Journal of
Endocrinology and Thyroid Research
ISSN: 2573-2188

Juniper Publishers
key to the Researchers

Commentary
Volume 2 Issue 3 - June 2017
DOI: 10.19080/JETR.2017.02.555586

J Endocrinol Thyroid Res

Idiopathic Hypereosinophilic Syndrome: Etiology, Diagnosis, Manifestations and Management

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Submission: June 08, 2017; **Published:** July 06, 2017

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Introduction

Hypereosinophilia is a clincial condition that can classified into secondary, clonal and idiopathic. Secondary hypereosinophilia is caused by tissue helminthic infection, parasitic infestations, atopy, allergic reactions, vasculitis, or drugs like allopurinol and carbamazepine. Clonal hypereosinophilia represents neoplastic proliferation of eosinophils as part of an underlying stem cell-derived myeloid malignancies and can accompany all types of leukemia and lymphoma [1,2]. Idiopathic hypereosinophilic syndrome (IHS) represents a multisystem disorder defined by sustained eosinophilia; 1.5 x 10(9)/L or greater for at least 6 months (a shorter duration is acceptable in the presence of symptoms that require eosinophil-lowering therapy), of an undetectable cause with evidence of organ system dysfunction (cardiovascular system, skin, central and peripheral nervous system, gastrointestinal tract, eyes). Endomyocardial fibrosis is considered the most classic and known complication of prolonged IHS. Further, skin lesions, multiple erythematous indurated plaques, can be the dominating and/or presenting symptom in about 50% of patients [1-5]. Diagnosis of IHS implies that both secondary and clonal eosinophilia have been ruled out as possible diagnoses. For example, the more accurate term to describe eosinophilia associated with clonal or phenotypically abnormal lymphocytes is lymphocytic variant hypereosinophilia, not lymphocytic variant IHS. Further, rare instances of congenital eosinophilia must be considered in pediatric cases [2]. In 1997, it was previously mentioned that over expression of IL-5 in transgenic mice was shown to lead to both peripheral blood eosinophilia and tissue eosinophilia [5]. Later, in 2007, a population of aberrant T cells that secretes interleukin-5 have been identified in a subset of patients, indicating the existence of lymphocyte-mediated hypereosinophilia. Further, the characterization of specific genetic alterations linked to clonal eosinophilia has been achieved. The most frequently encountered genetic aberrancy is the cryptic FIP1-like 1/platelet-derived growth factor receptor alpha (FIP1L1-PDGFRA) fusion transcript, which results in an eosinophilic, myeloproliferative disorder [6]. In addition, Further, BCR-ABL1 was recently suggested to be screened in those patients [7]. It's noteworthy that manifestations of IHS can be so diverse and may include among others: cholecystitis and recurrent gastroenteritis [8], monoarticularsynovitis with synovial fluid [9], hepatitis and achalasia [10], primary biliary cirrhosis [11], angina pectoris [12], or even chronic orofacial pain and mucosal inflammation [13]. Cotricosteroids are widely used in IHS treatment. Prompt responses usually were observed in patients who received prednisone at a dose of 1mg/kg daily.Hydroxyurea, imatinib, nilotinib, alemtuzumab, and mepolizumab have been also used in corticosteroid-unresponsive or relapsed patients depending on some markers e.g. usage of tyrosine kinase inhibitors in FIP1L1-PDGFRA-positive patients [2,6,14].

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DOI: 10.19080/JETR.2017.02.555586

How to cite this article: Mina T K.Idiopathic Hypereosinophilic Syndrome: Etiology, Diagnosis, Manifestations and Management. J Endocrinol Thyroid Res. 2017; 2(3): 555586.DOI: 10.19080/JETR.2017.02.555586