Tumor Localization in Ectopic Cushing Syndrome Using Combined PET/CT Imaging

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Abstract: Diagnosis of ectopic Cushing syndrome is challenging. The best imaging approach for localizing ectopic ACTH-secreting tumors is not defined. Here, we report on a 68-year-old woman with new-onset hypertension (>200/90 mm Hg) who was referred to our institution with suspicion of ectopic ACTH-secreting tumor for further work-up. Combined FDG PET/CT as a whole-body imaging modality revealed a neuroendocrine tumor of the pancreatic tail confirmed by surgical exploration.

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Conflicts of interest and sources of funding: none declared.

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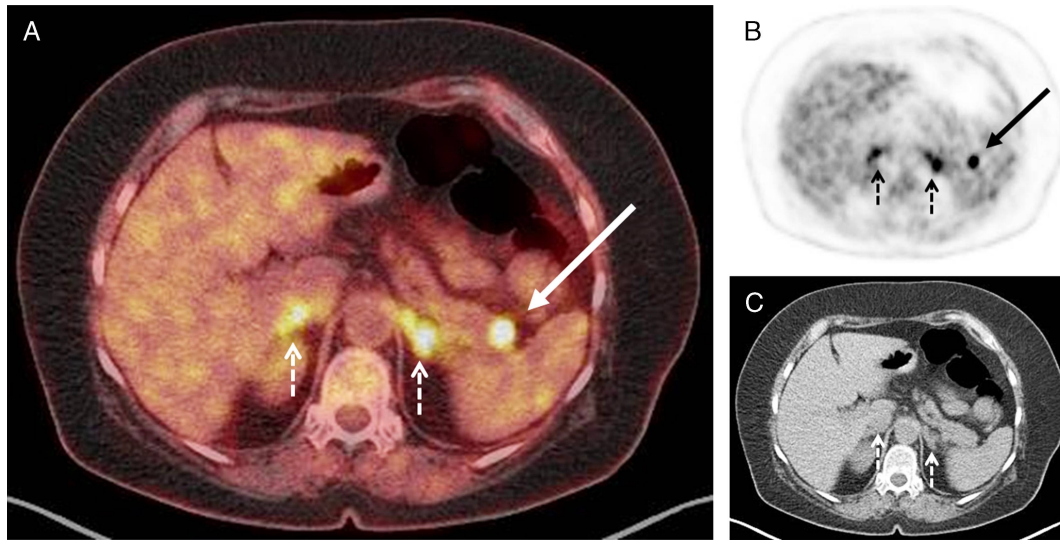

FIGURE 1. Here, we report on a 68-year-old woman with new-onset hypertension (>200/90 mm Hg) who was referred to our institution with suspicion of ectopic ACTH syndrome (EAS) for further work-up. ACTH and cortisol levels were highly increased; the dexamethasone suppression test was negative. Sinus petrosus catheterization confirmed EAS. No ectopic source was detected by CT and MRI. Combined ^{18}F-FDG PET/CT (**A**) as a whole-body imaging modality revealed an intense hypermetabolic focus in the pancreatic tail (*arrows*, **A** and **B**). Additionally, due to continuous ACTH stimulation, both hypertrophic adrenals showed reactive FDG uptake (*dotted arrows*, **A**–**C**). Surgical exploration was performed and revealed a 9-mm-sized, well-differentiated neuroendocrine tumor of the pancreatic tail with ACTH production which was removed in toto. Serum ACTH levels normalized within 24 hours.

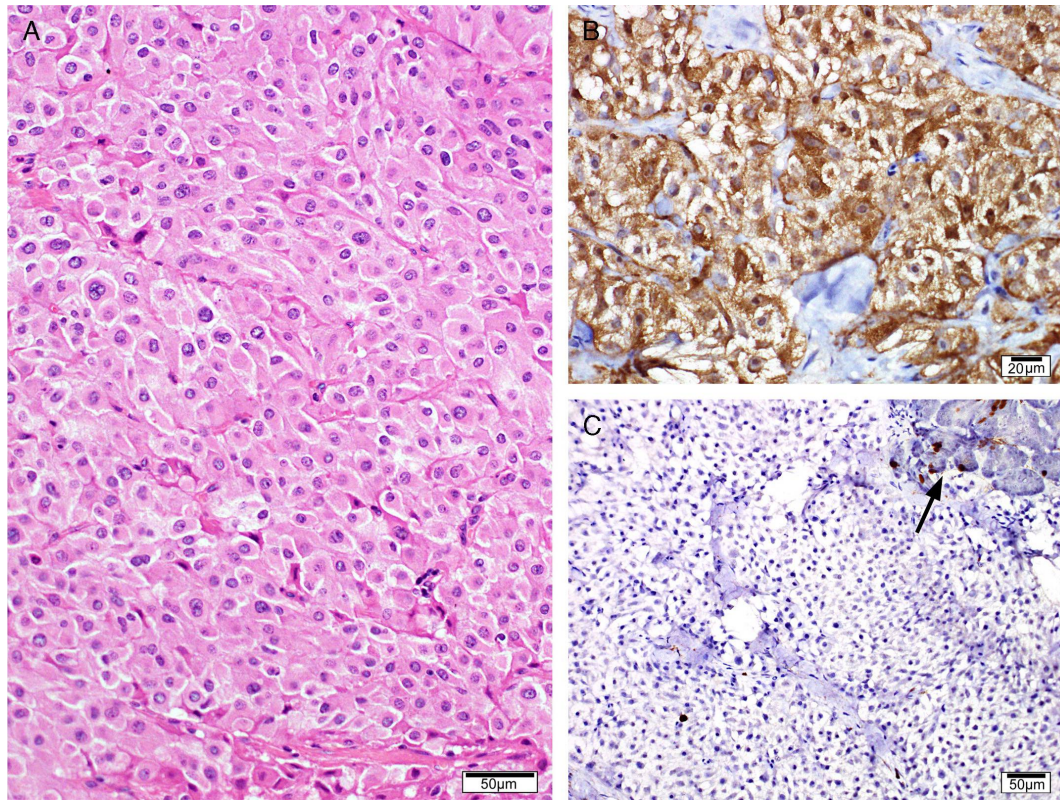

FIGURE 2. Histological examination revealed a well-differentiated tumor composed of monomorphic cells with broad eosinophilic cytoplasm arranged in solid nests (**A**). Ectopic ACTH production of the tumor was confirmed by immunohistochemistry (**B**) and staining for MIB-1 demonstrated a very low proliferation index in comparison with residual pancreas parenchyma (*arrow*, **C**). Cushing syndrome (CS) is the result of excess of endogenous or exogenous glucocorticoids. Clinical symptoms include hypertension, obesity, glucose intolerance, or muscle weakness.[1–3] Establishing the diagnosis is challenging since none of these symptoms is characteristic for CS. Endogenous CS is most frequently due to pituitary ACTH hypersecretion.[4] ACTH-dependent CS may also be caused by ectopic ACTH syndrome (EAS) which results in 50% from small-cell lung carcinomas.[5] Other types of cancer include carcinoid tumors, islet cell tumors, pheochromocytomas, and medullary thyroid carcinomas.[6] Diagnostic workup for ACTH-dependent CS comprises high-dose dexamethasone suppression test and corticotropin-releasing-hormone stimulation test.[4] In addition, blood sampling from the inferior sinus petrosus is performed.[1] The best imaging approach for localizing ectopic ACTH-secreting tumors is not defined.[7–10]