operation was performed, the tumor was found in the resected specimen in fifteen.
- **Extrapancreatic neoplasms** may rarely be associated with hypoglycemia that disappears on removal of the tumor. Most have been liver cell carcinomas or mesenchymal neoplasms. The latter have received diagnoses such as leiomyosarcoma, hemangiopericytoma, and fibrosarcoma, but the evidence strongly suggests that the large majority have the morphologic features of either **benign or malignant solitary fibrous tumor**. Most are found in the pleura or retroperitoneum, and they tend to attain a very large size.