EXTRAMEDULLARY BONE MARROW TUMOR IN THALASSEMIA

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This 24-year-old male thalassemia major patient was brought to our attention to be evaluated for marrow transplantation. He had been receiving regular transfusional treatment since the age of 30 months and was currently getting 2 units of pure red blood cells every two/three weeks. He had received more than 600 units of blood. Regular subcutaneous deferoxamine therapy had been continuing since the age of 15 years.

Routine chest X-rays showed a mediastinal mass caused by extramedullary erythropoietic

tissue (Figures 1 and 2). This tumor mass was not causing any symptoms. The diagnosis was subsequently confirmed by chest computed tomography in his hometown.

Extramedullary erythropoiesis is an infrequent but well-recognized complication of thalassemia intermedia. It can also occur in inadequately transfused thalassemia major patients, producing bizarre radiological images. With the advent of the hyper-transfusion regimen this finding has become uncommon in developed countries. Nevertheless it is still encountered in

ABSTRACT

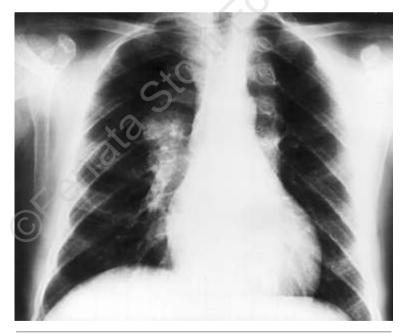


Figure 1. Posteroanterior chest roentgenogram: mediastinal tumor mass due to extramedullary erythropoietic tissue. A homogeneous mass with well-defined borders is clearly visible in the right paravertebral area.



Figure 2. Lateral chest roentgenogram: direct view of the mass of erythropoietic tissue in the posterior mediastinum.

selected cases, where it indicates the need for a more intense transfusional regimen.

This patient was excluded from the transplant program because of the presence, in addition to the mediastinal tumor, of severe iron overload and liver cirrhosis. He returned to his hometown with instructions to follow a high transfusion- and chelation regimen.

References

- Erlandson ME, Brilliant R, Smith CH. Comparison of sixtysix patients with thalassemia major and thirteen patients with thalassemia intermedia: including evaluation of growth, development, maturation and prognosis. Ann NY Acad Sci 1964; 119:727-31.
- Korsten J, Grossman H, Winchester PH, Canale VC. Extramedullary hematopoiesis in patients with thalassemia anemia. Radiology 1970; 95:257-61.