CASE REPORT

Extramedullary Hematopoiesis in a Giant Adrenal Myelolipoma: A Case Report

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Abstract

Introduction: Adrenal myelolipoma with extramedullary hematopoiesis (EMH) is an exceedingly rare entity with very few cases reported in the literature. We present a rare case of adrenal myelolipoma with EMH.

Case report: A 48-year-old man with the previous history of splenectomy for hereditary spherocytosis was presented with 12 × 12 cm right adrenal tumor and underwent laparoscopic adrenalectomy. The histopathological features confirmed the diagnosis of adrenal myelolipoma with EMH.

Conclusion: In any patient with a history of chronic hemolytic anemia and adrenal mass, it is imperative to consider EMH as a possible diagnosis.

Keywords: Adrenal myelolipoma, Case report, Extramedullary hematopoiesis, Hemolytic anemia.

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Introduction

Myelolipoma arising in the adrenal is a not so rare entity and is usually asymptomatic, diagnosed incidentally on radiological examination for other reasons. Extramedullary hematopoiesis is the formation of cellular components of blood, occurring outside the bone marrow. Adrenal myelolipoma with EMH is an exceedingly rare entity with very few cases reported in the literature.

Case Description

A 48-year-old man presented with a complaint of right flank pain for 10 years. He was diagnosed with hereditary spherocytosis and had undergone splenectomy 14 years ago. For flank pain, he was evaluated with ultrasonography of the whole abdomen and pelvis. It revealed hepatomegaly, gallstone, and a mixed echogenic mass in the right suprarenal region measuring 10 × 6 cm. The patient underwent further evaluation with contrast-enhanced computed tomography. The study revealed evidence of well-defined, lobulated outlined, mixed solid, and fat density focus on the right adrenal gland with nearly 30–40% fat contents seen in the lateral aspect of the focus. Mild enhancement of the solid components was seen. There was no evidence of any microcalcification. The focus measured about 12 × 8 cm. The right kidney was pushed inferiorly. The left adrenal gland was unremarkable. Multiple enlarged paravertebral soft tissue density foci were seen along the lower dorsal spine, the largest measuring 7 cm × 5 cm (Fig. 1). In view of the history of splenectomy, these sites were confirmed as EMH.

Due to his recurrent flank pain, laparoscopic right adrenalectomy was performed. Histopathological examination revealed a tumor composed of lobules of mature adipose tissue admixed with normal appearing hematopoietic element with all three-cell lineage namely myeloid cells, erythroid cells, and megakaryocytes. Extensive areas of hemorrhage and infarct-type necrosis were seen. Histological features were suggestive of myelolipoma with EMH (Fig. 2). The patient is doing well after 18 months of surgery.

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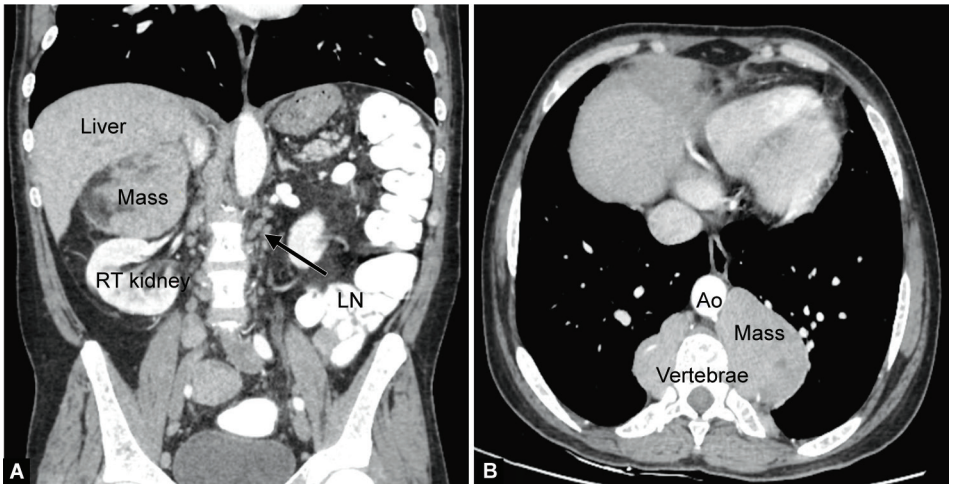
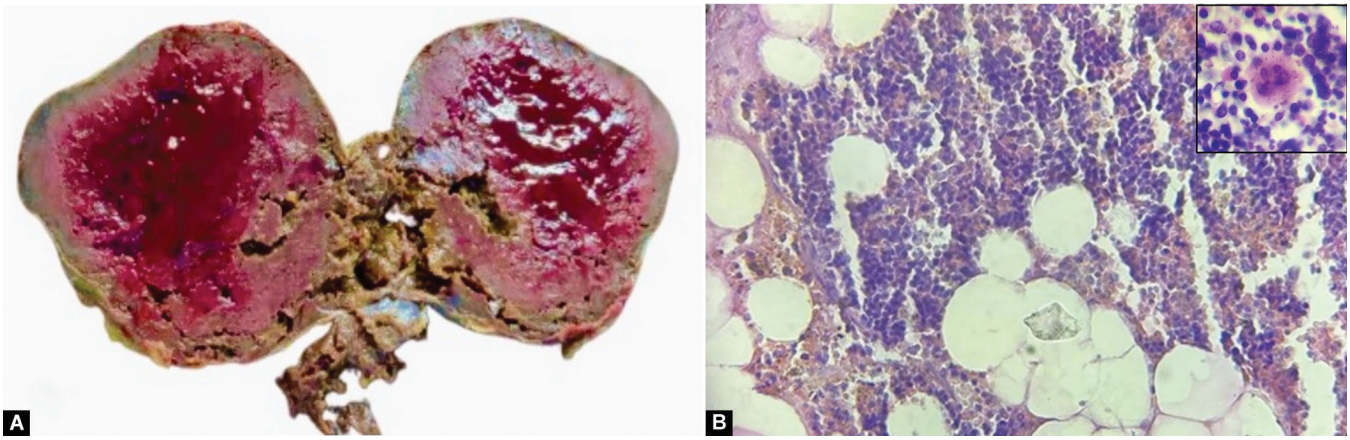
Discussion

Extramedullary hematopoiesis is a physiological compensatory phenomenon in response to altered hematopoiesis occurring secondary to inadequate bone marrow function.[1] Extramedullary hematopoiesis is commonly reported in the liver and spleen whereas it has rarely been reported in adrenal glands, breast, dura mater, and bowel as well.[1] There are several theories as to the pathogenesis of adrenal EMH. It may be that the adrenal gland contains primitive rests that have hematopoietic potential.[2] Another hypothesis is that the adrenal gland is directly invaded by hematopoietic bone marrow from adjacent bone erosions or fractures.[2] Few cases of EMH in the adrenal gland have been reported, which were clinically detected as incidental myelolipoma.[2–4] Adrenal EMH can be managed conservatively in asymptomatic patients, but symptomatic patients may require a blood transfusion, radiotherapy, hydroxyurea, or surgical resection.[1] Small asymptomatic lesions of <5 cm are usually



Figs 1A and B: Contrast-enhanced computed tomography (CECT) abdomen showing (A) well-defined, lobulated outlined, mixed solid, and fat density focus in the right adrenal gland with nearly 30–40% fat contents seen in the lateral aspect of the focus. Mild enhancement of the solid components is seen. (B) Multiple enlarged paravertebral soft tissue density foci (EMH sites)



Figs 2A and B: (A) Cut surface revealing large areas of hemorrhage and pale yellow colored areas consistent with adipose tissue. (B) H and E sections show adipose tissue and hematopoietic elements with areas of hemorrhage. Inset is showing megakaryocyte (high magnification: 40×)

observed for 1–2 years with imaging controls. However, tumors >10 cm have a potential risk of malignancy and hemorrhagic complications, and hence surgical resection is preferred.[3,4] In view of symptoms and large size, we proceeded with laparoscopic adrenalectomy in our patients, which is the standard procedure currently.

In conclusion, adrenal mass in patients with a history of hemolytic anemia could be an EMH site and should be managed accordingly.

Patient Perspective

My flank pain is now relieved after the surgery.

Authors' Contributions

SJ and SKY: Planning, implementation, data collection, writing, review, and editing.

NG, RM, and RY: Writing of pathology part, review, and editing.

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