IMAGE OF INTEREST

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A rare adrenocorticotropic hormone-producing nasal neuroendocrine tumor with multiple metastasis

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A 72-year-old woman who had experienced nasal stuffiness, peripheral edema, abdominal discomfort, and distension for 1 month visited our hospital. She had been diagnosed with diabetes mellitus before 3 months and hypertension, 4 years previously. The patient was taking metformin, vildagliptin for diabetes and candesartan for hypertension.

In a physical examination, she had red cheeks, buffalo hump, and thin arms and legs. The laboratory data revealed hypokalemia (1.99 mEq/L), high urine potassium (27.4 mEq/L), and metabolic alkalosis (pH 7.61, bicarbonate 43.7 mmol/L). Since the random cortisol level increased to 66.38 μg/dL and adrenocorticotropic hormone (ACTH) was extremely increased (1,111.1 pg/mL), sella turcica magnetic resonance imaging was performed to assess ACTH-dependent Cushing's syndrome.

Notably, a mass that occupied both nasal cavities and extended to both inferior frontal lobes was identified (Fig. 1A-1C), and the pituitary gland was intact. A biopsy was performed on the mass in the nasal cavity, and well-differentiated tumor cells with abundant eosinophilic cytoplasm and small hyperchromatic nuclei were observed in the focal lesions of the mass (Fig. 2A and 2B). Most of the cells in the mass were positive for chromogranin (Fig. 2C) and sysnaptosphysin (Fig. 2D), a specific marker for neuroendocrine tumors (NETs), as well as ACTH (Fig. 2E). Very low tumor cells expressed Ki-67 (Ki-67 index was < 1%) (Fig. 2F) and suggested the low-grade NET and the patient was diagnosed with Cushing's syndrome due to ectopic ACTH syndrome (EAS) by a NET of the nasal cavity. Fluorodeoxyglucose positron emission tomography (FDG-PET/CT) was performed to assess the stage of the NETs, and the PET/CT images revealed advanced NETs with multiple hypermetabolic lesions in the nasal cavity, cribriform plate, right retropharyngeal and left neck level II lymph nodes, both adrenal glands, multiple nodules in both lungs, and cecum (Fig. 1D-1J).

Although a few reports of extraadrenal ACTH-producing NET is exist, Cushing's syndrome due to ACTH-secreting NETs in the nasal cavity is extremely rare, and to our knowledge, there was no case of nasal NET with advanced stage with multiple metastasis. Although extremely rare, primary nasal NET should be considered as a cause of EAS, and we suggest that rigorous checks of the whole body to be performed for patients with NETs with low Ki-67, due to the risk of multiple metastases.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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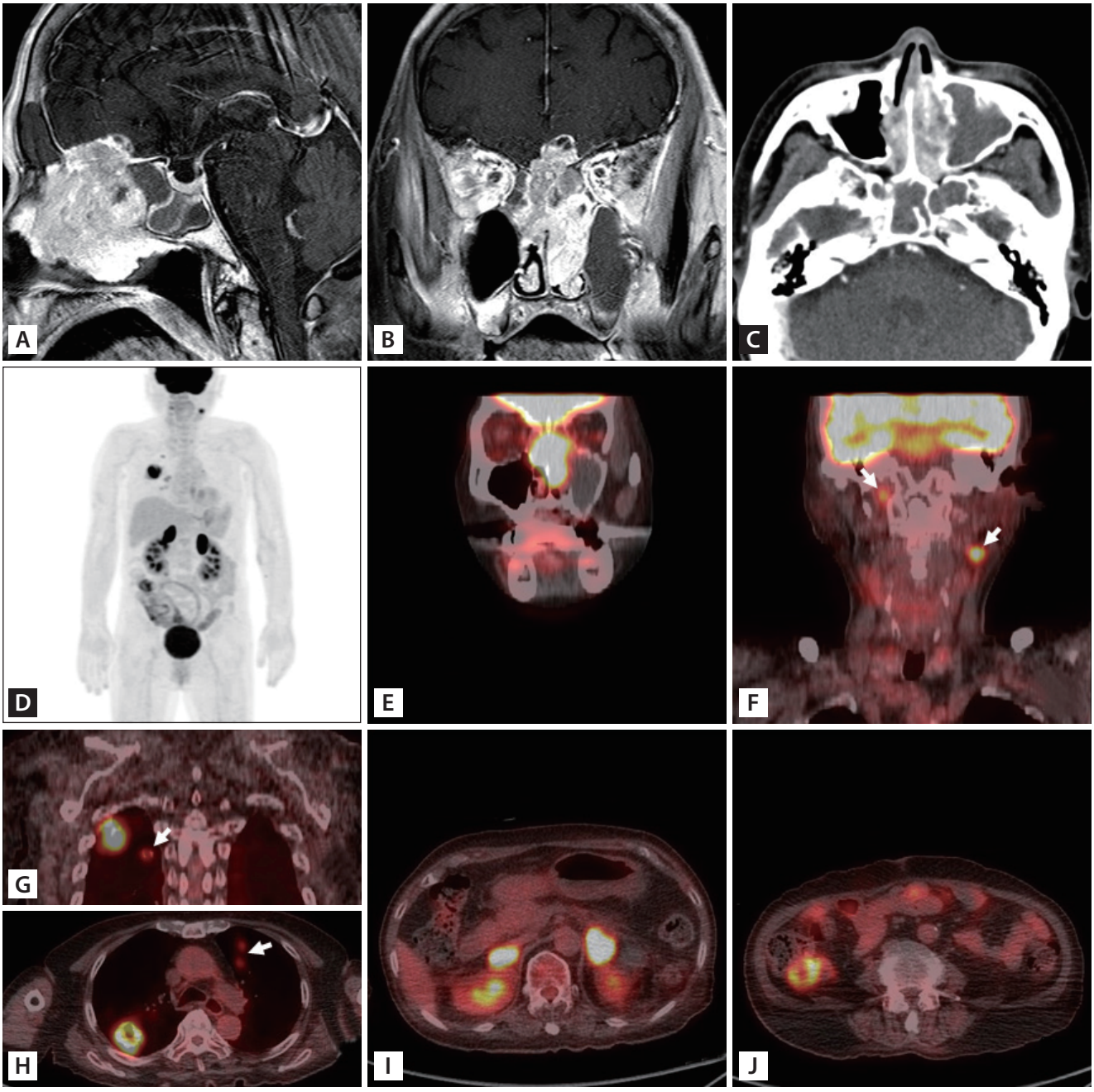

Figure 1. The representative radiologic (magnetic resonance imaging) and 2-[fluorine-18]-fluoro-2-deoxy-D-glucose-position emission tomography/computed tomography (FDG-PET/CT) images. A 4 × 3 × 3 cm heterogeneously enhanced mass occupying both sinonasal cavities with intracranial extension to the anterior cranial fossa. It destroyed the ethmoid bone structure and obstructed the left osteomata unit and pansinusitis (A, B, C). Representative coronal PET/CT images revealed intense FDG uptake at multiple sites (D), including the nasal cavity (E), lymph nodes (F), lung (G, H), adrenal gland (I), and cecum (J). Arrrows indicate the FDG uptake in the lymph node in the right retropharyngeal and level II (F) and in the multiple lung nodules (G). FDG uptake was observed in the tumor in the nasal cavity and cribriform plate.

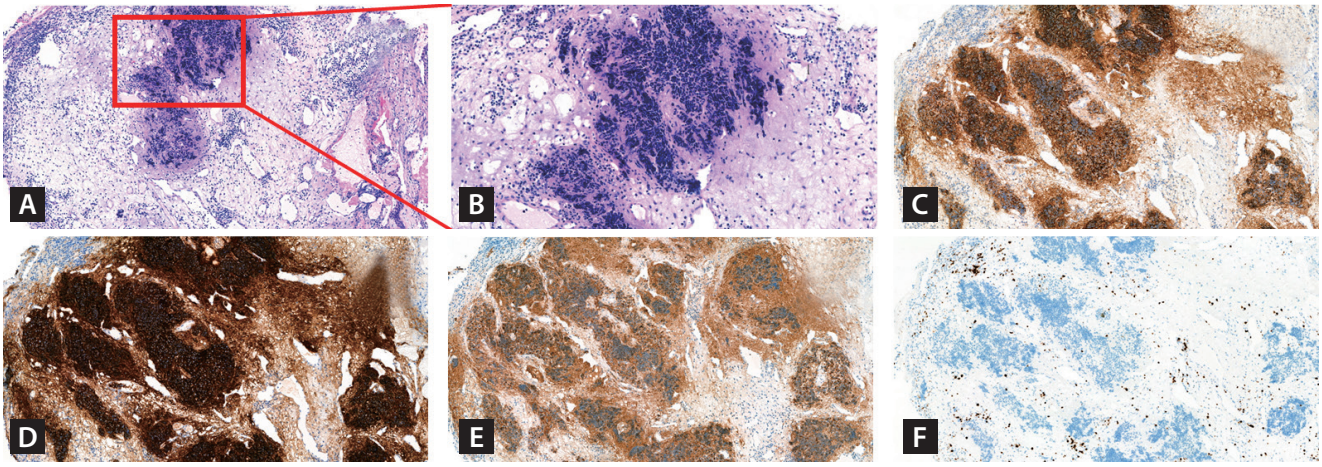

Figure 2. Pathologic images from the tumor in the nasal cavity. The tumor cells (red circle) seen at ×200 magnification were focally observed (A) and at ×400 magnification, well- differentiated small round cells with abundant eosinophilic cytoplasm and small hyperchromatic nuclei were observed (B). At ×400 magnification, the neuroendocrine tumor cells were strongly immuno-positive for chromogranin (C), synaptophysin (D), and adrenocorticotropic hormone (E). The proliferating cells were immuno-positive for Ki-67 stain (F) and Ki-67 index was extremely low; 1%.

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