Adrenal Extramedullary Hematopoiesis in a patient with Beta Thalassemia Major

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ABSTRACT

Introduction: **Extramedullary hematopoiesis in the adrenal gland is rare and is usually discovered incidentally. Thalassemia is one of the hematological disorders in which extramedullary hematopoiesis could be seen in many organs including adrenals**

Case report: **Our patient was a nineteen years old Iraqi man. He is a known case of thalassemia major since the age of 5 months. He was discovered to has a right suprarenal mass by ultrasound examination as part of investigation for nocturia. The mass was surgically excised and the histopathological examination revealed extramedullary hematopoiesis.**

Conclusion: **Awareness should be raised to consider the diagnosis of extramedullary hematopoiesis in any thalassemic patient with a tissue mass**

Key words: **Thalassemia, Extramedullary hematopoiesis, Adrenal**

تكون الخلايا المولدة للدم خارج النخاع في الغدة الكظرية لمريض مصاب بالثلاسيميا الكبيرة نوع بيتا– تسجيل حالة

المقدمة: ان تكون الخلايا المكونه للدم خارج النخاع في الغدة الكظرية هي حالة نادرة وعادة ما يتم اكتشافها صدفة. تعتبر الثلاسيميا هي واحدة من اضطرابات الدم التي يمكن أن ينظر فيها تكون الدم خارج الدم في العديد من اعضاء الجسم بما في ذلك الغدة الكظرية.

تقرير الحالة: كان مريضنا رجلًا عراقيًا يبلغ من العمر تسعة عشر عامًا .وهو حالة معروفة من مرض الثلاسيميا الكبرى منذ عمر ٥ أشهر تم اكتشاف أن لديه كتلة فوق الكلى اليمنى عن طريق فحص الموجات فوق الصوتية كجزء من التحقيق من أجل التبول الليلي .تم استئصال الكتلة جراحيا وكشف الفحص النسيجي المرضي "تكون الخلايا المكونه للدم خارج النخاع"

الاستنتاج : ينبغي رفع الوعي للنظر في تشخيص تكون الدم خارج النخاع في أي مريض بالثلاسيميا لديه كتلة نسيجية.

INTRODUCTION

Thalassemia is an autosomal recessive disease associated with chronic anemia. Iraq is one of the countries with the high incidence and prevalence of thalassemia.[1] Extramedullary hematopoiesis is a recognized complication of ineffective hematopoiesis with production of blood elements outside the bone marrow. Extramedullary hematopoiesis can be seen in chronic hemolytic diseases like thalassemia, sickle cell anemia and hereditary spherocytosis. It can also complicate myelofibrosis and polycythemia Vera.[2] Extramedullary hematopoiesis involves most commonly the reticuloendothelial system, but it can also be seen in other organs, like the pleura, lungs, gastrointestinal tract, breast, skin, brain, kidneys, paraspinal tissue and adrenal glands.[3-5]

CASE REPORT

Our patient is 19 years old Iraqi man. He is a known case of thalassemia major diagnosed at the age of five months. The patient was underwent splenectomy at the age of 9 years. During his life, he has had a pre-transfusion hemoglobin level of 6-9 g/dL with regular blood transfusion every 20-30 days. He was complaining of nocturia and accidentally found

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to have right supra renal mass by abdominal ultrasound examination and computed tomography (CT) scan. The mass was surgically excised because of a presumptive diagnosis of adrenal tumor that was suggested by the radiologist. On macroscopic examination, the excised mass was oval in shape, 7.2 cm. in maximum dimension, with a dark brown smooth outer surface. The cut surface of the mass was brown, lobulated and firm with several hard calcified areas (Fig-1). The histopathological examination revealed cellular proliferation of numerous myeloid and erythroid cells with several megakaryocytes (Fig-2). Immunohistochemical stains for megakaryocytes, myeloid and erythroid linages were positive. The pathological diagnosis was extramedullary hematopoiesis in the right adrenal gland.

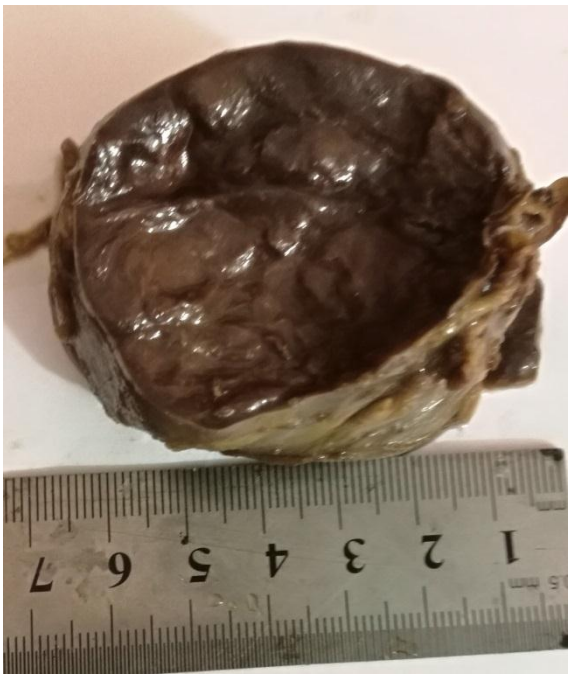

Fig 1. Macroscopic appearance of the mass with smooth outer surface and lobulated brown cut surface.

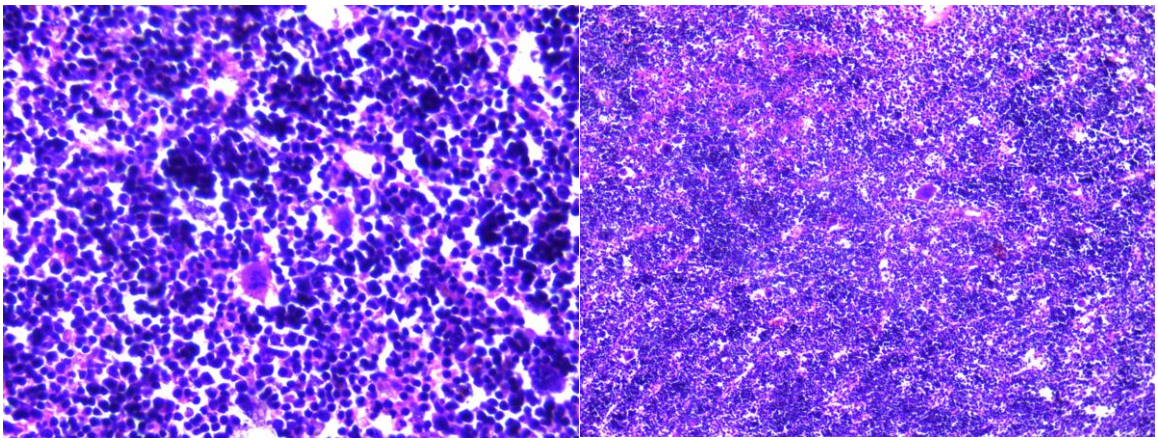

Fig 2. The microscopic examination of the mass revealed cellular proliferation of different hematopoietic cells

DISCUSSION

Extramedullary hematopoiesis commonly occurs in untransfused patients with thalassemia intermedia and less commonly in patients with β-thalassemia major when erythropoiesis is inadequately suppressed by blood transfusions.[6] While the incidence of extramedullary hematopoiesis in patients with thalassemia major is less than 1%, it may reach up to 20% in patients with thalassemia intermedia[2] Extramedullary hematopoiesis is frequently seen in the liver and spleen which can potentially produce fetal hemoglobin. However, it has been reported in other organs, like the pleura, lungs, gastrointestinal tract, breast, skin, brain, kidneys, paraspinal tissue and adrenal glands.[3-5] Adrenal gland Extramedullary hematopoiesis is rare, several cases had been reported worldwide.[7,8,10,11] Most cases of Extramedullary hematopoiesis are discovered incidentally. They are asymptomatic in most of the cases. The symptoms are due to mass effect and are related to the site affected, such as spinal cord compression. [9] Our patient was complaining of nocturia and he was subjected to ultrasound examination when the adrenal mass was initially discovered. In thalassemic patient with adrenal mass, the diagnosis of extramedullary hematopoiesis should be considered to avoid unnecessary surgical excision.[7] The diagnosis of adrenal Extramedullary hematopoiesis can be made by true cut needle biopsy guided by ultrasound or CT. scan. In our case the presumptive diagnosis of adrenal tumor suggested by the radiologist and the large size of the mass encouraged the surgeon to perform complete excision of the mass. However, surgical resection is inevitable in certain cases of adrenal extramedullary hematopoiesis particularly in the presence of a large adrenal mass.[10]

CONCLUSION

The diagnosis of extramedullary hematopoiesis needs to be considered in any patient with thalassemia who presents with a tissue mass. We believe that the publication of this case report may increase the awareness of the diagnosis of extramedullary hematopoiesis in thalassemic patients.

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