A CASE OF RECURRENT JAUNDICE

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Abstract:
13 YEAR OLD BOY PRESENTED WITH HISTORY OF FATIGUE, JAUNDICE AND COLA COLOURED URINE OF 2 MONTHS DURATION PRECEDED BY FEVER. HE HAD 2 SIMILAR EPISODES IN THE PAST AND HIS BROTHER IS ALSO SUFFERING FROM SIMILAR ILLNESS. BASED ON THIS HISTORY WE INVESTIGATED HIM, HE WAS FOUND TO HAVE ANEMIA, MILDLY RAISED BILIRUBIN AND LDH LEVELS. HIS PERIPHERAL SMEAR SHOWED OCCASIONAL BITE CELLS. THEN WE WENT ON TO DO A G-6PD ASSAY AND WAS FOUND TO BE SIGNIFICANTLY LOW. THE BOY WAS DIAGNOSED TO HAVE HEMOLYTIC ANEMIA DUE TO G-6 PD DEFICIENCY AND WAS TREATED SYMPTOMATICALLY.

Keyword: RECURRENT JAUNDICE, ANEMIA, BITE CELLS, G-6PD DEFICIENCY.

A 13 YEAR OLD BOY PRESENTED WITH HISTORY OF FATIGUE, JAUNDICE AND PASSING COLA COLOURED URINE OF 2 MONTHS DURATION. THE EPISODE WAS PRECEDED BY FEVER WITH NO RIGORS, HEADACHE OR VOMITING. HE HAD SIMILAR EPISODES WHICH WERE SELF LIMITING IN 2001 AND 2004. HIS YOUNGER BROTHER ALSO HAS SIMILAR HISTORY IN THE PAST. HIS ELDER BROTHER DIED IN EARLY NEONATAL PERIOD FOR UNKNOWN REASONS. HIS PARENTS AND OTHER FAMILY MEMBERS WERE APPARENTLY NORMAL. ON EXAMINATION HE WAS PALE AND MILDLY ICTERIC. AT THIS POINT OF TIME WE HAD 2 DIFFERENTIAL DIAGNOSIS, HEMOLYTIC ANEMIA PROBABLY AN ENZYME DEFICIENCY OR PARAXYSOMAL NOCTURNAL OR COLD HEMOGLOBINURIA AND MALARIA. ON INVESTIGATING FURTHER HEMOGLOBIN WAS 8.5gm /dl, bilirubin was 2.8 mg/dl WITH PREDOMINANT INDIRECT FRACTION. HIS LDH WAS 263 U/L, PERIPHERAL SMEAR REVEALED ANISOCYTOSIS.
POIKILOCYTOSIS WITH OCCASIONAL BITE CELLS AND NO PARASITES. USG ABDOMEN WAS NORMAL. THEN WE WENT ON TO DO A G-6PD ASSAY AND IT WAS FOUND TO BE SIGNIFICANTLY LOW, IT WAS 1 U/gm OF HEMOGLOBIN [REF RANGE WAS 4.6-13.5]. OTHER PARAMETERS INCLUDING PT, APTT, BT, CT WERE NORMAL. SINCE IT WAS A RECURRENT, FAMILIAL JAUNDICE PRECIPITATED BY INFECTION AND G-6PD LEVEL WAS LOW EVEN DURING THIS PERIOD IMMEDIATELY AFTER HEMOLYSIS WHERE THE LEVEL MAY BE NORMAL DUE TO THE RELEASE OF FRESH RBCS, HE WAS DIAGNOSED TO HAVE HEMOLYTIC ANEMIA DUE TO G-6PD DEFICIENCY. HE WAS TREATED SYMPTOMATICALLY.

DISCUSSION
G-6PD [GLUCOSE-6-PHOSPHATE DEHYDROGENASE] DEFICIENCY IS COMMON IN TROPICAL PARTS ESPECIALLY IN AFRICA, MIDDLE EAST AND SOUTH EAST ASIA WHERE IT MAY BE HAVING A SURVIVAL ADVANTAGE ALSO. IT IS X LINKED AND MOST OF THEMUTATIONS ARE MISSENSE MUTATIONS. THIS RESULTS IN DECREASED ENZYME ACTIVITY IN OLD RBCS. EMBDEN MEYERHOF PATHWAY PRODUCES NADPH WHICH VIA REDUCED GLUTATHIONE WILL PROTECT THE RBC AGAINST OXIDANT STRESS. THIS IS ALTERED IN G-6PD DEFICIENCY. IN THE PRESENCE OF OXIDANT STRESS HEMOGLOBIN IS DENATURED AND PRODUCE HEINZ BODIES, THOSE RBCS WITH HEINZ BODIES ARE REMOVED BY SPLEEN AND RESULTS IN BITE CELLS. THE COMMON TRIGGERS ARE INFECTIONS, DRUGS AND FAVA BEANS. DRUGS WITH DEFINITE RISKS ARE ANTIMALARIALS LIKE PRIMAQUINE, DAPSONE, COTRIMOXAZOLE, NALIDIXIC ACID, NAPHTALENE, METHYL ENE BLUE, ACETANILIDE. POSSIBLE AGENTS ARE CHLOROQUINE, CIPROFLOXACIN, HIGH DOSE ASPIRIN MORE THAN 3 gm, VIT K.

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I APOLOGISE THAT I COULD NOT CAPTURE THE BITE CELLS CLEARLY IN MY CAMERA AND BITE CELLS ARE NOT SEEN CLEARLY AMONG THE RBCS IN SMEAR PHOTO TAKEN.