Extramedullary Hematopoiesis Mimicking as an Adrenal Incidentaloma: A Case Report

Apanaree Bhekasuta* Phakthanidcha Lertweerasirikul** Pasathorn Potivongsajarn***
*,** Somdej Pranangchaosirikit Hospital, Chonburi Province
*** Rajavithi Hospital, Ministry of Public Health, Bangkok
* Corresponding Author: ploypds@gmail.com

Abstract

We report on a case of extramedullary hematopoiesis in a male in his 50s who presented with right adrenal incidentaloma from CT scan for right ureteric calculi. The patient denied having any symptoms of mass effect or functioning of adrenal tumor. The CT scan revealed a heterogeneous fatty density mass with minimal enhancement in the right adrenal region, measuring 9.9 x 8.8 x 10.7 cm. Open adrenalectomy was performed after exclusion of a functioning tumor. The pathology report confirmed massive extramedullary hematopoiesis in the fat tissue surrounding the normal adrenal gland. A hematologist was consulted to assess the cause of extramedullary hematopoiesis in this case, and no abnormal hematologic cause was found. This case highlights the need to consider extramedullary hematopoiesis as a rare differential diagnosis for adrenal masses, even in the absence of hematologic abnormalities, to guide accurate diagnosis and management.

Keywords: adrenalectomy, extra-medullary hematopoiesis, adrenal incidentaloma, case report, adrenal myelolipoma

Introduction

Extramedullary hematopoiesis (EMH) is a rare pathological process in which hematopoietic tissue develops outside the bone marrow, typically in response to chronic anemia or bone marrow dysfunction. It is most commonly found in the liver, spleen, lymph nodes, and paravertebral regions. Adrenal involvement is exceedingly rare, with only a few cases reported in the literature[1,2] EMH can mimic neoplastic processes, presenting as incidental masses or causing symptoms through mass effect. Accurate diagnosis is essential to avoid unnecessary treatments. We present a case of massive EMH surrounding a normal adrenal gland, emphasizing the importance of considering this rare diagnosis when evaluating adrenal masses with atypical imaging features.

Received: October 22, 2023; Revised: January 30, 2024; Accepted: February 3, 2024

Case Presentation

A 48-year-old Thai male presented at the internal medicine outpatient clinic at Somdej Pranangchaosirikit Hospital with a right adrenal incidentaloma discovered after a CT scan to evaluate a right ureteric stone. His underlying conditions included type II diabetes and dyslipidemia, which were routinely checked and controlled.

The patient had a history of hematuria but there was no history of abdominal pain, palpitations, flushing, excessive perspiration or unexplained weight loss or weight gain to suggest a functioning adrenal tumor. Clinical examination revealed no pallor or icterus, and his blood pressure was normal (130/84 mmHg). Abdominal examination showed no hepatosplenomegaly or other palpable mass.

Laboratory investigations revealed no anemia, a low mean corpuscular volume (60 fL), and a normal platelet count (239×10^9/L). An overnight 1 mg dexamethasone suppression test was done, and the result was suppressible. Plasma dehydroepiandrosterone sulfate (DHEAS), plasma aldosterone, plasma renin and plasma metanephrines were all within normal limits. A CT KUB scan found a fatty density mass with minimal enhancement at the right adrenal gland, measuring 9.9 x 8.8 x 10.7 cm in size, and a 1.6 cm stone at the right UPJ with mild right pyelectasis (Figure 1).

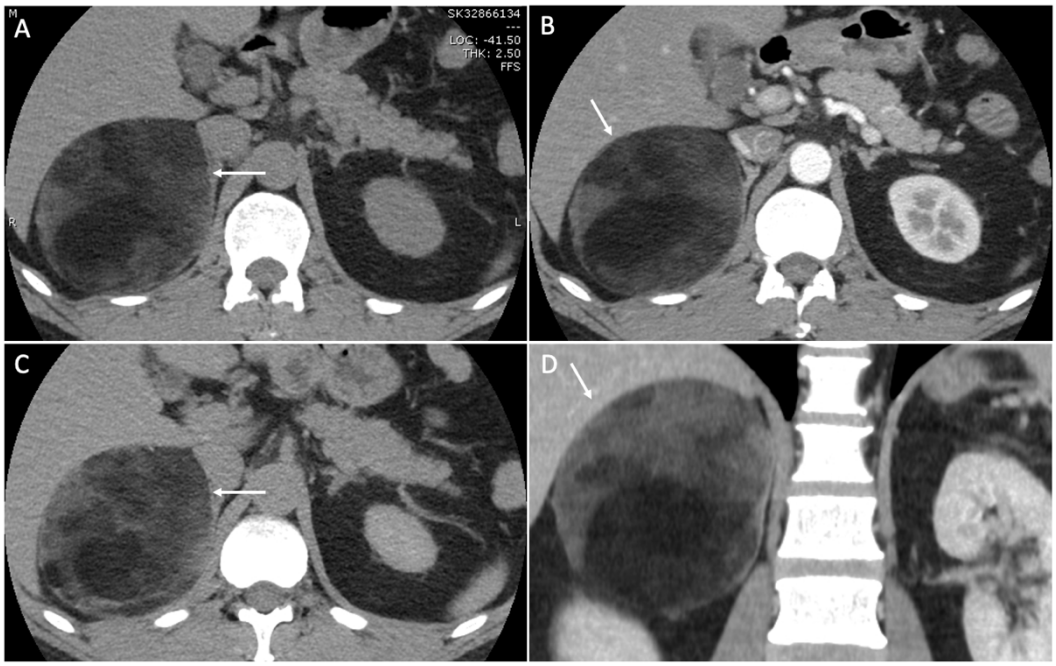

Figure 1. Axial precontrast (A), Arterial phase (B), Delayed phase (C), and Coronal delayed (D) CT images reveal a well-defined right adrenal lesion (Arrow).

After discussing with the patient the finding of a nonfunctioning adrenal incidentaloma causing pyelectasis, an open adrenalectomy was performed. Gross examination revealed a homogeneous, rubbery, fatty-like mass measuring 10 x 9.5 x 8 cm in size. The mass showed an area of hemorrhage (Figure 2). The remaining adrenal cortex showed no remarkable change. On microscopic examination, hematopoietic tissue consisting of numerous megakaryocytes, erythroid, and myeloid precursors, suggestive of extramedullary hematopoiesis surrounding the normal adrenal gland, was observed (Figure 3, 4). The final diagnosis was massive extramedullary hematopoiesis in the fat tissue surrounding the normal adrenal gland.

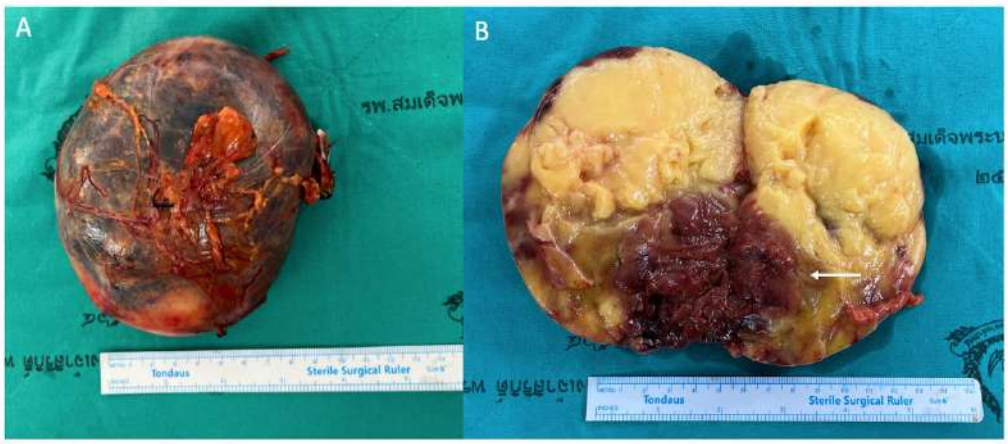

Figure 2. A homogeneous rubbery fatty-like mass measuring 10 x 9.5 x 8 cm. (A). The mass shows area of hemorrhage (Arrow)(B).

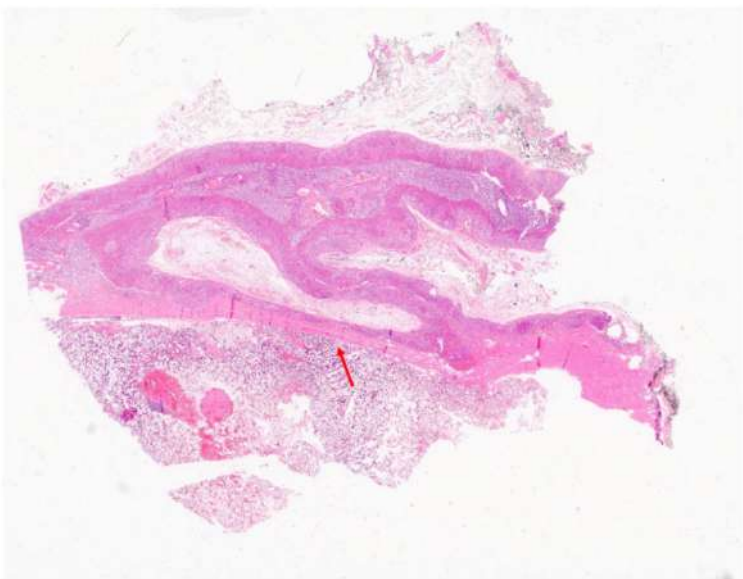

Figure 3. A low-power field of the adrenal gland (arrow) surrounded by myelolipoma.

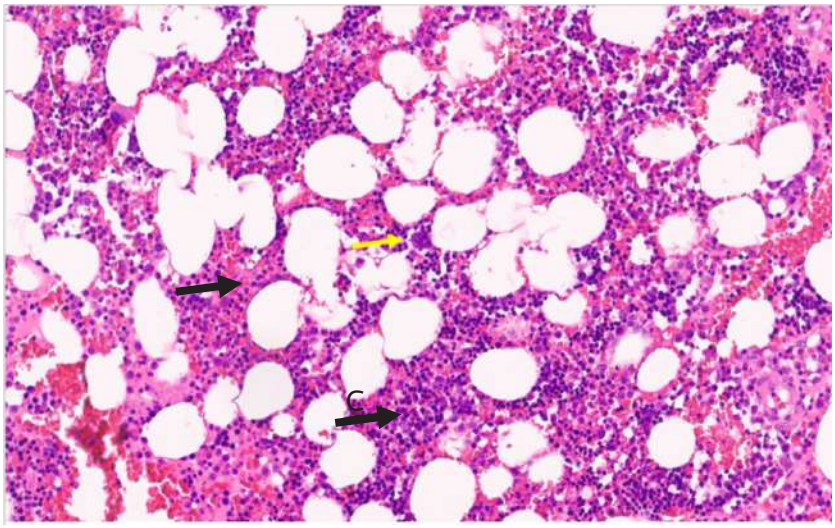

Figure 4. A high-power field showing myelolipoma with megakaryocytes (A), erythroid precursors (B), and myeloid precusors (C).

A hematologist was consulted to investigate a hematologic cause. Hb electro-phoresis was performed, which revealed Hemoglobin A at 97.4% and Hemoglobin A2 at 2.6%. PCR-DNA for Alpha-thalassemia tested positive for Alpha-thalassemia 1 (--SEA), diagnosing the patient with alpha-thalassemia 1 trait, which could not cause EMH. Comprehensive markers for myeloproliferative neoplasms were analyzed, and all tests were negative for JAK2 V617F, CALR, and MPL mutations.

This research was approved by the Research Ethics Committee of the Naval Medical Department under protocol number RP023/67 on June 25, 2024. The study details were fully disclosed to the patient, who provided informed consent for publication.

Discussion

Myelolipomas and extramedullary hematopoiesis (EMH) share a similar histological composition, comprising mature adipocytes and trilinear hematopoietic cells, akin to bone marrow elements. Despite this resemblance, they represent distinct pathological entities. EMH is a physiological process during fetal development that may persist in response to hematological disorders, such as myeloproliferative diseases and hemoglobinopathies. Conversely, myelolipomas are benign, encapsulated tumors containing both fat and hematopoietic components, typically unrelated to hematologic abnormalities and found in patients with otherwise healthy bone marrow.[3]

In this case, a massive EMH was identified surrounding a normal adrenal gland in a patient without any detectable hematologic disorder. This finding is highly unusual, as EMH typically presents in the context of systemic hematologic conditions.

The absence of such underlying pathology in this patient makes the case noteworthy, emphasizing the need to consider EMH in the differential diagnosis of adrenal masses, particularly when imaging features suggest a heterogeneous fatty composition.

The management of adrenal myelolipoma should be individualized due to the absence of formal guidelines. Most myelolipomas are incidentally discovered and asymptomatic; they are often smaller than symptomatic tumors. When imaging clearly identifies a myelolipoma and no symptoms are present, treatment is unnecessary. Surgical resection is the primary treatment option if needed.[2]

For adrenocortical adenomas, the latest guidelines from the European Society of Endocrinology recommend that no further imaging is necessary if an incidentaloma is homogeneous, imaging features are consistent with a benign adrenal mass (< 10 HU), and it is smaller than 4 cm in diameter.[5,9-12] In contrast, myelolipomas are typically larger and slow-growing. Due to insufficient data, it is challenging to determine a specific threshold diameter for follow-up imaging. However, monitoring may be warranted for giant myelolipomas (greater than 10 cm), though the appropriate frequency for follow-up remains undetermined. Given the benign nature of myelolipoma, it is conceivable that these patients would require a long-term follow-up, and an initial surgical approach seems justified since routine imaging may not reliably distinguish a benign lipid-poor mass from a malignant one.

What sets this case apart from existing literature is the identification of extensive EMH surrounding a structurally normal adrenal gland, with no associated hematologic disorder. This underscores the importance of thorough histopathological examination to differentiate EMH from neoplastic processes, especially in cases involving atypical adrenal masses.

The contribution of this case to the literature lies in highlighting EMH as a rare but possible diagnosis, even in the absence of systemic hematologic conditions. Clinicians should be aware of this possibility when encountering adrenal masses with unusual imaging characteristics. Further research and documentation of similar cases are needed to better understand the pathogenesis and optimal management strategies for EMH in such atypical presentations.

Conclusion

Extramedullary hematopoiesis surrounding a normal adrenal gland is a very rare site, which was observed in the microscopic examination of this case. Meanwhile, adrenal myelolipoma is composed of fat and myeloid tissue, and is listed in the group of mesenchymal and stromal tumors of the adrenal cortex. This case emphasizes the diagnostic challenges and management complexities in patients with incidental findings of extramedullary hematopoiesis as adrenal mass, particularly when not accompanied by a hematologic medical history.

References

1. Lau HY, Lui DCY, Ma JKF, Wong RWC. Sonographic features of adrenal extramedullary hematopoiesis. J Ultrasound Med 2011;30:706-7.
2. Kamran H, Haghpanah A, Dabbaghmanesh MH, Defidio L, Bazrafkan M, Dehghani A, et al. Simultaneous adrenal and retroperitoneal myelolipoma resected by laparoscopic surgery: a challenging case. BMC Urol 2023;23(1):114
3. Decmann Á, Perge P, Tóth M, Igaz P. Adrenal myelolipoma: a comprehensive review. Endocrine 2018;59(1):7-15.
4. Fassnacht M, Tsagarakis S, Terzolo M, Tabarin A, Sahdev A, Newell-Price J, et al. European Society of Endocrinology clinical practice guidelines on the management of adrenal incidentalomas, in collaboration with the European Network for the Study of Adrenal Tumors. Eur J Endocrinol 2023;189(1):G1-G42.
5. Calissendorff J, Juhlin CC, Sundin A, Bancos I, Falhammar H. Adrenal myelolipomas. Lancet Diabetes Endocrinol 2021;9(11):767-75.
6. Young WF. Clinical practice. The incidentally discovered adrenal mass. N Engl J Med 2007;356:601-10.
7. O'Regan KN, Jagannathan J, Krajewski K, Zukotynski K, Souza F, Wagner A, et al. Imaging of liposarcoma: classification, patterns of tumor recurrence, and response to treatment. AJR Am J Roentgenol 2011;197(01):W37-43.
8. Lam AK. Update on adrenal tumours in 2017 World Health Organization (WHO) of endocrine tumours. Endocr Pathol 2017;28(3):213-27.
9. Esfandbod M, Kabootari M. Adrenal extramedullary hematopoiesis: a case report. J Clin Case Reports 2012;2(16):1000226.
10. Ebbehoj A, Li D, Kaur RJ, Zhang C, Singh S, Li T, et al. Epidemiology of adrenal tumours in Olmsted County, Minnesota, USA: a population-based cohort study. Lancet Diabetes Endocrinol 2020;8(11):894-902.
11. Jing Y, Hu J, Luo R, Mao Y, Luo Z, Yang J, et al. Prevalence and characteristics of adrenal tumors in an unselected screening population: a cross-sectional study. Ann Intern Med 2022;175(10):1383-91.
12. Hamidi O, Raman R, Lazik N, Iniguez-Ariza N, McKenzie TJ, Lyden ML, et al. Clinical course of adrenal myelolipoma: a long-term longitudinal follow-up study. Clin Endocrinol (Oxf) 2020;93(1):11-8.