A Case of Late-Onset Hereditary Angioedema in a Patient with Non-Hodgkin's Lymphoma, Common Variable Immunodeficiency, and Antiphospholipid Antibody Syndrome

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Abstract

Introduction

Hereditary angioedema (HAE) with C1q deficiency is a rare hereditary condition characterized by repeated attacks of angioedema. This condition most frequently affects the skin or mucosal tissues of the upper respiratory and gastrointestinal tracts. HAE can be associated with lymphoproliferative disorders such as splenic marginal zone non-Hodgkin's lymphoma and should be considered in the differential for patients with recurrent attacks of angioedema or abdominal pain that is unresponsive to treatment. HAE is underdiagnosed and continued research and education on diagnosis and management of the disease are warranted.

Case Presentation

A 71-year-old woman presented to the ED with a complaint of abdominal pain. Her symptoms included cramping, constipation, diarrhea, nausea, and severe vomiting. The patient's previous medical history consists of splenic marginal zone non-Hodgkin's lymphoma, treated successfully with chemotherapy; common variable immunodeficiency with ongoing IVIG treatment; positive anticardiolipin antibody testing with daily 81 mg of aspirin and no current thrombotic events. Subsequent diagnostic testing results indicated a diagnosis of late-onset C1q hereditary deficiency angioedema was made.

Discussion

HAE due to acquired C1-INH deficiency is difficult to identify and is most commonly diagnosed after many years of recurring attacks. Currently, HAE treatment relies on short- and long-term prevention and treatment for acute attacks. The goal of HAE therapy is to reduce the frequency and severity of angioedema and improve patient quality of life. Due to the variable clinical presentation and severity of the disease, if untreated, it is essential to recognize and initiate appropriate treatment for HAE.