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RESPIRATORY MANIFESTATIONS OF CHRONIC GRANULOMATOUS DISEASE; A CLINICAL SURVEY OF PATIENTS FROM IRANIAN PRIMARY IMMUNODEFICIENCY REGISTRY

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Abstract:

Chronic granulomatous disease represents a group of inherited disorders of phagocytic system wherein recurrent infections are seen at different sites especially in the respiratory system. To determine the clinical spectrum of respiratory manifestations in chronic granulomatous disease patients, in this retrospective study, we used data from Iranian Primary Immunodeficiency registry. The diagnosis was based upon WHO criteria for chronic granulomatous disease. We reviewed the records of 38 patients (26 males, 12 females), related to 33 families, 73% of whom were consanguineous. The median age at the time of the study was 12yrs (3mo-22yrs). The median onset age of symptoms was 4 months (lmo-12yrs), and that of diagnostic age was 5yrs (lmo-20yrs), with a diagnostic delay of 4.15 yrs, on an average. Sixty three percent of our patients had respiratory involvement in the course of their illness, including pneumonia (18pts, 75%), tuberculosis (llpts, 46%), aspergillosis (3pts, 12.5%), pulmonary abscess (3pts, 12.5%), and bronchiectasis (lpt,4%). Only 4 of our patients presented with respiratory problems as their first manifestation. Lymph nodes were the first common site and the lungs were the second sites of involvement in chronic granulomatous disease patients; however, it is noteworthy that only in a few of our patients, it was the first manifestation of the disease. Thus special attention should be paid to the pulmonary complications while managing this disease.

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