Relato de Caso / Case Report

Regression of extramedullary hematopoiesis with hydroxyurea therapy in β-thalassemia intermedia

Regressão da hematopoese extramedular na talassemia intermédia após terapia com hidroxiuréia

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*Excessive ineffective erythropoiesis in thalassemia intermedia may cause extramedullary hematopoiesis (EMH), resulting in spleen and liver enlargement or masses in several tissues, mainly paravertebrally. Other less frequent locations of diffuse compensatory EMH are kidneys, adrenal glands, breasts, spinal cord, pleura, pericardium, duramater, adipose tissue and skin, although intrathoracic extramedullary hematopoiesis is a rare condition. Management strategies have included radiation and transfusion therapy. Hydroxyurea with transfusion therapy has been associated with clinical regression of EMH in thalassemia. We report an uncommon case of intrathoracic EMH in a patient with β-thalassemia intermedia, that showed significant recovery with HU therapy. Rev. bras. hematol. hemoter. 2006;**28**(1):71-72.*

Key words: *Hemoglobinopathies; extramedullary hematopoiesis; thalassemia; hydroxyurea.*

Introduction

Extramedullary hematopoiesis (EMH) is a compensatory mechanism that occurs in patients with hematological dysfunctions, such as thalassemia major or intermedia and sickle cell anemia.[1] EMH most commonly occurs in organs that have physiological hematopoiesis during embryonic life, especially the liver, spleen and lymph nodes.[2] Other less frequent locations of diffuse compensatory EMH are the kidneys, adrenal glands, breasts, spinal cord, pleura, pericardium, duramater, adipose tissue and skin. Intrathoracic extramedullary hematopoiesis is a rare condition.[2,3]

Transfusions have been successful in reducing EMH in patients with thalassemia intermedia. On the other hand, hydroxyurea (HU), an inhibitor of the cellular enzyme ribonucleotide reductase, has been associated with clinical improvement in sickle cell anemia, but there is limited experience with HU in thalassemia.[4,5]

We present a case of regression of intrathoracic EMH with HU therapy in a patient with β-thalassemia intermedia.

Case report

A 39-year-old woman with β-thalassemia intermedia (genotype IVS1-6/ IVS1-110) was asymptomatic when she suffered a traumatic break of her left scapula. The whole blood count was as follows: hemoglobin 8.8 g/dL; hematocrit 28.0%; mean corpuscular volume 77.0 fl; mean corpuscular hemoglobin 24.0 pg; leukocytes 9.4 x109/L; platelets 635 $x10^9$/L. A chest X-ray demonstrated that there was a break of left scapula and also it showed a middle-lower mediastinal mass.

A thoracic computed tomography (CT) showed a right smoothly-bordered paravertebral mass located on the T3 - T9 levels without bone destruction (Figure 1). The patient was treated with HU at an initial dose of 8.5 mg/Kg/day, with the dose increased every 8 weeks to a final dose of 16.0 mg/Kg/day.

A follow-up CT was obtained one and two years later with EMH regression (Figure 2). There were no side effects during HU treatment.

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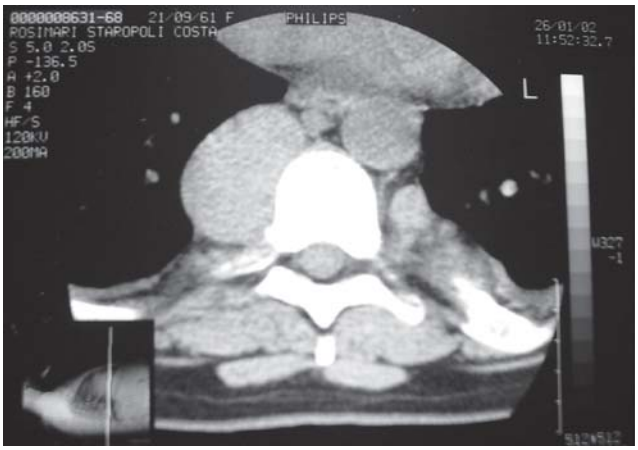

Figure 1. Thoracic computed tomography with right paravertebral mass without bone destruction

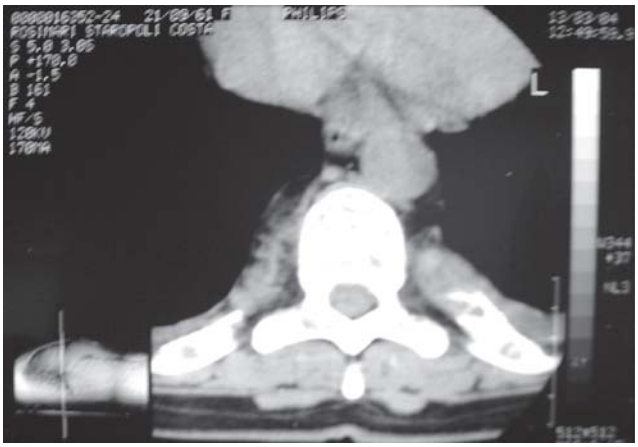

Figure 2. Follow-up thoracic computed tomography after hydroxyurea therapy with reduction of paravertebral mass

Discussion

Excessive ineffective erythropoiesis in thalassemia intermedia may cause EMH, resulting in spleen and liver enlargement or masses in several positions, specifically paravertebrally.

Management strategies have included radiotherapy, blood transfusions and, occasionally, surgery.[1,2] Spontaneous recovery with no therapeutic intervention has also been reported, but may take several months to occur and is subject to frequent recurrence.[6]

HU has been associated with increase of the total percentage of fetal hemoglobin and the number of F-cells, as well as the total hemoglobin concentration and mean corpuscular volume in sickle cell patients.[7] Although there is limited experience with HU in thalassemia, some studies have demonstrated successful regression of EMH with HU therapy.[4,5,7]

Our report calls attention to the fact that asymptomatic EMH patients may have important clinical improvement with conservative therapy such as HU. However, prospective studies to define influence factors to response and rate of EMH recurrence after HU use are necessary.

Resumo

*A excessiva eritropoese ineficaz na talassemia pode causar hematopoese extramedular (HEM), resultando em hepatomegalia, esplenomegalia e massas de tecido hematopoético em diversos tecidos. Localizações de HEM compensatória menos freqüentes são rins, glândulas adrenais, canal medular, pleura, pericárdio, duramáter, tecido adiposo e pele. Entretanto, HEM intratorácica é condição rara. Estratégias terapêuticas incluem radiação e transfusões sanguíneas. O uso de hidroxiuréia concomitante a terapêutica transfusional foi associado à regressão clínica da HEM na talassemia. Nós descrevemos um caso de HEM intratorácica em paciente portadora de talassemia intermédia, com significante regressão do quadro após terapêutica isolada com hidroxiuréia. Rev. bras. hematol. hemoter. 2006;**28**(1):71-72.*

Palavras-chave: *Hemoglobinopatia; hematopoese extramedular; talassemia; hidroxiuréia.*

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