Reviewer’s report

**Title:** Thrombotic thrombocytopenic purpura complicating pregnancy. Perimortem cesarean in a case of unfavorable course

**Version:** 2  **Date:** 24 March 2013

**Reviewer:** Mario Ojeda-Uribe

*Which of the following best describes what type of case report this is?:* An unexpected event in the course of observing or treating a patient

*Has the case been reported coherently?:* Yes

*Is the case report authentic?:* Yes

*Is the case report ethical?:* Yes

*Is there any missing information that you think must be added before publication?:* Yes

*Is this case worth reporting?:* Yes

*Is the case report persuasive?:* No

*Does the case report have explanatory value?:* No

*Does the case report have diagnostic value?:* No

*Will the case report make a difference to clinical practice?:* No

*Is the anonymity of the patient protected?:* Yes

**Comments to authors:**

Comments

1. **Abstract**

   Introduction

   Authors report a case of TTP, but neither ADAMTS13 nor antibodies to ADAMTS13 were assessed.

   (I mean it is important to try to differentiate between idiopathic and Upshaw-Schulman Syndrome as a high frequency of this entity is found out in pregnant women, see Veyraider et al Blood 2012).
I disagree with authors the situation was critical at admission (Hemoglobin 5 g/dL) and very low platelet count 5X10^9/L) plus high LDH values.

Conclusions’ paragraph: the authors mentioned a triad of anemia, thrombocytopenia and neurological changes. The need to clarify the type of anemia: non autoimmune.

(Many cases of chronic ITP can be associated to anemia and this does not mean a TMA).

You can not wait for results of ADAMTS13 measurements in order to decide what to do. A suspicion of TTP related to pregnancy has some well defined therapeutic rules that is important to respect.

2. Introduction

Which are the references that authors can include to support the association of an « hypercoagulability state » and TTP in pregnancy?

What do they mean when they talk about the increased frequency of TTP in pregnant women with low ADAMTS13 levels? Do they talk about patients with a history of idiopathic TTP? and/or patients with known or unknown Upshaw-Schulman syndrome?

Coombs’s test (DAT)

Reference 7 which gives such a high level of neurological complications has to be updated.

Authors talk about an improvement in the last 30 years because of the use of plasma therapy. They are right but they cannot say that is due in priority to infusions of fresh frozen plasma (in addition it is not only fresh plasma!!) (cf…Other treatments such as plasma exchange help to remove…). This is confusing for authors. Plasma exchange adds ADAMTS13 that is present in the plasma infused and removes also as they say potential antibodies and very large VWF multimers. This paragraph needs to be largely modified.

Authors have to mention that ADAMTS13 and potential auto-antibodies to ADAMTS13 were not assessed in this case. Platelets values must be written according to standards rules.

I don’t know what was the death cause? If it was due to pulmonary oedema, was it triggered by a cardiac failure related to cardiac microangiopathic lesions or to occlusion in larger coronary vessels? Do they have data about a necropsia assessment of the cardiac tissue? I think this could be an interesting aspect to develop. The figure of the brain is interesting but histological findings of this and other tissues could add some value to this work.

Authors describe multiple micro-thrombi so I guess this histological features were assessed and might be available.

3. Discussion
The authors mentioned studies highlighting the need for regular and close follow-up of patients with recurrent TTP. This is an important issue. Which are these studies?

What do they mean when they say that « as patients have prior episodes the risk of recurrence is known »?

Because most of the authors are affiliated to the Obstetric Department of their Institution it could be worth value if they try to give more information about recommended therapeutic options at the different trimesters of pregnancy and what are the expected outcomes in case of the interruption of pregnancy based on published data.

4. Conclusions

Same as previously you can not wait for ADAMTS13 values to decide what to do.

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