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Editorial

Abdominal Attacks May Reveal Hereditary Angioedema!

Naouel Lemdaoui[1,2], Nada Boutrid[3,4], Hakim Rahmoune[3,4]*, Mounira Amrane[3], Abderraouf Bataiche[1,2] and Hala Boutrid[5]

[1]General Surgery "B" Department, Ben Badis University Hospital of Constantine, Algeria

[2]Faculty of Medicine, Constantine-3 University, Algeria

[3]LMCVGN Research Laboratory, Setif-1 University, Algeria

[4]Department of Pediatrics, University Hospital of Setif, Setif-1 University, Algeria

[5]Department of Obstetrics-Gynecology, University Hospital of Babeloued, Algiers-1 University, Algeria

***Corresponding Author:** Hakim Rahmoune, LMCVGN Research Laboratory, and Department of Pediatrics, University Hospital of Setif, Setif-1 University, Algeria.

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We received a 14-years old girl suffering from recurrent acute attacks of abdominal pain and nausea with facial swelling and labial angioedema, reoccurring irregularly since 3 years without a clear trigger.

A large panel of radiological and laboratory investigations could not unveil the cause of these episodes, until she was readmitted in the ward during a similar crisis with afebrile abdominal pain, face swelling and a transient crural macular rash.

The patient is on her first day of menses and is free from urticaria or pruritus.

This clinical stereotyped sequence is highly suggestive of Hereditary Angioedema (HAE), the most common type of complement related, non-allergic angioedema; with peritoneal involvement and intestinal swelling revealed by pain and vomiting.

Repeated screening tests and an exhaustive review of the patient history and evolution, pooling clinical and serological elements, allowed the final diagnosis of the rare type III HAE, non-deficient and oestrogens-related.

The HAE is a genetic disease caused by a deficient or a dysfunctional Complement C1-inhibitor protein, with subsequent vasodilation due to bradykinins [1].

These HAEs, including the rarest form of type III oestrogenic HAE, are often diagnosed with long delay because of the non-specific and heterogeneous clinical presentation frequently encompassing subcutaneous oedema and digestive signs [2].

In fact, before diagnosis, patients frequently present nausea, vomiting and abdominal pain (due to intestinal wall swelling), as well as subcutaneous oedema during many years.

Sharp look to patient history and accurate physical examination are obviously mandatory, along with prompt and adequate biological exploration to resolve such clinical enigma.

Different drugs are now available for this scarce complement disorder. Specifically, androgens are cheap and easy-to-use drugs for the oestrogenic type III HAE and might be dramatically efficient to diminish the severity and frequency of attacks [3,4].

In sum, these recurrent abdominal signs, ranging from nausea/vomiting to severe abdominal pain, deserve a particular attention in order to unmask some uncommon conditions like bradykinin-associated angioedema.

These angioedemas present with a myriad of clinical signs, including digestive ones; and patients may experience a long (and sorrowful!) odyssey before diagnosis.

Recent guidelines provide strong diagnostic and therapeutic algorithms for these peculiar angioedema [5].

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