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Paroxysmal Cold Hemoglobinuria: A Rare Case Presentation

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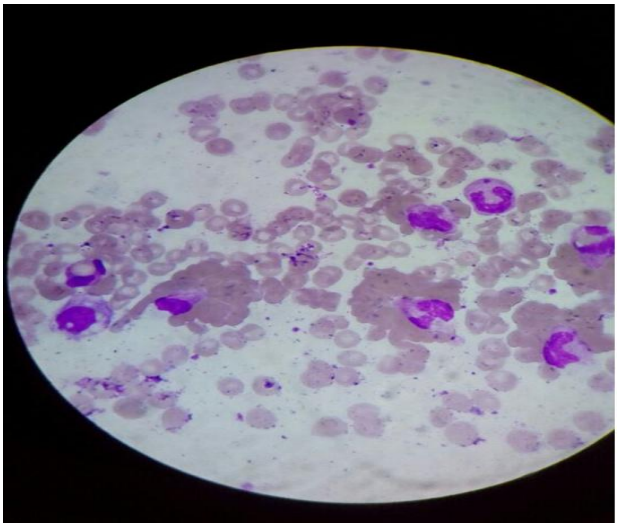
Background
Paroxysmal cold hemoglobinuria (PCH) is a rare cause of acquired autoimmune hemolytic anaemia (AIHA) commonly seen in children post viral infections caused by complement dependent cold acting autoantibodies that produce intravascular hemolysis and hemoglobinuria in vivo and gives rise to Donath-Landteiner (D-L)reaction in vitro.

Case Report
A 25 yr old female patient with last menstrual period 15days back came with complaints of fever with chills and generalise weakenss for 4 days, vomiting and loose stools for 2 days and reddish brown discolouration of urine more during the morning hours for 2 days. On examination patient is febrile pallor ++, icterus++ with no significant findings on systemic examination. Investigations reveal wbc 9700cells/cumm, hb 4mg/dl, platelets 2.24cells/cumm, total bilirubin 5.3mg/dl, LDH 2262U/L, urine analysis was positive for RBCs , HCV/Hbsag/VDRL serology was negative, ultrasound abdomen and pelvis were WNL DIRECT COOMBS TEST:++ with polyspecific anti IGg and anti C3d with ice cold saline wash and negative with routine room temperature saline. PERIPHERAL smear: RBC: few nucleated RBC of late erythroblastic type are seen. WBC: mild neutrophilic predominance. Many neutrophils are showing erythrophagocytosis. RBC rosette are seen surrounding engulfing neutrophils .peripheral smear, elevated LDH and direct coombs test positive for anti C3d with poly specific antisera and characteristic clinical findings are S/O AIHA with PCH as differential diagnosis. D-L autoantibody test was positive confirming the diagnosis. Due to unavailability of specific blood group, she was referred to higher centre for further management. The repeated hemoglobin in a span of 48 hours dropped down to 1.8mg/dl and in an hour patient died.



Conclusion

PCH is a rare disease, early diagnosis based on clinical and pathological profile and aggressive treatment with red cell transfusion and immunosupression may help in prevention of any untoward events.