THE ROLE OF THE SPLEEN IN REMOVAL OF ABNORMAL RED CELLS
IN THALASSEMIC PATIENTS

BY
SUPAPORN SUWIWAT

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SUPAPORN SUWIWAT

Department of Pathobiology, Faculty of Science, Mahidol University
Rama VI Rd., Bangkok 10400, Thailand

ABSTRACT

The role of spleen in removal of the aged, abnormal and pathologic red blood cells in human as well as in experimental animals is rather well established. This study is, however, the first one in pursuing and emphasizing the precise role of the spleen in removing and producing abnormal red cells in thalassemic patients. In order to accomplish the aim set, red blood cells from the peripheral blood, red cells from the splenic artery, and red cells from the splenic vein were collected, morphologically analyzed and quantitated using scanning electron microscopy in patients with various types of thalassemia. The splenic events were also studied by histopathological and transmission electron microscopical examination of the splenic tissue. This reported group of patients composed of β-thalassemia/Hb E (9 cases), homozygous β-thalassemia (2 cases), and α-thalassemia/Hb CS (1 case). These 12 patients showed typical signs
of hypersplenism and underwent the splenectomy at the time of study. The study therefore composed of 3 phases: red cell profile in presplenic phase (SEM); intrasplenic event (LM, SEM and TEM); and red cell profile in postsplenic phase (SEM).

Intra-splenic phase as studied by light microscope revealed prominent and marked dilatation of the sinusoidal spaces, hypertrophy and hyperplasia of RE and sinus lining cells. Iron deposition was detected in RE cells, basement membrane, and of interest in the sinus endothelial lining cells. Extramedullary hematopoiesis was readily noticeable in every case. Fibrosis was moderately observed. Details of culling and cellular fragmentation processes in the spleen were electron microscopically visualized. By the same technique, both forms of iron associated granules, ferritins and hemosiderins, were observed in lysosomal system and also in the cytoplasm of RE and endothelial cells.

The analysis of percentage of discocytes in the pre-splenic (splenic artery) as compared to the post-splenic phase (splenic vein) revealed the following results: 42.51 ± 10.04 to 51.09 ± 11.39 with P value of 0.004 in β-thalassemia/Hb E; 46.60 to 61.45 in homozygous β-thalassemia; and 52.40 to 61.40 in α-thalassemial/Hb CS. In addition, red cells having high surface area to cell volume ratio (codocyte, leptocyte and torocyte) and cells having low surface area to cell volume ratio (spherocyte and stomatocyte) were higher in the splenic arteries than in the splenic veins in patients with β-thalassemia/Hb E with P values of 0.082 and 0.020 respectively. The immediate and important role of spleen in removal of the abnormal
red blood cells is therefore, emphasized by the above findings.

The inverse correlation between percentage of discocyte and percentage of cells with high and low surface area to cell volume ratio in the splenic veins of patients with \( \beta \)-thalassemia/Hb E strengthened the belief that spleen would allow only the red cells with optimal surface area to cell volume ratio to successfully pass through its microcirculation with relative ease.

Higher percentage of cells with low surface area to cell volume ratio in splenic vein than in the peripheral blood of patient with \( \beta \)-thalassemia/Hb E suggested the existence of other non-splenic site of abnormal red cell destruction or trapping. Liver was likely to be one such possible site.

Obvious positive correlations between splenic weights and the percentage of the abnormal red cells in the splenic vein were demonstrated in \( \beta \)-thalassemia/Hb E (r value of 0.679 and P value of < 0.05) and in all cases (r value of 0.724 and P value of < 0.005). This findings indicated the splenic role in formation of some abnormal red cells particularly those resulted from the process of cellular fragmentation namely, dacryocyte, keratocyte, and schizocyte.

Both open and close types of circulation are known to exist in the human spleen. Relatively higher percentage of fragmented red cells in the splenic veins of larger spleens implies predominancy of the open pathway of circulation in the spleens of thalassemic patients in this series of study.