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Acta Medica Iranica
2009;47(4) : 37-40

Clinical and Laboratory Findings in Iranian Children with Cyclic Neutropenia

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Abstract:

Cyclic neutropenia is a rare immunodeficiency syndrome, characterized by regular periodic oscillations in the circulating neutrophil count from normal to neutropenic levels through 3 weeks period, and lasting for 3-6 days. In order to determine the clinical features of cyclic neutropenia, this study was performed.

Seven patients with cyclic neutropenia (3 males and 4 females), who experienced neutropenic periods every 3 weeks (5 with severe and 2 with moderate neutropenia), were investigated in this study. They had been referred to Iranian Primary Immunodeficiency Registry during 23 years (1980-2003).

The range of patients' ages was from 7 to 13 years (median 11 years). The median age at the onset of the disease was 12 months (1 month- 2 years) and the median age of diagnosis was 2 (1.5-5) years, with a median diagnosis delay of 1 year (2 months- 5 years). Neutropenia was associated with leukopenia (3 patients), anemia (3 patients), and thrombocytopenia (1 patient). Patients were asymptomatic in healthy phase, but during the episode of neutropenia suffered from aphthous ulcers, abscesses and overwhelming infections. The most commonly occurred manifestations were: otitis media (6 cases), oral ulcers (5 cases), abscesses (4 cases), pneumonia (3 cases), diarrhea (3 cases), oral candidiasis (3 cases), cutaneous infections (2 cases), and periodontitis (2 cases). One of these patients subsequently died because of recurrent infections.

Unusual, persistent or severe infections should be the initiating factors to search for an immune deficiency syndrome such as cyclic neutropenia, because a delay in diagnosis may result in chronic infection, irretrievable end-organ damage or even death of the patient.

TUMS ID: 3955

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