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Ironoverload and Malignancies in patients with Haemoglobinopathies : a single center experience.

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Background

Thalassemias are a group of hereditary blood disorders characterized by abnormalities in the synthesis of the haemoglobin chains. The pathophysiology of thalassemia is, to a certain extent, associated with the generation of labile plasma iron (LPI) in the pathological red blood cell (RBC). Recent literature considers the possible key role of iron as a carcinogenic agent and supports the hypothesis that the cases of malignancies in thalassemic patients could be ascribed to elevated levels of ferritin. The aim of our study was to determine the frequency, characteristics, and pattern of malignancies in a group of patients with β thalassemia major (BTM), β thalassemia intermedia (BTI) and Hemoglobinosis H (HbH).

Patients

The patients were recruited from “Thalassemia and Hemoglobinopathies” Center of University Hospital of Messina (Sicily). Seventy-four patients with BTM (M=41, F=33); 14 with BTI (M=8, F=6), and 3 sisters with HbH were included in the study. The standard of care for our BTM patients involved regular follow-up, blood transfusion every 2–3 weeks, and iron chelation therapy. Patients with BTI and HbH did not need for blood transfusion or needed only two or three transfusions per year.

All thalassemic patients were subjected to genotypic analysis.

Results

Media age of our patients was distributed as following: 35.1 years ±8.0 in BTM patients, 42.2 years±6.0 in BTI subjects, and 30.4 years±4.2 in the 3 sisters with HbH. We found 5 cases of cancer in the group of 91 patients (5.5%): 4 malignancies in the 74 BTM patients (5.4%) (mean age of 35.6±8.4 years); no case cancer in the group of BTI (0%), and 1 case in the 3 HbH patients (33.3%). In particular, 3 cases of thyroid papillary carcinoma (75%) and 1 case of gastric cancer (25%) were found in BTM patients. A thyroid papillary carcinoma was found in one female with HbH.

Mean Hb and ferritin concentrations of patients with malignancies were distributed as following: 9.4±0.2 g/dl and 1521±163 ng/ml in the 4 BTM subjects; 10.4±0.2 g/dl and 421±98 ng/ml in BTI group; and 9.0±0.6 g/dl and 921±101 ng/ml in the 3 sisters with HbH.

Conclusion

This study in a Sicilian cohort of patients with BTM , BTI and HbH showed that the coexistence of malignancy and thalassemia is not rare and that it could be correlate to elevated levels of ferritin. Some types of cancer, such as thyroid papillary carcinoma, seem to occur more frequently than other types in patients with thalassemia.