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"Clinical and Radiological Aspects of Chronic Granulomatous Disease in Children: A case Series from Iran "

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Abstract:

Chronic granulomatous disease (CGD) is a rare disorder of phagocytes, predisposes patients to bacterial and fungal infections. The main purpose of this study was to determine the clinical, radiological, pathologicial features, outcome and response to treatment of children with CGD. Thirteen patients with CGD, who had been referred to National Research Institute of Tuberculosis and Lung Disease (NRITLD), were reviewed during a 6 year period (1999-2005). There were 10 (76%) male and 3(24%) female cases. The median age of the patients was 9 years (1 month-12 years).Family history of CGD was reported by 7 patients. The median diagnostic age was 8 years, with a diagnostic delay of 4.5 years. The most common manifestations of CGD were pulmonary infections and skin involvement, followed by generalized lymphadenopathy. The most common radiological findings were multiple lymphadenopathy in mediastinal region and fibrotic changes in lung fields. Two patients died of pulmonary infections. Based on the results of this research, immunologic evaluations especially evaluation for CGD is highly recommended in children suffering from recurrent pulmonary infections, cutaneous or hepatic abscesses, or infections caused by uncommon pathogens. Early diagnosis and prophylactic treatment both, prevent further development of the lesions, irreversible complications and decreasing mortality and morbidity rates in children suffering from CGD.

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