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Extramedullary Hematopoiesis: Uncommon Case of Giant Adrenal Incidentaloma Mimicking Tumor Mass

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Abstract

Extramedullary Hematopoiesis (EMH) is a compensatory phenomenon to insufficient bone marrow function to maintain the circulatory demands. EMH occurs most commonly in the reticuloendothelial system such as spleen, Liver and lymph nodes, or in para osseous sites. EMH in the adrenal is rare with fewer than 10 cases reported, presenting as a giant adrenal mass are extremely rare. We report 38-year-old women with history of β-thalassemia since childhood. Computed Tomography (CT) was revealed a giant right adrenal mass with heterogeneous density mimicking a tumor mass with moderate splenomegaly associated, after excluding hormonal secretion. Ultrasound-guided biopsy was performed and reported fibrocollagenous tissue displaying trilineage haematopoiesis in favor of adrenal EMH, thus malignancy is eliminated. As a giant mass, treatment was an excision surgery associated with iterative transfusions to prevent recurrence. In daily practice, practitioners should pay attention to the differential diagnosis in any patients with chronic hematologic disorders suggestive for EMH even of uncommon location. A confirmatory preoperative biopsy is recommended to avoid unnecessary procedure.

Keywords: Extramedullary hematopoiesis • Adrenal incidentaloma • Surgery • Pathology

Introduction

While EMH is physiological during fetal development, pathogenic EMH can emanate when physiological hematopoiesis in the bone marrow is ineffective and hematopoietic stem cells migrate to other tissues, as is often seen in chronic disorders of the hematopoietic system, such as thalassemia, myelofibrosis, polycythemia, leukemia, lymphoma, after bone marrow irradiation, poisoning, and neoplastic conditions [1]. Cytokine induction leads to increased cell production and a greater availability of pluripotent stem cells. This phenomenon occurs mainly in embryonic hematopoiesis sites (spleen, liver, lymph nodes), sometimes in other organs like the kidney, lung and digestive tract. To the best of our knowledge, we documented an extremely rare reported case of adrenal EMH presenting as a giant mass which is clinically referred to as incidentaloma.

Case Report

We report the case of 38-year-old women who had been known to have β-thalassemia since childhood, presented to the hospital with non-specific upper abdominal pain and progressive worsening of asthenia. On physical exam, he was pale, icteric with a blood pressure of 130/80 mm Hg. He had frontal bossing and prominent facial bones. Abdominal exam revealed moderate splenomegaly and no other palpable mass.

Laboratory investigations showed thrombocytopenia (100 × 10 9 G/L platelets; Hemoglobin 8 × 10 G/L; WBC count 7.4 × 10 9 G/L, MCV 64.7 fl, MCH 20.8 pg. The results from biochemical investigations were: serum bilirubin 12.5 μmol/L, conjugated bilirubin 2.7 μmol/L, blood urea 5.8 mmol/L, serum creatinine 130.0 μmol/L, serum calcium 1.8 mmol/L, serum potassium 4 mmol/L, serum sodium 140.0 mmol/L, alanine aminotransferase 20.1 U/L, aspartate aminotransferase 19.2 U/L. Urinalysis in sediment, showed many leukocytes and erythrocytes. Serum cortisol and 24-hour urinary metanephrines were within normal limits excluding tumor hormonal secretion.

He had splenomegaly and a large incidental hypoechoic right suprarenal mass in the ultrasonography of the abdomen. On further characterisation by a CT scan, the mass was found to arise from the right adrenal gland of size 13 × 11 cm with heterogeneous density, cystic and necrotic changes and a speck of calcification (Figure 1). In the light of these radiological and biological data, the differential diagnosis included a malignant non-secreting adrenal tumor. Ultrasound-guided biopsy is performed and reported fibrocollagenous tissue displaying trilineage haematopoiesis in favor of adrenal EMH, thus malignancy is eliminated. As a giant mass with bleeding risk and locoregional compression, a surgical excision is indicated. The excised specimen was 13 × 11 × 5 cm, weighed about 400 g, a well-circumscribed brownish mass, fragile and homogenous with no obvious fat component. A rim of normal looking adrenal tissue was noted at the periphery (Figure 2). Histology revealed the mass to be composed of mature hemopoietic elements, with a predominance of erythroid series. A few megakaryocytes were also noted (Figure 3). There was no need for an immunohistochemical complement in view of the typical histological appearance to rule out another differential diagnosis. The absence of lymphoid follicles in multiple sections allowed us to exclude an accessory spleen that

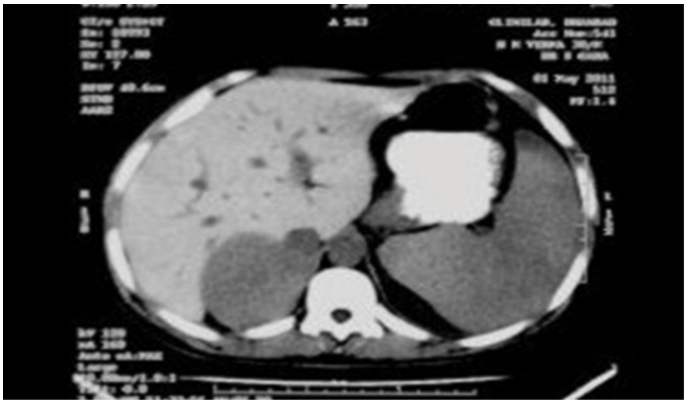

Figure 1. Contrast enhanced computed tomography showing right adrenal mass in a lower section. The right kidney is still not seen in this section, as the adrenal mass had pushed it inferiorly. Note the splenomegaly associated on the left.

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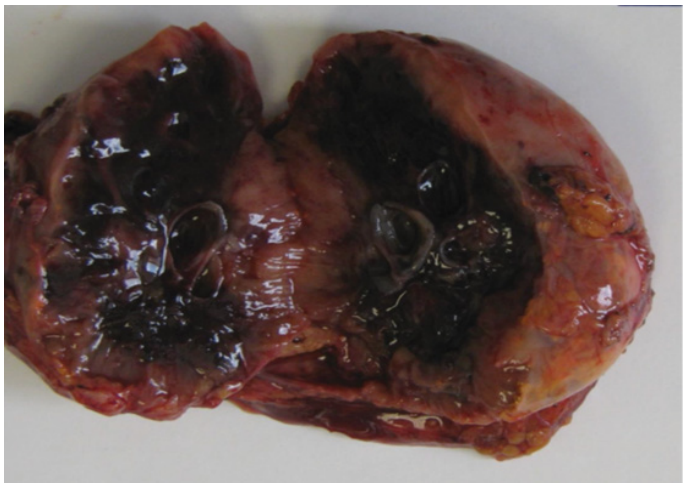

Figure 2. Excised specimen of the right adrenal showing a reddish-brown surface with large haemorrhagic changes.

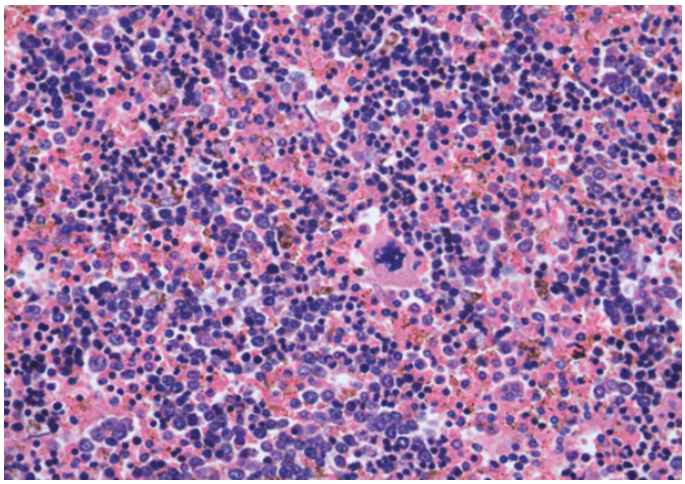

Figure 3. Hematoxylin and eosin stain showing granulocytic, erythroid and megakaryocytic hematopoietic line.

may be present at this location. The absence of a lipomatous component allowed us to rule out myelolipoma, which is another histological differentiation diagnosis. The final diagnosis was EMH of the right adrenal gland. Control examination and CT scan was recommended 6 months after surgery and also monthly evaluation of his peripheral blood count with iterative transfusions.

Discussion

EMH is usually asymptomatic and discovered incidentally. Symptomatic cases occur due to mass effect with compression to adjacent organ or hemorrhagic complications. The adrenal glands are the seat of a wide variety of diseases. However, EMH in the adrenal is uncommon, with less than 10 cases reported [2]. Several hypotheses are suggested trying to explain the mechanism of migration of hematopoietic cells in the adrenal gland. The adrenal gland is hematopoietic capacity during the fetal period and EMH may develop from rests in a sick state. Other scientists believe that the embolization of hematopoietic stem cells and a return to the adrenal gland may occur. Chronic hypoxia is another suspected cause of HME [3]. Our patient had only a chronic positive history of hemolytic anemia by hemoglobinopathy β-thalassemia major.

Initially it was whether it was an adrenocortical carcinoma or a non-functioning adrenal adenoma. Adrenal adenomas normally do not grow larger than 3 cm. Moreover, the densities in adrenal adenomas are less than 0 in an unenhanced CT [4]. By size criteria, heterogeneously enhancing and highly vascular [4,5] the likelihood of this mass being an adrenocortical carcinoma was high. The anemia and splenomegaly could have been pointers towards the diagnosis of an underlying hematological disorder. The practitioners should pay attention to this entity, and an image-guided biopsy of the adrenal gland in most instances is likely to give the diagnosis.

Different treatment options include surgical decompression, blood transfusion, radiotherapy, hydroxyurea or a combination of these modalities. Treatment depends on severity of symptoms, mass size, clinical status and previous treatment

Most reported giant cases due to EMH have been treated with surgical decompression [6] with or without radiation therapy in emergency cases, intravenous steroids may be used as the temporizing measure until definitive treatment is applied.

Radiation is an applicable therapy [7] and this avoids surgical procedure and associated risks. Suppression of bone marrow due to radiation may be observed in already anaemic patients, also may cause special risk (e.g. heart failure).

Hydroxyurea [8] successfully increases fetal haemoglobin in patients with sickle cell disease but there is limited experience with hydroxyurea in thalassaemia.

Hypertransfusion [9] to keep the hemoglobin level at 12-14 g/dL seems to be an acceptable method of treatment that should be recommended as a first-line approach or as an adjuvant therapy to other methods.

In our case, the size of the tumor was 13 cm in diameter. Due to our preoperative histological diagnosis, regardless of the carcinogenic risk, enucleation of the mass was performed rather in relation to the compression of adjacent organs and the risk of a hemorrhagic complication. Iterative transfusions to prevent recurrence were associated.

Conclusion

We have presented an unusual case of EMH as a giant adrenal mass that requires surgical excision. In case of a chronic hematologic disorder, a biopsy is indicated for a better management of adrenal incidentaloma. If it reveals extramedullary haematopoesis, the therapeutic procedure will be well adapted.

References

1. Alam, MR, Habib MS, Guru Dhakal P and Khan MR, et al. Extramedullary hematopoiesis and paraplegia in a patient with hemoglobin e-Beta thalassemia. *Mymensingh Med J* 19(2010): 452-457.
2. Christos, S, Georgiades, Edward G, Neyman, Isaac R, Francis and Michael B Sneider, et al. Typical and atypical presentations of extramedullary hematopoiesis. *AJR Am J Roentgenol* 179(2002):1239-1243.
3. Bastounis ,EA, Anastasios J, Karayiannakis, Lydia Nakopoulou and EL Papalambros, et al. Incidentalomas of the adrenal gland: Diagnostic and therapeutic implications. *Am Surg* 63(1997): 356-360.
4. King, Bernard F, Kopecky, Kenyon K, Baker, Martin K and Clark, Steven A. Extramedullary hematopoiesis in the adrenal glands: CT characteristics. *J Comput Assist Tomogr* 11(1987): 342-343.
5. Yee Lau, Hang, Dilys Choi Yu Lui, Johnny Ka Fai Ma and Richard Wing Cheuk Wong. Sonographic features of adrenal extramedullary hematopoiesis. *J Ultrasound Med* 30(2011): 706-713.
6. Dore, F, Cianciulli P, Rovasio S and Oggiano L, et al. Incidence and clinical study of ectopic erythropoiesis in adult patients with thalassemia intermedia. *Ann Ital Med Int* 7(1992): 137-140.
7. Berhouma, Moncef and Jemel Hafedh. Management of spinal cord compression caused by extramedullary hematopoiesis in beta-thalassemia. *Intern Med* 47(2008): 1125-1128.
8. Saxon, Ben R., David Rees and Nancy F. Olivieri. Regression of extramedullary haemopoiesis and augmentation of fetal haemoglobin concentration during hydroxyurea therapy in β-thalassaemia. *Br J Haematol* 101(1998): 416-419.

9. Tai, SM, Chan JS, Shau Yin Ha and Young BW, et al. Successful treatment of spinal cord compression secondary to extramedullary hematopoietic mass by hypertransfusion in a patient with thalassemia major. *Pediatr Hematol Oncol* 23(2006): 317-321.

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