

SHORT COMMUNICATIONS

Presence of Erythrocytosis in a Patient with Diagnosis of Beta Thalassemia Trait

Ahmet Ifran, Kürşat Kaptan and Cengiz Beyan

Department of Hematology, Gülhane Military Medical Academy, Ankara, Turkey

INTRODUCTION

The patient, a 68 years old woman, was admitted to our clinic with the complaints of itching on the lower legs, easy fatigue and night sweating. She was previously evaluated in the hematology clinic 4 years ago as an outpatient and was diagnosed as a case of β -thalassemia trait with microcytosis, HbA₂ level 4.7%; red blood cell count $9.31 \times 10^{12}/L$, hemoglobin $>15.5 \text{ g dl}^{-1}$, hematocrit 59% and white blood cell count $> 18.5 \times 10^9/L$ on the last admission. The patient was reevaluated for polycythemia. On physical examination she had a palpable spleen and plethora, other findings were normal. Her arterial O₂ saturation was 96%. Erythropoietin level was 9.8 mU ml^{-1} (normal range $9-30 \text{ mU ml}^{-1}$). The diagnosis of polycythemia rubra vera was made according to these findings. Doppler ultrasonography of the lower extremities was normal. Hematocrit levels restored to levels below 50% with two phlebotomies and the patient was discharged with low dose aspirin.

A substantial number of hemoglobinopathies with increased oxygen affinity that cause erythrocytosis are reported [1-3]. The red cell count more than 9 millions in our patient is hard to explain with a simple condition of thalassemia trait. The complaints related to hyperviscosity made it logical to search for a myeloproliferative condition. The best way to distinguish both conditions or in other words to rule out a myeloproliferative condition is to determine the erythropoietin (EP) level. As described in many reports, thalassemic patients even though they are just carriers for thalassemia gene have higher levels of EPO when compared to normal population [4, 5]. Erythropoietin level in our patient was lower than of

normal range which suggests that the bone marrow is no more under the control of erythropoietin. We have excluded the possible reasons of secondary erythrocytosis in our patient. The presence of erythrocytosis in a patient with diagnosis of β -thalassemia trait draws no attention at first evaluation, but if the symptoms of hyperviscosity are present, the patient should be evaluated further for a concurrent myeloproliferative disorder.

REFERENCES

1. Ntazi, G.A., S. Shibata, 1986. Hemoglobinopathies due to abnormal functional properties of hemoglobin molecule. Stable abnormal hemoglobins with high O₂ affinity and erythrocytosis. *Kawasaki Med. J.*, 12: 1-12.
2. Harteveld, C.L., J.H. Groeneveld, B. van Dam, P. Van Delft, N. Akkerman, S. Arkesteijn, P.C. Giordano, 2005. Hb Zoeterwoude [β 23(B5) Val \rightarrow Ala]: a new beta-globin variant found in association with erythrocytosis. *Hemoglobin*, 29: 11-17.
3. Pagano, L., A.M. Salzano, V. Carbone, D. Iannelli, A. Viola, F. Pollio, L. Prossomariti, O. David, G. Ricco, P. Pucci, 2004. Hb Cardarelli [β 86(F2)Ala \rightarrow Pro]: A new unstable and hyperaffine variant in association with β (+)-thalassemia. *Hemoglobin*, 28: 103-115.
4. Chaisiripoomkere, W, S. Jootar and S. Chanjarunee, 1999. Ungkanont A. Serum erythropoietin levels in thalassemia major and intermedia. *Southeast Asian J. Trop. Med. Public Health*, 30: 786-788.
5. Vedovato, M., G. Salvatorelli, M. Taddei Masieri and C. Vullo, 1993. Epo serum levels in heterozygous β -Thalassemia. *Haematologia (Budap)*, 25: 19-24.