Case #56

NAME Educational Activities Committee

Case provided by:

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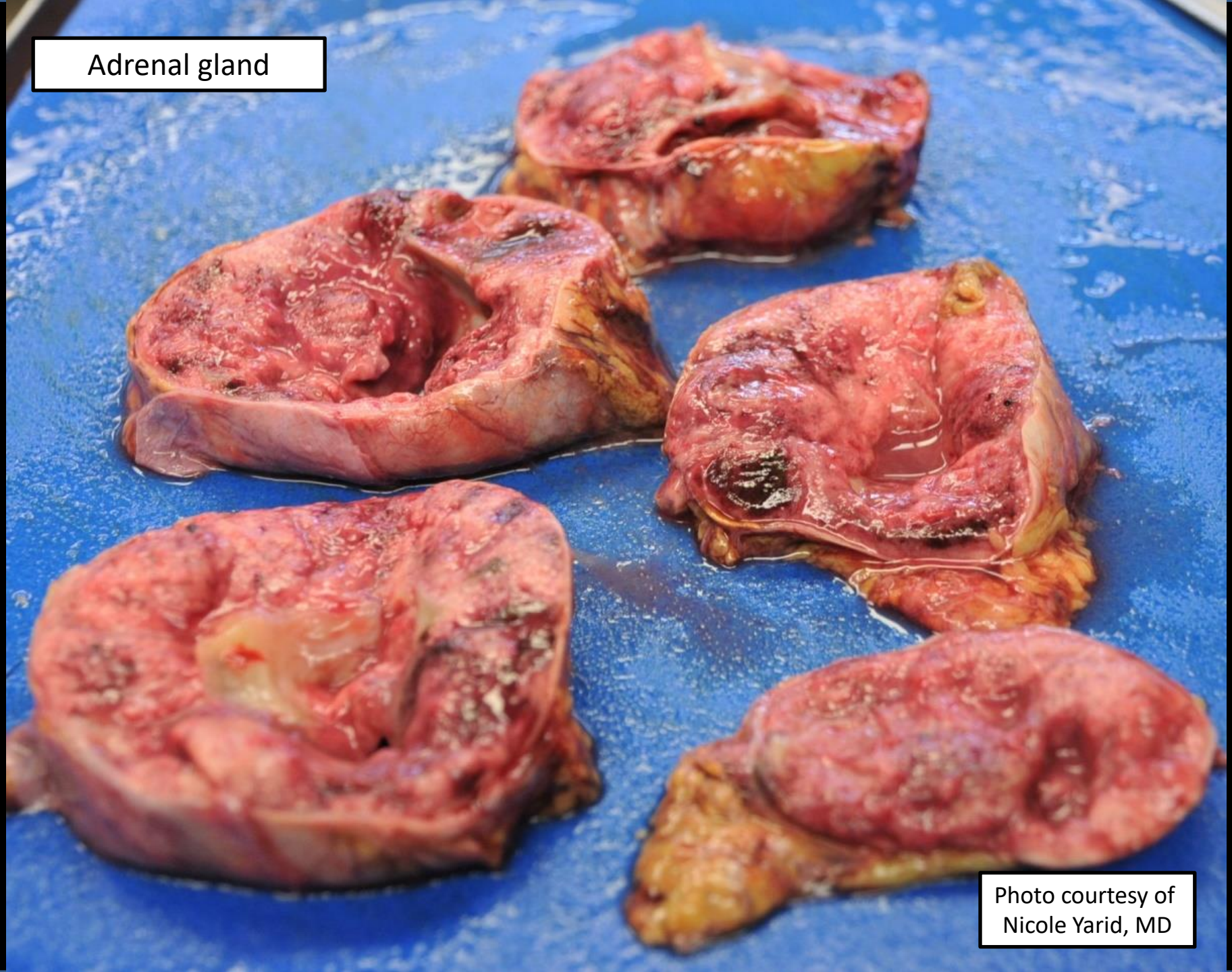
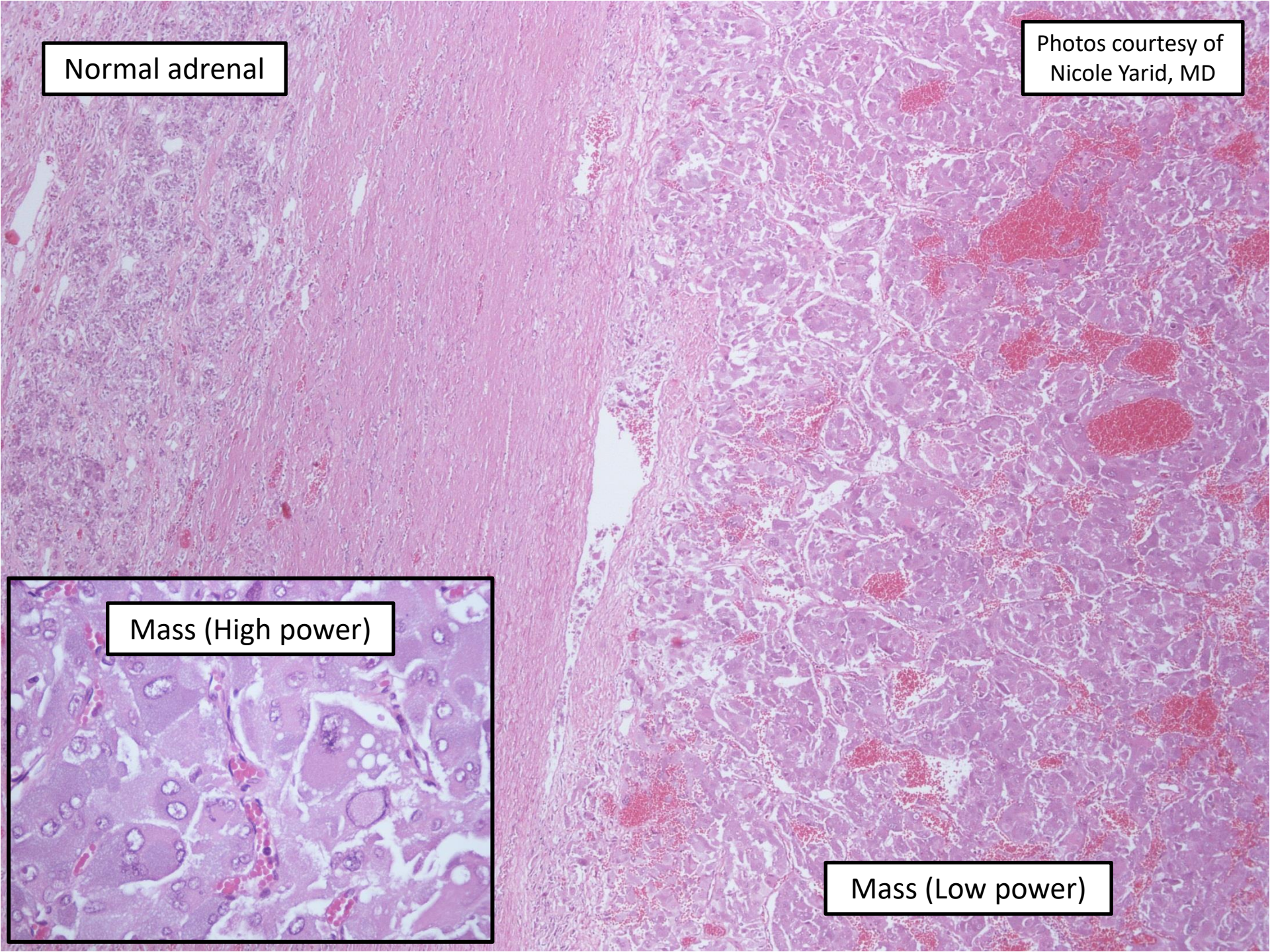

Adrenal gland
Photo courtesy of
Nicole Yarid, MD


Normal adrenal
Photos courtesy of Nicole Yarid, MD
Mass (High power)
Mass (Low power)

A 30-year-old male with neurofibromatosis type 1 and hypertension went to the dentist for an extraction. Upon administration of a local anesthetic (lidocaine and epinephrine) he became acutely short of breath, coughed up pink froth, and complained of abdominal pain. Hospital CT showed acute respiratory distress syndrome, pulmonary hemorrhage, “tooth” in airway, and a large adrenal mass. Autopsy revealed massive acute myocardial infarction and the adrenal mass above. What is the diagnosis?

A. Adrenocortical carcinoma

B. Pheochromocytoma/paraganglioma

C. Metastasis

D. Plexiform neurofibroma

E. Myelolipoma

ANSWER...

B. Pheochromocytoma/paraganglioma (CORRECT ANSWER, 79.13% of answers)

The clinical history and autopsy findings are most consistent with pheochromocytoma/paraganglioma (PPGL). PPGLs are neuroendocrine tumors derived from chromaffin cells of the adrenal medulla (called pheochromocytomas) or from the sympathetic or parasympathetic autonomic ganglia (paragangliomas). These tumors account for up to 0.6% of cases of adult hypertension and NF1 carries an increased risk of PPGLs. PPGLs are well-circumscribed and unencapsulated with cut surfaces that are tan to red with areas of hemorrhage. Microscopically, they are composed of large, polygonal, uniform or extensively vacuolated cells in a nested (zellballen), trabecular, or solid arrangement. Other less common cardiovascular manifestations such as arrhythmias, angina pectoris, acute myocardial infarction, dilated cardiomyopathy, acute heart failure, and cardiogenic shock have occasionally been reported with PPGLs. Therefore, this tumor was deemed to contribute to the patient's cause of death.

Other Responses

A. Adrenocortical carcinoma (14.31% responses)

Adrenocortical carcinoma (ACC) has a bimodal distribution, with the first peak in children <5 years and the second peak during the $4^{th}/5^{th}$ decades of life. Adult ACC can also be associated with virilization alone or a mixed syndrome (virilization and Cushing's syndrome). Macroscopically, ACC are typically solitary, bulky and yellow-tan or red-brown with a median size of 10 - 12 cm. The cut surfaces are heterogeneous with areas of necrosis and hemorrhage. Microscopically, these are encapsulated tumors composed of large, often pleomorphic cells with granular clear to eosinophilic cytoplasm, arranged in variably sized nests, large sheets, and trabeculae. While the macro- and microscopic characteristics are akin to pheochromocytoma, the clinical history of hypertension, cardiogenic shock, and NF1 are not consistent with ACC. Immunohistochemical stains can also differentiate between the two entities but were not performed in this case.

C. Metastasis (2.58% responses)

Adrenal glands are the fourth most common site of metastases in malignant disease and metastases are the most common malignant lesions involving the adrenal gland. Adrenal metastasis can occur from primaries in the lung (39%), breast (35%), melanoma, gastrointestinal tract, pancreas, and kidney among other places; therefore, metastases to the adrenal glands are heterogeneous lesions and frequently affect both glands. The lack of primary on autopsy makes metastasis unlikely, especially considering the other macroscopic and microscopic findings and decedent clinical history.

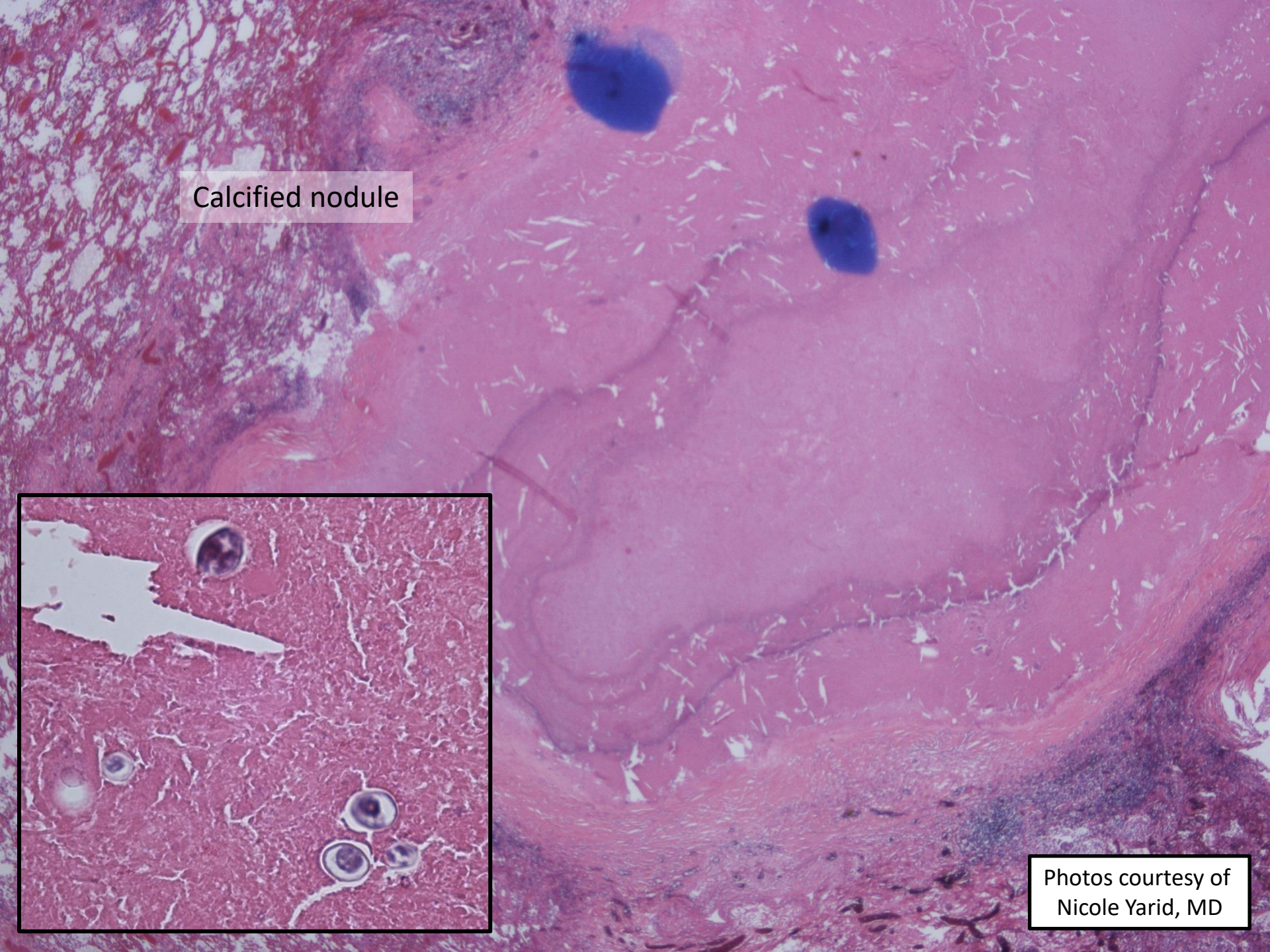
D. Plexiform neurofibroma (3.18% responses)

Plexiform neurofibroma (PN) is a benign peripheral nerve sheath tumor that surrounds multiple nerve fascicles giving them their characteristic irregularly thickened, distorted, and tortuous structure. As opposed the adrenal tumor shown, these are typically large tumors attached to major nerve trunks in the neck or extremities. They typically form in childhood and are part of diagnostic criteria for neurofibromatosis type 1. Microscopically, these are hypocellular spindle cell tumors with a myxoid background.

E. Myelolipoma (0.8% responses)

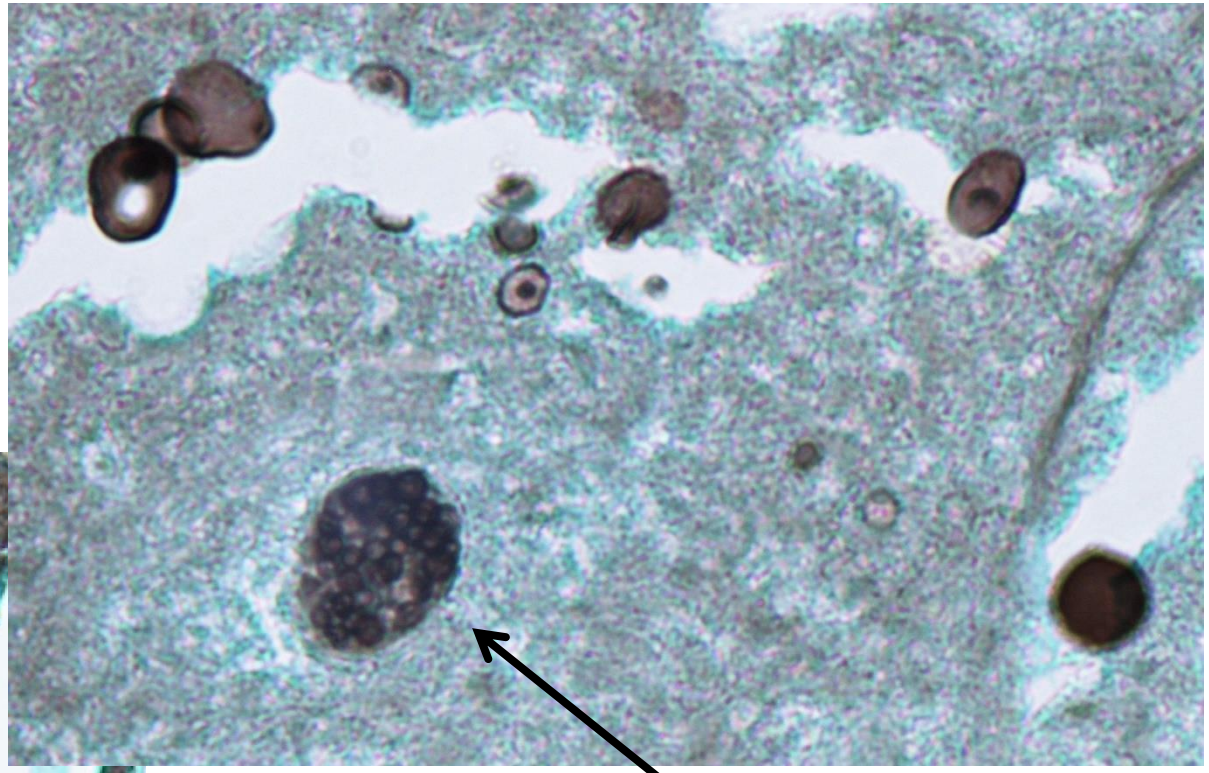
These are nonfunctional tumors typically seen in the fifth to seventh decade of life. They are benign and usually unilateral tumors composed of hematopoietic precursor cells and mature fat. Larger tumors may have hemorrhage, necrosis, calcification, and cysts; however, the microscopic findings and decedent clinical history in this case are incompatible with the diagnosis of myelolipoma.

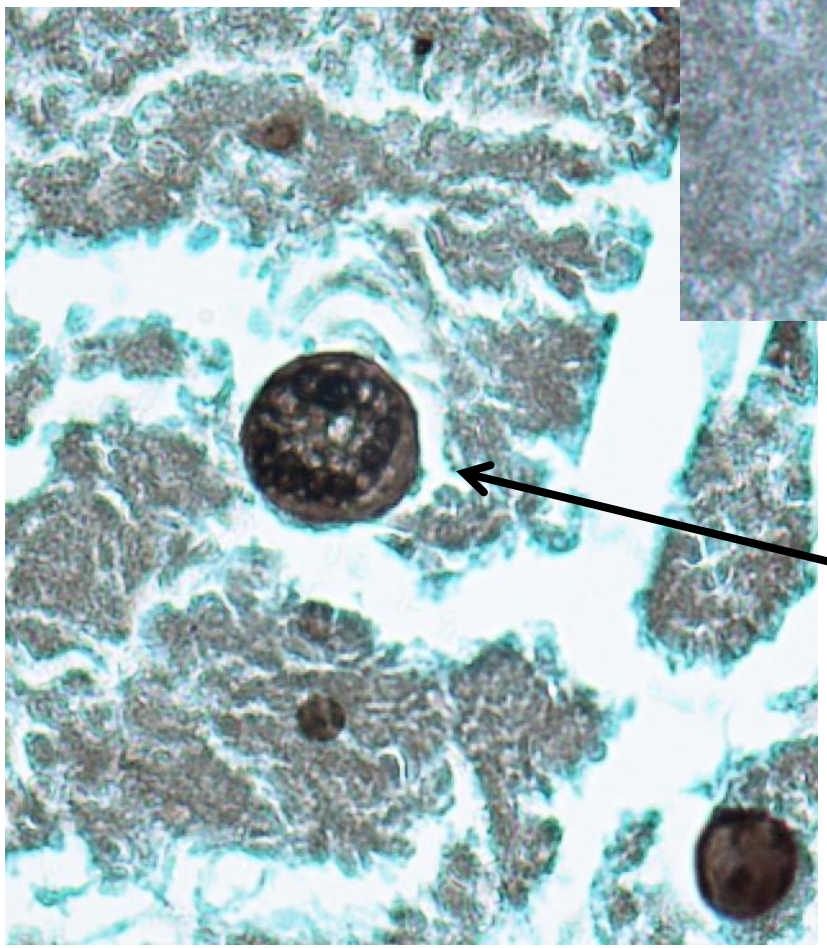
Other fun autopsy finding:
"Tooth" seen on CT turned out to be a calcified nodule….


Calcified nodule
Photos courtesy of
Nicole Yarid, MD

Photos courtesy of Nicole Yarid, MD

GMS stain highlights fungal organisms!





Morphology consistent with Coccidioides spherules with endospores

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