Reviewer's report

Title: Leukocytosis and high hematocrit levels yield misinterpretation in abdominal emergency of hereditary angioedema (HAE)

Version: 6 Date: 20 March 2013

Reviewer: andrea zanichelli

Reviewer's report:

Major Compulsory Revisions
I do agree with authors that it is important to increase awareness of HAE among physicians and that often this rare condition is misdiagnosed.

Nevertheless the aim of the study is not well defined. This study compare laboratory findings in HAE patients during remission and during an acute attacks, does not compare lab tests in patients with HAE during an abdominal attacks and patients with abdominal pain due to other diseases to explore the cause of misdiagnosis of HAE.

It is already known that WBC increase during an acute abdominal attack. This study confirm this data. The increase in WBC does not help in making a differential diagnosis. Other markers might be helpful (see manuscript Cugno M et al, Plasma biomarkers of acute attacks in patients with angioedema due to C1-inhibitor deficiency, Allergy 2009: 64: 254–257

Minor essential revisions
Keywords:
- C1 inhibitor concentrate

Background:
- “absence or defect of the C1INH". HAE patients have a deficit of C1 INH. HAE is an autosomal disease, levels of C1 INH are reduced, <50% of normal individuals, but C1 INH is present.

- Plasma-derived C1 INH is not the only treatment for acute attacks, there are several drugs available (Icatibant, Ecallantide, Ricombinat C1 INH). If plasma derived C1 INH is the only treatment available in Japan the authors have to specify that this is the situation in Japan

Patients and method:
- authors should report if patients were on prophylactic treatment with androgens because these drug might increase the hematocrit levels

Results:
- authors should report the value of C3, C4 and functional C1 INH and the normal range

Conclusions:
- conclusions are unsatisfactory

References:
- authors should include this reference “Cugno M e al, Plasma biomarkers of acute attacks in patients with angioedema due to C1-inhibitor deficiency, Allergy 2009: 64: 254–257” in place of reference number 6

**Level of interest:** An article of limited interest

**Quality of written English:** Needs some language corrections before being published

**Statistical review:** Yes, but I do not feel adequately qualified to assess the statistics.

**Declaration of competing interests:**
I declare that I have no competing interests