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Adrenal Mass Revealing Extramedullary Hematopoiesis

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Abstract **Case Report**

Adrenal hematopoiesis is a rare condition. It must be suspected in case of any incidentaloma in a patient treated for chronic hemolytic pathology. Surgery remains the most effective treatment for large symptomatic masses. We report a case of large adrenal mass in young patient with history of beta-Thalassemia intermedia, managed by open resection. The post-operative course was uneventful.

Keywords: Extramedullary hematopoiesis, adrenal mass, Beta-Thalassemia, incidentaloma, hydroxyurea, transfusion, case report.

INTRODUCTION

Adrenal hematopoiesis is a rare condition. It is defined by abnormal development of hematopoietic tissue in the adrenal gland. This type of extramedullary hematopoiesis (EMH) is secondary to medullary hyperstimulation to compensate for the insufficient production of blood cells at its normal site, the bone marrow, or a chronic destruction of these cells in the case of chronic hemolytic pathologies. This abnormal production can occur in other organs such as digestive tract, kidneys, lungs ... But more rarely adrenals [1]. Clinically, this phenomenon usually remains asymptomatic for a long time, but is sometimes revealed by symptoms of compression of adjacent organs or pain of a gravity type.

We report the observation of a young patient operated in our surgical clinic for a large symptomatic adrenal mass, in which the histopathological examination revealed a hematoma caused by adrenal hematopoiesis. The aim of this work is to illustrate through this observation the clinical, radiological and therapeutic characteristics of this pathology.

OBSERVATION

A 21-year-old female who has been treated for Beta-Thalassemia intermedia for 15 years, during which time she had received 3 transfusions. An abdominal ultrasound was performed on her for right hypochondrium pain resistant to symptomatic treatments, which revealed a mass on the upper pole of the right kidney. An abdominal CT scan showed a large right retroperitoneal mass, well limited, with regular contours and heterogeneous contrast, delimiting areas of necrosis and containing an upper polar calcification, it measured 122x105x146 mm (Fig1). A biological workup including urinary methoxylates and cortisol levels was without abnormalities. The patient underwent an adrenalectomy through open surgical approach, removing a large friable dark red mass. The post-operative course was simple. Histological examination of the surgical specimen revealed a normal adrenal parenchyma surrounding a huge hematoma which contains cells at different stages of hematopoiesis confirming the diagnosis.

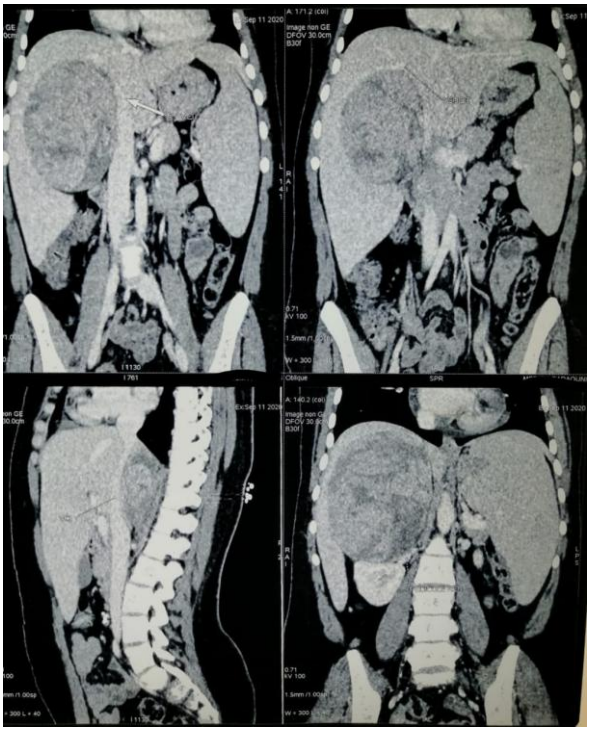

Fig-1: CT scan shows a heterogenous well defined right adrenal mass delimiting areas of necrosis

DISCUSSION

Extramedullary hematopoiesis (EMH) is a compensatory response to chronic anemia in non-transfused beta-thalassemia intermedia patients. Its incidence is 15 to 20% in non-transfused patients [2, 3], and less than 1% in patients correctly transfused since birth [4, 5]. Thus, the therapeutic protocol in thalassemia is to maintain a pre-transfusion hemoglobin level above 9.5 g / dl [6], which allows suppression of ineffective erythropoiesis and thus to slow down the phenomenon of EMH. The most common site of this phenomenon is the dorsal spine followed by the liver and spleen. Adrenal localization is very rare [7].

Adrenal hematopoiesis remains asymptomatic for a long time, and its discovery is generally inadvertent on imaging or, more rarely, following compression syndrome of neighbouring organs or pain when the mass is very large [8], as in the case of our patient. Indeed, an incidentaloma in a patient followed for a hematological pathology such as beta thalassemia should suggest extramedullary hematopoiesis.

Diagnosis is oftenly mentioned by imaging, the CT scan shows in the active hematopoiesis phase a well-limited mass, with tissue density, without or with minimal enhancement after injection of contrast product [9], in the chronic inactive phase the lesions are of fatty density (hypodenses) [10, 11].

Biopsy is often technically difficult and associated with high risk of bleeding [12]. It can be proposed in elderly patients when differential diagnosis with neoplasia is difficult [4, 10]. Histological examination in this case shows hematopoietic tissue containing immature and mature erythroid and myeloid cells [13].

The therapeutic attitude towards EMH is not standardized in literature; it depends on location, symptomatology, size of the mass and its evolution. Several options have been proposed, alone or in combination, for other locations than the adrenal: repeated transfusions, radiotherapy, corticosteroid therapy, hydroxyurea and surgery. For adrenal localization, the surgical indication will depend on the size of the mass. Karami *et al*. recommend for small asymptomatic masses less than 5 cm, radiological monitoring for 1 to 2 years [14]. Chakrabarti *et al*. suggests monthly monitoring for 6 to 12 months for small masses, and reserve surgery for symptomatic masses greater than 10 cm given the risk of bleeding complications, spontaneous or traumatic rupture, or malignant transformation [15]. Our patient had a symptomatic mass measuring approximately 15cm, which was a real indication for adrenalectomy.

Laparoscopy is the preferred approach for small masses [16], whereas for large masses greater than 10 cm, open surgery is required [17].

After treatment, EMH requires a long-term monitoring because of the risk of recurrence, as well as prevention by repeated transfusions to maintain a hemoglobin level above 9.5 g / dl.

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