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Case Report

Recurrent Vomiting due to Intestinal Angioedema

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Abstract

Estrogen-dependant (type III) is the rarest form of hereditary angioedema, with a myriad of clinical presentations including acute gastrointestinal symptoms.

Patients may have a long odyssey before diagnosis and do need a dedicated prophylaxis.

Keywords: Hereditary Angioedema; Gastrointestinal Symptoms; Androgens

Introduction

Recurrent episodes of vomiting with abdominal pain may unmask a hidden metabolic cause; particular attention is so mandatory to find clues in such conditions.

Detailed history and accurate examination, along with adequate investigations are mandatory to resolve such clinical enigma.

Case Presentation

A teenage girl of 14 years is admitted for an acute episode of abdominal pain and recurrent vomiting/nausea with acute swelling of face and feet.

She had been hospitalized several times -during the last 3 years- for similar conditions, misdiagnosed as acute glomerulonephritis or as severe viral infection.

The patient is at her first day of menses, and clinical examination is free from rash or pruritus.

Clinical suspicion of hereditary angioedema led to check complement serology (C4 and C1-inhibitor) but results were negative.

Pooling history with clinical and serological elements, the diagnosis of type III Herediatry Angioedema was considered.

Supportive care was beneficial and the patient relieves within three days.

Long-term prophylaxis with androgens (Danazol) due to precedent attacks severity was efficient after a 6 months follow-up.

Discussion

Hereditary angioedema (HAE) are inherited disorders characterized by subcutaneous and submucosal edema, mainly in respiratory and gastrointestinal tracts.

HAE type I (synthesis deficiency) and type II (functional deficiency) result from a deficiency in the plasma level of functional C1 inhibitor. The SERPING1 mutations are well-recognized as responsible for thes two autosomal dominant disorders.

Type III is a rare form of HAE that occurs in women and is precipitated or worsened by high estrogen levels. It is associated with mutations in the gene for Factor XII that result in its increased activity [1]. Its most distinguishable feature is the clinical phenotype as estrogen-dependent; with normal C4 and normal C1 INH level and function [2,3].

Clinically, all HAE types may manifest with swelling of the face, extremities, and upper airways. Gastrointestinal symptoms are

also common and may include abdominal pain, vomiting, and diarrhea. Pancreatitis is also reported, as well as surgical-like abdomen [2,4,5].

Treatment of type III HAE can be categorized as treatment of attacks (on-demand therapy) and prophylactic therapy (short- and long -term). Both options were used in managing our case with a dramatic response to the attenuated androgen [6].

Safety, availability and low-cost of attenuated androgens are highly appreciated, especially in low-middle income countries [7].

Conclusion

Before being diagnosed, HAE patients suffer during many years from recurrent respiratory and/or digestive symptoms with subcutaneous edema, and gastrointestinal involvement is frequently at the forefront of HAE manifestations.

Response to androgens, alongside with other non-specific therapies, are peculiar of the type III "feminine" HAE.

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