

## PAROXYSMAL NOCTURNAL HEMOGLOBINURIA

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### INTRODUCTION

Paroxysmal Nocturnal Hemoglobinuria is a rare hemolytic anemia, affecting young adults. We report a case and present the clinical and laboratory data of this peculiar disorder. Its complications and treatment are also discussed.

### CASE REPORT

A 32 years old school teacher was admitted in February, 1994 in Medical B unit Lady Reading Hospital with a two years history of fatigue, faintness and palpitations. Clinically he was anemic and mildly jaundiced. Spleen was moderately enlarged. We started investigating his anaemia. Laboratory data revealed hypochromic microcytic anaemia with marked reticulocytosis. Bone marrow was aspirated from the posterior iliac crest. It was hyper cellular and iron stores were absent. On urine examination urobilinogen was present in excess as well as haemoglobin in late hours of night. Glucose-6-phosphate dehydrogenase and Pyruvate kinase screening test were found negative. RBC haemolysis started at 0.6% NaCl and completed at 0.35% NaCl. Coomb's test and Ham's test were negative. On clinical grounds and laboratory data, it was diagnosed as "Hereditary Spherocytosis" and was discharged on Folic acid. We followed him two months later in OPD. He was still anemic and there was reticulocytosis. In order to know the

main site of RBC destruction "Splenic Scan" with denatured (Heat) RBCs was performed. It showed large spleen with almost all labelled RBCs taken up into it. There was no activity in bone marrow and liver. In June 1994 splenectomy was performed. Histopathology of Spleen showed Haemosiderin laden macrophages. Patient's condition did not improve even after splenectomy.

In August, 1994 he was referred to Sheikh Zayed Hospital. There Ham's test was positive. Haemosiderin granules were present in urine deposits along with epithelial cells. Neutrophil Alkaline Phosphatase score was low. Patient was diagnosed as paroxysmal Nocturnal haemoglobinuria.

### DISCUSSION

PNH is an acquired intrinsic abnormality of the RBC membrane which causes increased susceptibility to lysis by complement. The incidence is 2-5 percent of all acquired Haemolytic anemias. Certain proteins like acetylcholinesterase and decoyaccelerating factor are attached to cell membrane. Deficiency of these is responsible for membrane. It begins insidiously in a previously healthy person. Hemolysis is often increased at night, because CO<sub>2</sub> retention during night leads to a slight fall in plasma pH. This facilitates activation of complement.

there by causing the patient's urine to be very dark in color. Laboratory findings include anemia with reticulocytosis, haemosiderinuria and excess urinary urobilinogen, positive HAM's test and sucrose lysis test and low leucocyte Alkaline Phosphates score. The complications include Thrombosis, Iron deficiency and development of acute leukaemia. Literature shows a high maternal mortality among PNH patients (5.8%). The most common cause of death is Liver vein thrombosis. No definite treatment is available. Blood transfusion is the mainstay of treatment. Androgens particularly Methyltestosterone and Oxymethalone stimulate erythropoiesis. In a young patient with an HLA-compatible sibling, bone marrow transplantation is considered.

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