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Paroxysmal nocturnal hemoglobinuria: A multicenter study from Saudi Arabia

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Background & Aim: Paroxysmal nocturnal hemoglobinuria (PNH) is a rare acquired clonal hematopoietic stem cell disorder, characterized by the deficiency of glycophosphatidylinositol (GPI) that anchors proteins in cell membranes. PNH is manifested variously with hemoglobinuria, thrombosis, or bone marrow failure. This multi-central retrospective study was aimed at assessing the incidence and characteristics of patients diagnosed with PNH in Saudi Arabia.

Methods: Patients referred for PNH diagnosis at King Faisal Specialist Hospital and Research Centre, Riyadh during the 3-year period (2007-2009) and the 2-year period (2012-2013), also at King Faisal Specialist Hospital and Research Centre, Jeddah, King Abdulaziz Medical City (NG), Riyadh, King Abdulaziz Medical City (NG), Jeddah data during the last year 2014, were included in the analysis. Peripheral blood samples were used for multi-parametric flow cytometry analysis based on fluorescent inactive aerolysin (FLAER), and the markers, CD235a and CD59 on red blood cells (RBCs), and CD14, CD45, CD64, CD24, and CD15 on white blood cells (WBCs) exclusively monocytes and granulocytes. Univariate analysis of the disease characteristics was performed.

Results: Of the 843 samples submitted for PNH screening, 40 were positive (4.7%). Of the 40 patients analyzed, 22 patients (55%) presented with aplastic anemia, 4 patients (10%) with pancytopenia, and 3 patients with thrombosis (7.5%) one with Budd-Chiari syndrome and 2 with portal vein thrombosis. Hemolytic anemia represented 5% (2 patients) of all cases. Other positive cases were presented with unrelated diseases such as immune thrombocytopenic purpura (ITP) in 2 cases (5%) and non-specific diagnosis in the rest. All samples showed type II and III GPI-deficient clones with a median clone size of 12 (range, 0.04%-85%) in the RBCs, and 56 (range, 1%-100%) in WBCs (monocytes and granulocytes).

Conclusions: This study confirms the rarity of PNH and its predominant presentation as aplastic anemia or thrombosis in a Saudi Arabian population, similar to the worldwide incidence.

Biography

Salem Khalil completed his MBBS at King Saud University Riyadh in 1984; fellowship of the Royal College of Pathologists of Australasia, Australia in 1992 and; fellowship in Molecular Hematopathology at MD Anderson Cancer Center, Huston, Texas, USA.

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