

Extramedullary Hematopoiesis in a Man With β -Thalassemia: An Uncommon Cause of an Adrenal Mass

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Introduction: Extramedullary hematopoiesis (EMH) commonly occurs in the spleen, liver and lymph nodes. Rare cases of EMH in the adrenal gland have been reported.

Case Presentation: We report the case of a 33-year-old man from the South of Iran suffering from major β -thalassemia, who underwent open left adrenalectomy and the histopathology revealed EMH.

Conclusions: In patients in which a history of hematologic disorders exists, careful imaging and hormonal assay should be done to certify a diagnosis of EMH. However, the surgical management becomes inevitable in certain cases.

Keywords: Extramedullary hematopoiesis; Thalassemia; Adrenal

1. Introduction

Extramedullary hematopoiesis (EMH) is a compensatory mechanism for deficient formation or function of red blood cells (1). It can present, uncommonly, as a soft tissue mass. Most frequently, it occurs in the spleen, liver, lymph nodes and less frequently, in the lung, pleura, thymus and breasts. Rare cases of EMH were seen in adrenals (2, 3). The EMH may occur in congenital or acquired hemolytic diseases, ineffective erythropoiesis, loss of stem cell differentiation or non-myeloid neoplastic diseases (4). The β -thalassemia is caused by the absence or reduction of b-globin chains. Even patients with mild forms may develop EMH (5). We report the case of a 33-year-old man underwent left adrenalectomy for an incidentally detected adrenal mass.

2. Case Presentation

A 33-year-old man, from the South of Iran, suffering from major β -thalassemia with weight loss and loss of appetite few months prior to the presentation, was admitted in the Hazrat Aliasghar Hospital. He had a positive history previous splenectomy. He was pale yet he did not present jaundice. Laboratory data revealed anemia hemoglobin (Hb) = 11 g/dl, white blood cell count (WBC) = $10000 \times 10^6/L$, platelets (PLT) = $257000 \times 10^6/L$, normal liver function tests (LFT) and normal adrenal hormone levels. Abdomino-pelvic sonography showed left adre-

nal mass, measuring 8 × 8 cm, with hypoechoic, homogenous ecostructure. Computed tomography (CT) of the abdominopelvic area revealed a 9 × 8 cm hyperdense and heterogenous structure with certain degree of enhancement (Figure 1) Magnetic resonance imaging (MRI) findings described a 9 × 7 × 7 cm heterogenous hypoechoic

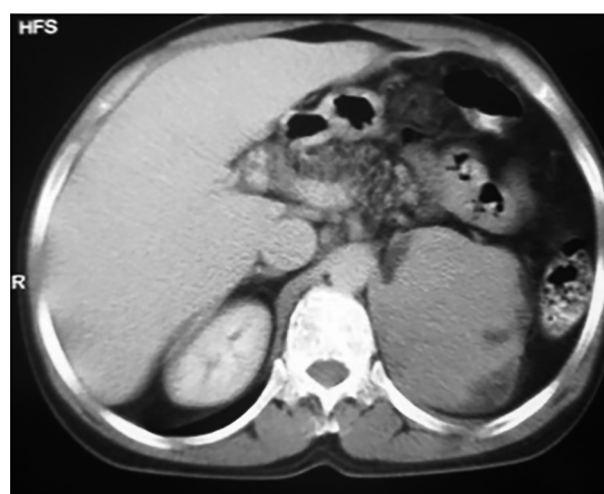


Figure 1. Contrast Enhanced Computed Tomography Scan, Showing Left Adrenal Mass

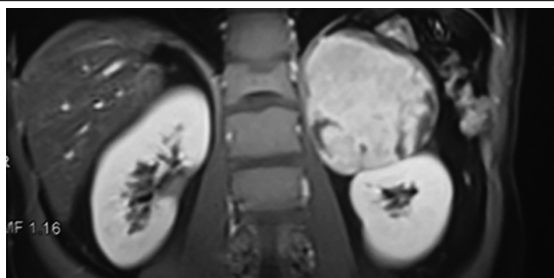


Figure 2. Coronal Magnetic Resonance Imaging of the Abdomen Demonstrates a Solid Left Suprarenal Mass



Figure 3. Excised Specimen of Left Adrenal Shown Surface

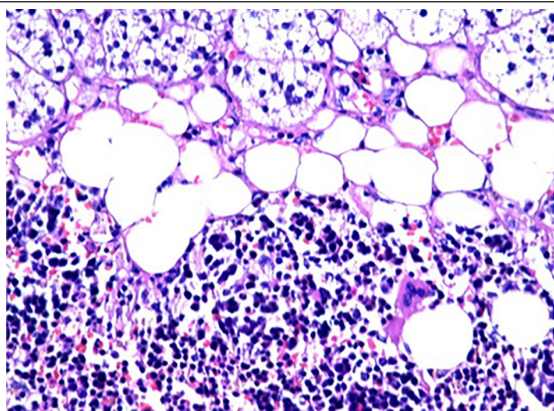


Figure 4. Adrenal Examedullary Hematopoiesis, Normal Adrenal Parenchyma (Upper Part of the figure) and Extramedullary Hematopoiesis (Lower Part of the figure) Including Various Stages of Hematopoietic Cells

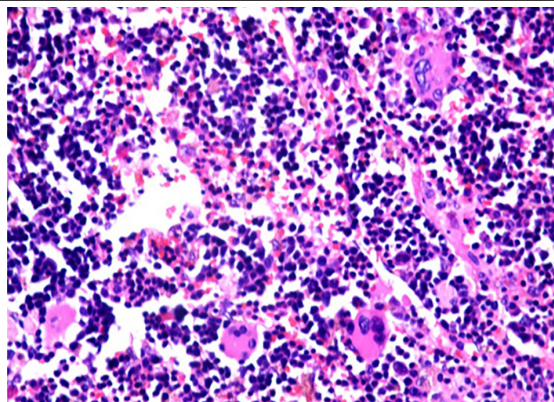


Figure 5. Various stages of Hematopoietic Cells with Predominance of Erythroid Series and Few Megakaryocytes

mass, with enhancement after contrast injection (Figure 2). The patient underwent left adrenalectomy via anterior subcostal incision. We preferred open surgery because of previous surgery (splenectomy) and probable adhesions. The specimen was removed with a rim-like adrenal gland on it. Gross examination showed a well-circumscribed dark brown mass, measuring $8 \times 6 \times 5$ cm (Figure 3). Histopathologic examination demonstrated extramedullary hematopoiesis, with foci of hemorrhage (Figures 4 and 5). Despite severe nausea and vomiting for 48 hours, the post-operative period was uneventful.

3. Discussion

The EMH is defined as the synthesis of hematopoietic elements outside the bone marrow. It can cause masses that are almost always small and, commonly, it is an incidental finding on autopsy. However rarely, EMH may have a relatively large size, with significant mass effect (6). Its prevalence is 3.6% of all surgically removed adrenal incidentalomas (3). The EMH in the adrenal is uncommon and few cases have been reported till now. Patients with adrenal EMH are generally known to have major β -thalassemia or other major hemoglobinopathies. When facing an adrenal mass, the size criteria is an important feature for making a decision (7). Masses larger than 6 cm should be considered malignant, until proven otherwise (8). When EMH presents as a soft tissue mass, it often mimics a more serious diagnosis, requiring radically different treatment options (2). The EMH could present special features on radiologic investigations that help in diagnosis. On ultrasonographic examination, it presents like a homogenous, hypoechoic and well defined round mass. The CT scans show it as a non-homogenous low-density tumor (3). The MRI, which is another diagnostic modality, the mass has low to intermediate signal on T1-weighted sequence and low to high signal on T2-weighted images (1). Radionuclide bone marrow imaging, with technetium-99m sulfur colloid may be helpful in diagnosis (1, 6). Also, biopsy is suggested sometimes to avoid surgery, although it has its own limitations: not reliable and not always safe (8). Although the fine needle aspiration has pitfalls regarding the cytologic features, it could be helpful when considering all the patients' data (9). If any hematologic disorder exists in a patient, careful imaging and hormonal assay should be considered and, at times, surgical management is inevitable.

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Authors' Contributions

Shahriyar Zeighami: case finding, literature review, critical appraisal of final manuscript, administrative and funding support, design and primary idea; Seyed Ali Eslahi: case finding, literature review, critical appraisal of fi-

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References

1. Singer A, Maldjian P, Simmons MZ. Extramedullary hematopoiesis presenting as a focal splenic mass: a case report. *Abdom Imaging*. 2004;**29**(6):710-2.
2. Calhoun SK, Murphy RC, Shariati N, Jacir N, Bergman K. Extramedullary hematopoiesis in a child with hereditary spherocytosis: an uncommon cause of an adrenal mass. *Pediatr Radiol*. 2001;**31**(12):879-81.
3. Porcaro AB, Novella G, Antonioli SZ, Martignoni G, Brunelli M, Curti P. Adrenal extramedullary hematopoiesis: report on a pediatric case and update of the literature. *Int Urol Nephrol*. 2001;**33**(4):601-3.
4. Rajiah P, Hayashi R, Bauer TW, Sundaram M. Extramedullary hematopoiesis in unusual locations in hematologically compromised and noncompromised patients. *Skeletal Radiol*. 2011;**40**(7):947-53.
5. Goerner M, Gerull S, Schaefer E, Just M, Sure M, Hirnle P. Painful spinal cord compression as a complication of extramedullary hematopoiesis associated with beta-thalassemia intermedia. *Strahlenther Onkol*. 2008;**184**(4):224-6.
6. Gupta P, Eshaghi N, Ghole V, Ketkar M, Garcia-Morales F. Presacral extramedullary hematopoiesis: report of a case and review of the literature. *Clin Imaging*. 2008;**32**(6):487-9.
7. Banerji JS, Kumar RM, Devasia A. Extramedullary hematopoiesis in the adrenal: Case report and review of literature. *Can Urol Assoc J*. 2013;**7**(5-6):E436-8.
8. Wein LRK, Andrew CN, Alan WP, Craig AP. *Campbell-walsh urology*. 10th ed Philadelphia: Saunders; 2011.
9. Policarpio-Nicolas ML, Bregman SG, Ihsan M, Atkins KA. Mass-forming extramedullary hematopoiesis diagnosed by fine-needle aspiration cytology. *Diagn Cytopathol*. 2006;**34**(12):807-11.