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CASE REPORT

Extramedullary Hematopoiesis in Adrenal Incidentaloma: Case Report of an Atypical Histopathological Finding

Jorge Montalvo-Hernández[1] · José Alfredo Álvarez-López[2] · Rafael Silva[3] · Rafael Humberto Pérez-Soto[4]

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Abstract
Adrenal gland extramedullary hematopoiesis is extremely rare, and when present, imaging studies may mimic an incidentaloma suspicious of malignancy. We present a 55-year-old woman without a history of hematologic disorder or neoplasms. There was an incidental finding of a 3.2 × 2.4-cm heterogenous adrenal mass and enhanced by contrast abdominal CT scan. Suspicious of malignancy was established, and laparoscopic left adrenalectomy was performed. Histopathological examination and immunohistochemical staining exhibited myeloid and erythroid precursors, as well as megakaryocyte confirming the diagnosis of extramedullary hematopoiesis in the adrenal gland.

Keywords Adrenal incidentaloma · Laparoscopic adrenalectomy · Extramedullary hematopoiesis

Introduction

Adrenal incidentaloma (AI) is becoming a more common adrenal disorder as a result of an increased detection in the last decades by the widespread use of abdominal ultrasound, computed tomography (CT), and abdominal magnetic resonance imaging (MRI). Assessments of functional status and malignancy risk are the two main objectives of preoperative workup for these patients. Extramedullary hematopoiesis (EMH) is the production of erythroid and myeloid progenitor cells outside the bone marrow as a compensatory mechanism, and in adults, it is typically seen in the clinical scenario of myeloproliferative neoplasms (leukemia/lymphoma), thalassemia, prolonged iron deficiency anemia, sickle cell disease, myelofibrosis, or polycythemia [1]. EMH occurs more frequently in the liver and spleen followed by the lymph nodes, lung, breast, small bowel, and thymus. Adrenal EMH is extremely rare, and when present, imaging studies may mimic an incidentaloma suspicious of malignancy.

Case Presentation

A 55-year-old woman with a history of primary hypothyroidism and hypertension was seen at the outpatient clinic complaining of chronic diarrhea and a 10-kg weight loss in

✉ Jorge Montalvo-Hernández
jorgemontalvo@yahoo.com

José Alfredo Álvarez-López
dr.alfredoalvarez@hotmail.com

Rafael Silva
rsilvaf82@gmail.com

Rafael Humberto Pérez-Soto
rafael.perez.dr@gmail.com

[1] Endocrine and Advanced Laparoscopic Surgery Service, Deparment of Surgery, North Central Hospital PEMEX, Calle Campo Matillas 52, San Antonio, Azcapotzalco, 02760 Mexico City, Mexico

[2] Clinical Endocrinology, North Central Hospital PEMEX, Calle Campo Matillas 52, San Antonio, Azcapotzalco, 02760 Mexico City, Mexico

[3] Deparment of Anatomical Pathology, North Central Hospital PEMEX, Calle Campo Matillas 52, San Antonio, Azcapotzalco, 02760 Mexico City, Mexico

[4] Endocrine and Advanced Laparoscopic Surgery Service, Deparment of Surgery, National Institute of Medical Sciences and Nutrition Salvador Zubirán, Vasco de Quiróga No. 15 Colonia Sección XVI, Tlalpan, 14080 Mexico City, Mexico

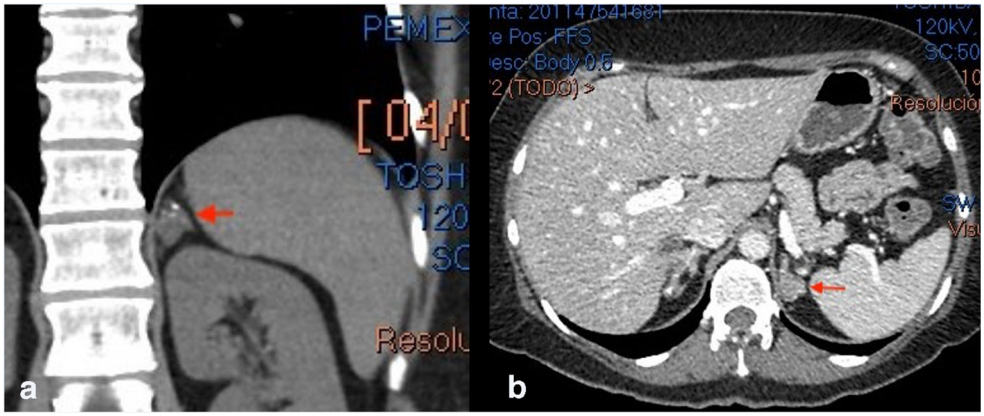

Fig. 1 Non-contrast coronal CT scan (**a**) shows a solid left adrenal incidentaloma (arrow) with calcifications inside. **b** Axial contrasted CT scan shows enhanced

the last 4 months. Physical exam was unremarkable. During her medical evaluation, an abdominal CT scan was performed with the incidental finding of a 3.2 × 2.4 cm of heterogeneous mass with small calcifications inside the left adrenal gland with a 28 HU in the non-contrast phase, enhancement in the arterial phase, and an absolute washout of 58% (Fig. 1). Biochemical workup showed hemoglobin 15.8 g/dL, hematocrit 47.2%, WBC count 9.8 × 10^9 G/L, neutrophils 70%, platelet count 167 × 10^9/L, serum glucose 100 mg/dL, serum sodium 141 mEq/L, serum potassium 3.8 mEq/L, TSH 1.01 μUI/mL, renin activity 6.1 ng/mL/h, aldosterone 275.3 pg/mL, PAC/PRA 4.5, a negative 1-mg-dexamethasone suppression test, 24 h of urine metanephrines, serum catecholamines, and vanillylmandelic acid between the normal ranges. A diagnosis of a non-functional left adrenal incidentaloma with imaging findings of suspicious of malignancy was established; laparoscopic left adrenalectomy was offered to the patient, which she consented. Surgery went uneventful through three ports, and the patient was discharged in postoperative day 2. Histopathological examination exhibited a 20-g adrenal gland, with normal cortex, hemorrhagic zones, and cystic degeneration of the medulla with myeloid, erythroid precursors, as well as megakaryocyte (Figs. 2 and 3). Immunohistochemical staining was positive for myeloperoxidase, CD117, and CD45 confirming the presence of bone marrow in the adrenal gland (Fig. 4).

Discussion

EMH in the adrenal gland is extremely rare with only a few cases described in the scientific medical literature. Despite

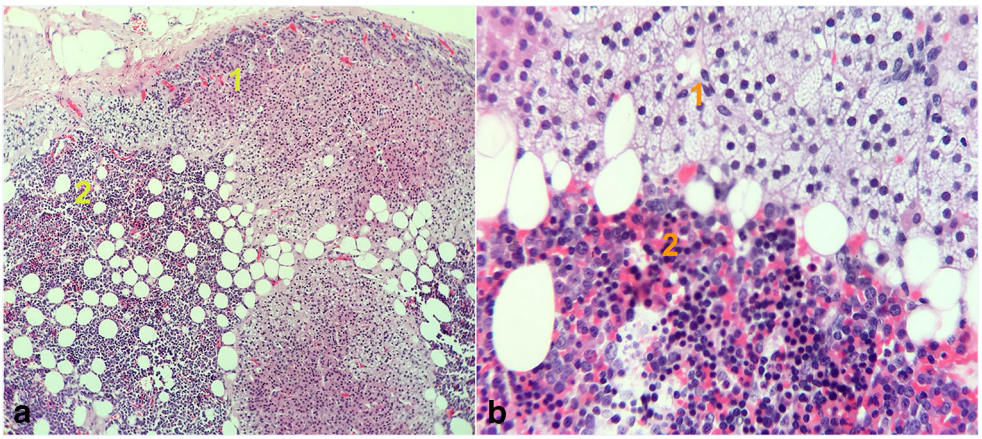

Fig. 2 **a** Hematoxylin and eosin staining photomicrography of normal adrenal cortex (1) and erythroid and myeloid precursors (2). **b** A × 4 magnified view

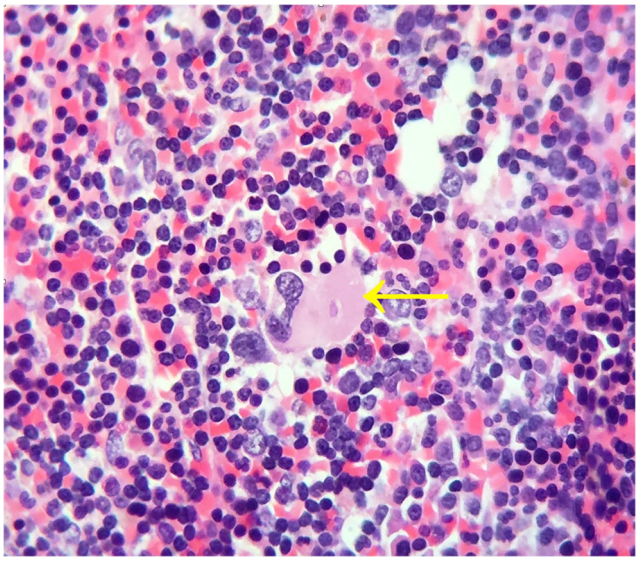

Fig. 3 A × 40 view of hematopoietic precursors; yellow arrow points a megakaryocyte

novel improvements in imaging acquisition, preoperative adrenal EMH identification by imaging studies remains a challenge, and, at this moment, there are no specific diagnostic findings. On abdominal CT scans, the majority of case reports describe a hypodense and heterogeneous adrenal mass with regular and well-defined borders, small calcifications, and enhancement after IV contrast administration [2]. In our case, we also performed a 12-min delayed CT protocol, obtaining an absolute washout of 58% that describes a vascularized mass and lipid-poor content. At MRI, adrenal EMH appears as a heterogeneous mass with variable signal intensity and enhancement on T1- and T2-weighted images. On T1- and T2-weighted images, internal fatty components show a hyperintense signal [3].

The vast majority of published cases have been diagnosed by histopathology, having undergone surgery [4, 5]. The presence of myeloid, erythroid precursors, as well as megakaryocyte (platelet precursors) is pathognomonic for EMH. We propose to add to the diagnostic approach of adrenal incidentaloma the EMH suspecting diagnosis in the clinical scenario of a hematologic disorder/neoplasms (with chronic anemia) and imaging characteristics previously discussed by CT and/or MRI. Diagnosis can be established through an image-guided adrenal biopsy with previous normal adrenal biochemical workup. Sekar et al. [6] from India achieved diagnosis by USG-guided biopsy in a beta-thalassemia patient and King et al. [7] from the USA through CT-guided biopsy in a myeloid metaplasia. Also, through functional imaging with technetium 99m-labeled sulfur colloid scintigraphy, it confirms the presence of bone-marrow elements achieved by Wat et al. [8]. All of them avoided surgery with success. A follow-up of these lesions should be the standard treatment in asymptomatic patients.

Unfortunately, preoperative diagnosis outside the previously mentioned clinical scenario is almost impossible, such as in our case. Asymptomatic patients with AI should entail a complete biochemical evaluation, focused on not only functional status (Cushing's, pheochromocytoma, and Conn's disease) but also a complete hemogram as blood disorders are the main comorbidities associated with EMH.

Although it is a rare finding, as cross-sectional imaging use is increasing, a rise in the detection and diagnosis of adrenal EMH is expected. Primary care physicians, radiologist, hematologist, endocrinologist, and endocrine surgeons should be aware of this possibility.

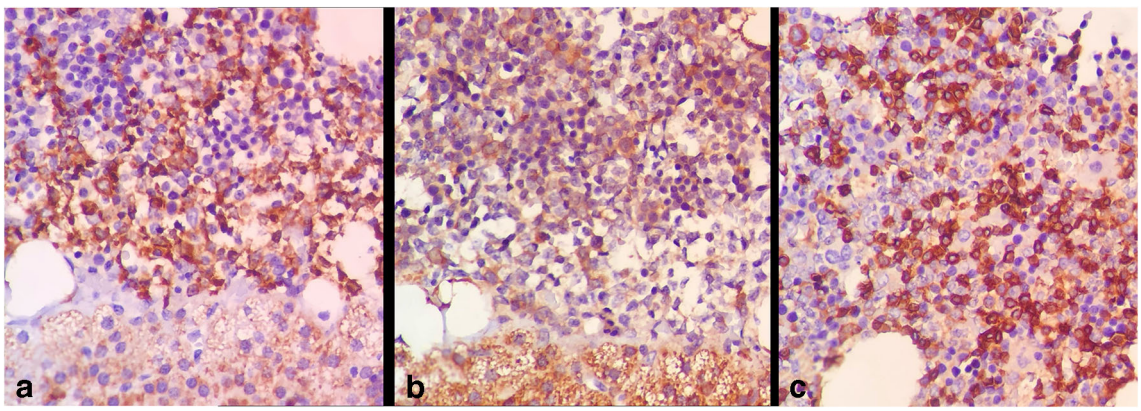

Fig. 4 Positive immunostains for myeloperoxidase (**a**), CD117 (**b**), and CD45 (**c**) confirm hematopoiesis tissue in adrenal gland

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

References

1. Fan F, Lavu S, Hanson CA et al (2018) Extramedullary hematopoiesis in the absence of myeloproliferative neoplasms: Mayo Clinic case series of 309 patients. Blood Cancer J 8(12):119
2. Salemme M, Rodella R, Fisogni S et al (2011) Nodular extramedullary hematopoiesis involving the adrenal gland. An uncommon cause of adrenal "incidentaloma". Pathologica 103:46–49
3. Sargar KM, Khanna G, Hulett BR (2017) Imaging of nonmalignant adrenal lesions in children. Radiographics 37(6):1648–1664
4. Kolev NH, Genov PP, Dunev VR et al (2020) A rare case of extramedullary hematopoiesis in adrenal mass. Urology Case Reports 30. https://doi.org/10.1016/j.eucr.2020.101120
5. Banerji JS, Kumar RM, Devasia A (2013) Extramedullary hematopoiesis in the adrenal: case report and review of literature. Can Urol Assoc J 7:E436–E438
6. Sekar S, Burad D, Abraham A et al (2015) Adrenal incidentaloma caused by extramedullary haematopoiesis: conservative management is optimal. BMJ Case Rep. https://doi.org/10.1136/bcr-2015-211014
7. King BF, Kopecky KK, Baker MK et al (1987) Extramedullary hematopoiesis in the adrenal glands: CT characteristics. J Comput Assist Tomogr 11:342–343
8. Wat NM, Tse KK, Chan FL et al (1998) Adrenal extramedullary haemopoiesis: diagnosis by a non-invasive method. Br J Haematol 100:725–727