AACE Clinical Case Rep. 9 (2023) 25–26

Contents lists available at ScienceDirect

AACE Clinical Case Reports

journal homepage: www.aaceclinicalcasereports.com

Visual Vignette

Abdominal Pain, Progressive Weakness, and Weight Loss in an 83-Year-Old Man

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ARTICLE INFO

Article history:
Received 10 August 2022
Received in revised form
9 September 2022
Accepted 13 September 2022
Available online 19 September 2022

Case Presentation

An 83-year-old man presented to the emergency room with nonspecific abdominal pain, and a subsequent computed tomography (CT) scan revealed bilateral solid adrenal masses measuring 5.5cm on the right and 2.7cm on the left. The average noncontrast Hounsfield unit of adrenal lesions was 32 HU. One month later, a positron emission tomography–CT scan showed hypermetabolism and interval growth of lesions, measuring 6.5cm on the right and 3.9cm on the left, with extensive hypermetabolic retroperitoneal lymphadenopathy (Fig. 1). Follow-up was delayed by the patient; in the subsequent 4 months, he experienced progressive weakness, impaired ambulation, abdominal pain, persistent nausea and vomiting, and weight loss (20 lbs). Upon hospital admission, his baseline 8 AM cortisol and adrenocorticotropic hormone levels were 6.2 mcg/dL and 357.0 pg/mL, respectively. He underwent adrenocorticotropic hormone–stimulation testing, which showed a peak serum cortisol level of 7.3 mcg/dL. The levels of plasma aldosterone, renin activity, plasma normetanephrine, and plasma metanephrine were

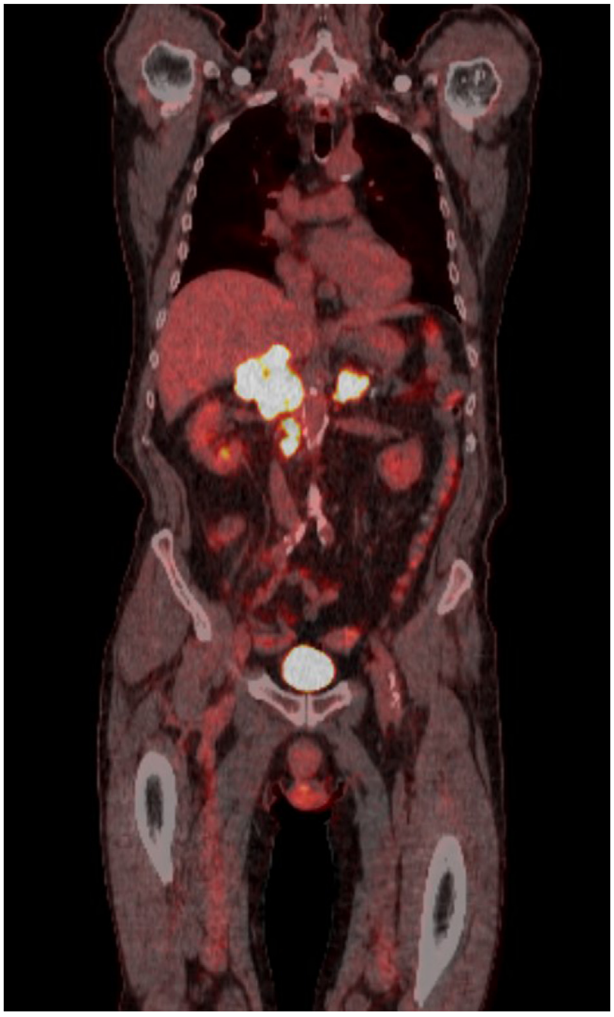

Fig. 1.

Abbreviations: CT, computed tomography; HU, Hounsfield unit; lbs, pounds; PAL, primary adrenal lymphoma; VHL, Von Hippel-Lindau; RET, rearranged during transfection; SDHB, succinate dehydrogenase complex subunit B; RCHOP, rituximab, cyclophosphamide, doxorubicin hydrochloride (hydroxydaunorubicin), vincristine sulfate (Oncovin), and prednisone.

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https://doi.org/10.1016/j.aace.2022.09.004
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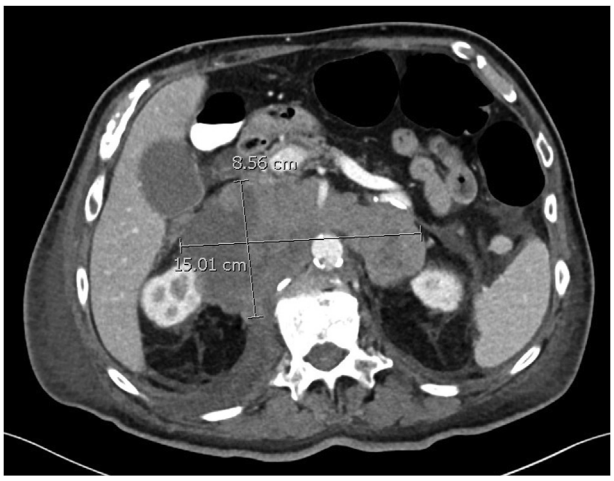

Fig. 2.

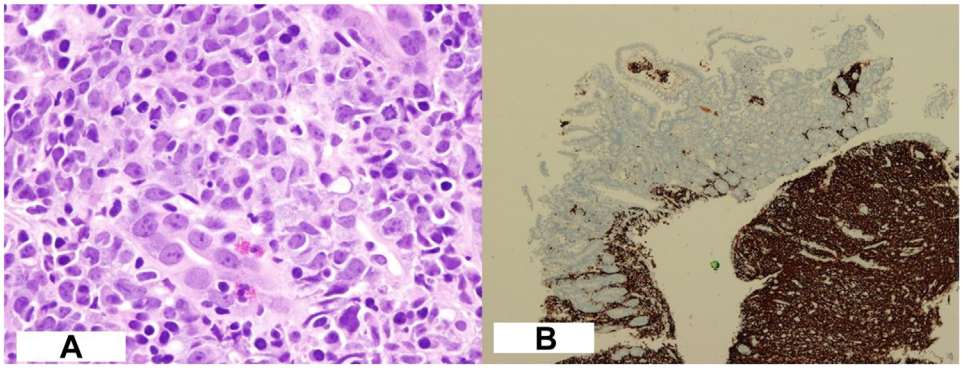

Fig. 3.

<1.0 ng/dL (0.0-30.0), 1.102 ng/mL/h (0.167-5.380), 35.6 pg/mL (0.0-297.2), and <10.0 pg/mL (0.0-88.0), respectively. His dehydroepiandrosterone sulfate level was 13.5 mcg/dL (20.8-226.4). He also developed hypercalcemia, with a parathyroid hormone level of 9.7 pg/mL (15-65) and a 1,25-OH_2-vitamin D level of 108.0 pg/mL (19.9-79.3). A repeat CT scan showed further interval growth and confluence of his adrenal lesions, with inferior vena cava displacement and compression of the second part of his duodenum (Fig. 2). An esophagogastroduodenoscopy performed for melena revealed a 3.0-cm friable ulcer in the second part of the duodenum—where the right adrenal mass was compressing—and a mucosal biopsy revealed diffuse neoplastic cells with nuclear pleomorphism, prominent nucleoli, and mitotic figures with positive CD20 immunohistochemical staining (Fig. 3 *A* and *B*).

What is the diagnosis?

Answer

Bilateral primary adrenal lymphoma (PAL) and associated primary adrenal insufficiency. PAL is very rare, accounting for 1% to 4% of all non-Hodgkin lymphomas.[1] PAL typically presents with abdominal pain and symptoms of adrenal insufficiency; up to 70% of cases are bilateral.[1,2] PAL can be distinguished biochemically from adrenocortical carcinoma or metastatic pheochromocytoma because these entities do not cause adrenal insufficiency. Moreover, adrenocortical carcinoma typically presents as a large, heterogeneously enhancing unilateral mass, and up to 60% of these have excess steroid production.[2] Metastatic pheochromocytoma can present bilaterally, most often affiliated with familial syndromes, such as *VHL*, *RET*, *SDHB*, or other genetic mutations. Classically, pheochromocytoma has marked heterogeneous contrast enhancement.[3] Lastly, one clue to the ultimate diagnosis was hypercalcemia with an elevated 1,25-OH_2-vitamin D level, indicating upregulation of 1α-hydroxylase. The patient was started on steroid replacement and is currently receiving acalabrutinib and RCHOP therapy (rituximab, cyclophosphamide, doxorubicin hydrochloride (hydroxydaunorubicin), vincristine sulfate (Oncovin), and prednisone). This case highlights the importance of early recognition and diagnosis of adrenal insufficiency secondary to adrenal lymphoma.

Disclosure

The authors have no multiplicity of interest to disclose. The views expressed in this article are those of the authors and do not reflect the official policy of the Department of Army/Navy/Air Force, Department of Defense, or the U.S. Government.

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