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Case Report

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Paraspinal extramedullary hematopoiesis in alpha-thalassemia with concurrent monoclonal gammopathy of undetermined significance: an uncommon case report

Abstract

Extramedullary hematopoiesis is a rare phenomenon, which consists of the formation of hematopoietic tissue in various parts of the human body due to the existence of a hematological disorder. Alpha–thalassemia is a great example of a disease that can result in extramedullary hematopoiesis. The present study reports an unusual case of a 64-year–old male, who was diagnosed with hemoglobin H disease and secondary paraspinal extramedullary hematopoiesis with a concurrent monoclonal hypergammaglobulinemia. The lesion presented itself as a paraspinal mass of 4,5 cm extending longitudinally between the T9 and T10 vertebral bodies and it was accompanied by mild anemia and splenomegaly. The presence of monoclonal hypergammaglobulinemia and the fact that the patient was until recently a blood donor complicated our final diagnosis, which proved to be the coexistence of hemoglobin H disease and extramedullary hematopoiesis with monoclonal gammopathy of undetermined significance. This is the main reason why the diagnosis, differential diagnosis and treatment options of this rare case are discussed.

Keywords: hematopoiesis, extramedullary, alpha-thalassemia, hemoglobin H, monoclonal gammopathy of undetermined significance

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Abbreviations: EMH, extramedullary hematopoiesis; ESR, erythrocyte sedimentation rate; Hb, hemoglobin; MGUS, monoclonal gammopathy of undetermined significance; MRI, magnetic resonance imaging; PLTs, platelets; RBC, red blood cells; T, thoracic vertebra; WBC, white blood cells

Introduction

Extramedullary hematopoiesis (EMH) functions as a compensatory phenomenon in cases of insufficient bone marrow function and refers to the hematopoiesis that takes place outside the medulla of the bone. It is usually associated with various hematologic disorders, including thalassemia.[1–4] Hemoglobin H disease is a type of a-thalassemia consisting of the loss of 3 genes (--/-a) and is often manifested as a mild anemia with thalassemic morphological changes and the existence of erythrocyte inclusions. Hemoglobin H is a tetramer composed of four beta globin chains and a great decrease in alpha chain availability.[5] In this hemolytic disorder, splenomegaly may occur, as well as mandibular and bone marrow hyperplasia and pathological fractures. Reported herein is the case of a 64-year-old male who suddenly developed a mild anemia, splenomegaly, monoclonal hypergammaglobulinemia and a paraspinal mass and was admitted to our hospital for further investigation. The patient was eventually diagnosed with hemoglobin H disease with concurrent MGUS. The present case report discusses the diagnostic methods, problems and treatment strategies that were applied and seeks to further examine this uncommon clinical manifestation.

Case presentation

The present study reports an unusual case of a 64-year-old male, former blood donor, smoker, heavy alcohol drinker, who, after a fall on the ground followed by intense back pain, underwent a CT scan and a full blood analysis. The patient was admitted to our clinic because of the occurrence of mild anemia, splenomegaly, marked monoclonal hypergammaglobulinemia and the appearance of a paraspinal mass of 4,5 cm extending longitudinally between the T9 and T10 vertebral bodies. On admission his temperature was 36,4 C, blood pressure 150/90 mmHg, respiratory rate 15/min and pulse rate 97/min. His laboratory studies revealed anemia (Hb 9,4 g/dl), normal WBC and PLTs (WBC 8100/μl and PLT 261.000/μl respectively), elevated RBC (5.070.000 /μl), normal ESR (4mm/hr) and slightly increased c-reactive protein (6,15 mg/l) as well as marked monoclonal hypergammaglobulinemia (1,90 g/dl). Serum glucose, electrolytes, renal and hepatic function (except for an elevated γ- Gt attributed to the patient's history of alcohol consumption) were normal. In addition to the above, there was an elevated ferritin level (954 ng/ml). The existence of a paraspinal mass in combination with the monoclonal hypergammaglobulinemia leaded to additional tests, such as hemoglobin electrophoretic studies, MRI, bone marrow scan, bone marrow aspiration and biopsy that contributed to the diagnosis.

Due to the absence of renal insufficiency as well as the absence of plasma cells on the bone marrow examination, the positive bone scan (Tc 99m nanocoloid) showing signs of an overfunctioning bone marrow (Figure 1) and mainly due to a hemoglobin electrophoresis

and an MRI consistent with hemoglobin H disease, a rapid and extremely interesting diagnosis were made.

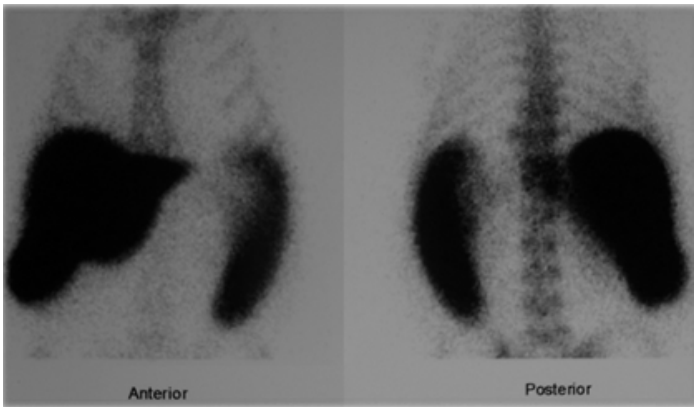

Figure 1 Tc 99m nanocolloid scanning.

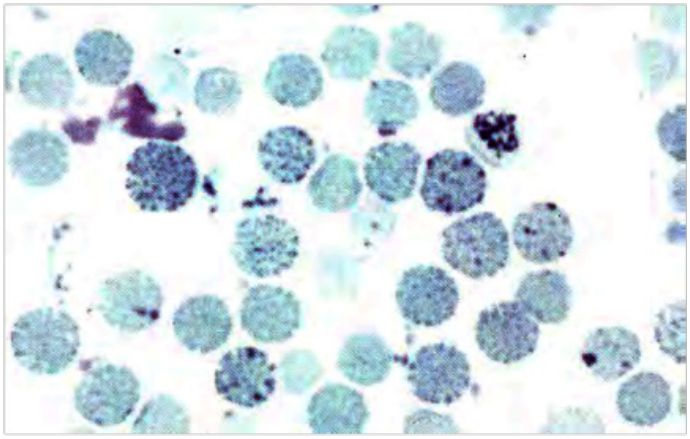

Figure 2 Erythrocyte inclusions.

The hemoglobin H disease was actually diagnosed by the presence of erythrocyte inclusions in the hemoglobin electrophoresis (Figure 2), although the magnetic resonance imaging had already suggested the possibility of the paraspinal mass being an example of extramedullary hematopoiesis (Figure 3). Ultimately, genotypic analysis was performed with the patient' s consent that confirmed the diagnosis.

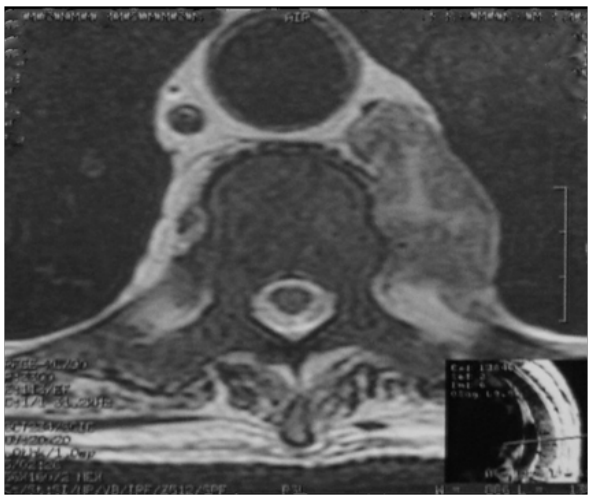

Figure 3 MRI depicting the paraspinal mass.

As far as the monoclonal hypergammaglobulinemia was concerned, since the bone marrow biopsy revealed erythroid hyperplasia and none of the criteria for multiple myeloma were met, it was inevitably attributed to monoclonal gammopathy of undetermined significance. Because no serious symptoms were reported or manifested by the patient, it was suggested that he should remain under close observation and only undergo a blood transfusion, when he ceases to be asymptomatic. Up to this time the patient remains in a good health with a mild anemia demonstrated in this blood analysis and an asymptomatic monoclonal hypergammaglobulinemia characterized as MGUS.

Discussion

For a paraspinal mass the potential clinical differential diagnoses are vast. EMH, however rare, should always be included in the diagnostic process. It is characterized by the presence of hematopoietic tissue outside the bone marrow and is usually associated with hematologic disorders.[6] Typically, the spleen and the liver are affected and occasionally the lymph nodes.[4,6] Sometimes, soft tissue masses can develop in the paravertebral regions, that cause no symptoms at all and are usually diagnosed by chance.[1] The development of extramedullary hematopoiesis in the clinical context of thalassemia is well documented.[1–3,6,7] For this reason, in the presence of anemia in combination with a paraspinal mass the diagnosis of EMH should be considered. It was this specific combination that eventually led to the diagnosis of the hemoglobin H disease, although the presence of the monoclonal hypergammaglobulinemia complicated the diagnostical process.

MGUS occurs in over 3% of individuals over 50 years old and consists of the presence of an asymptomatic serum monoclonal protein.[8] Some of these cases ultimately lead to MM, which signifies the importance of further follow-up in specific circumstances.[9] Our patient underwent a bone marrow aspiration and biopsy in order to exclude the possibility of MM and remains still under regular supervision. As far as the therapeutic options are concerned, it has been observed that most patients with hemoglobin H disease rarely require blood transfusions.[5] In case of a large mass of EMH, recurrent blood transfusions have been shown to result in its shrinkage.[1] However, this comes with the price of iron overload. Radiotherapy is an alternate treatment option, as well as surgical excision, hydroxyurea or a combination of these options.[1,2]

Conclusion

The question remains what is to be done in this kind of situations. Surely these patients should remain under observation. Whether or not they should receive a blood transfusion relays upon the better judgement of the physician in charge, who is after all responsible for weighing the advantages and disadvantages of multiple blood transfusions, especially since many of these patients will later on require chelation therapy in order to treat the iron overload.

Acknowledgments

None.

Conflicts of interest

Authors declare that there is no conflict of interest.

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