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Study of α -1-antitrypsin phenotypes frequencies in patients with primary antibody deficiencyFazlollahi, M.R.^{ab}, Aghamohammadi, A.^a, Hosseini, R.F.^b, Lotfi, A.S.^c, Khoshdel, A.^c, Farhodi, A.^a, Movahedi, M.^a, Gharagozlou, M.^a, Mozaffari, H.^a, Zandieh, F.^a, Mansouri, M.^a, Ghaffari, J.^d, Rezaei, N.^a^a Department of Allergy and Clinical Immunology, Children Hospital Medical Center, Tehran University of Medical Sciences, Tehran, Iran^b Department of Allergy and Clinical Immunology, Mashhad University of Medical Sciences, Mashhad, Iran^c Department of Clinical Biochemistry, Tarbiat Modarres University, Tehran, Iran^d Department of Pediatrics, Mazandaran University of Medical Sciences, Sari, Iran

Abstract

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Primary antibody deficiencies are the most frequent primary immunodeficiency disorders. Bronchiectasis as a feature of these disorders may be developed due to some factors such as α -1-antitrypsin deficiency. In order to determine the prevalence of two common α -1-antitrypsin deficiency alleles (PI*Z and PI*S) in Iranian patients with antibody deficiency, this study was performed. The prevalence of PI*M, PI*S, and PI*Z allele combinations was determined in 40 patients with primary antibody deficiency (with and without bronchiectasis) and compared with 60 healthy control subjects. Phenotyping was performed by isoelectric focusing. The phenotype frequencies among patients were as follows: M in 92.5%, S in 2.5% and Z in 5%. There was no significant difference in distribution of alleles or phenotypes between patients and control subjects. Moreover, no significant difference was found between patients with and without bronchiectasis. We did not find evidence to support an association between α -1-antitrypsin phenotypes and primary antibody deficiencies in a small, controlled study. Larger studies will be required to clarify the relationship between α -1-antitrypsin genotype and susceptibility to bronchiectasis in patients with antibody deficiency. Copyright © 2006, Iranian Journal of Allergy, Asthma and Immunology. All rights reserved.

Reaxys Database Information

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Allele; Alpha-1-Antitrypsin; Antibody Deficiency; Bronchiectasis

Indexed Keywords

EMTREE drug terms: alpha 1 antitrypsin

EMTREE medical terms: alpha 1 antitrypsin deficiency; article; bronchiectasis; clinical article; controlled study; female; gene frequency; human; humoral immune deficiency; isoelectric focusing; male; phenotype; prevalence; allele; genetics; immune deficiency; immunology

MeSH: Alleles; alpha 1-Antitrypsin; Bronchiectasis; Female; Gene Frequency; Humans; Immunologic Deficiency Syndromes; Male

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