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Neutropenia in Patients with Primary Antibody Deficiency Disorders

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Abstract:

Neutropenia is characterized by decrease in the absolute number of circulating neutrophils and an increase susceptibility to infections. The current study was performed in order to explain the clinical and laboratory findings of patients with antibody deficiency disorders associated neutropenia. The patients' records of 19 neutropenic cases out of 207 patients with antibody deficiencies, who had been referred to Children's Medical Center and enrolled in Iranian primary immunodeficiency registry, were reviewed. Nineteen cases (14 male and 5 female), with a mean age of 10.7 ± 5.7 years, were associated with neutropenia (9.2%). The disorders with associated neutropenia were Hyper IgM syndromes (3 of 8), Common variable immunodeficiency (13 of 109), and X-linked agammaglobulinemia (3 of 45). The median age for the onset of disease and diagnosis age were 15 months (1-134) and 3.8 years (6 months-13 years), respectively. The most common infections during the course of illness were pneumonia (13 cases), diarrhea (12 cases), oral candidiasis (9 cases), otitis media (6 cases), sinusitis (6 cases), cutaneous infections (5 cases), and abscess (5 cases). Other less frequent infections were: conjunctivitis, oral ulcers, meningitis, and osteomyelitis. Three neutropenic patients died because of recurrent infections. Neutropenia may occur in any of the primary immunodeficiency disorders. Persistent or severe infections always pose a supposition, which deserves further evaluation for detecting an underlying immune deficiency syndrome and neutropenia, since a delay in diagnosis may result in a serious organ damage or even death of the patient.

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