Reviewer's report

Title: Encephalopathic Susac's Syndrome associated with livedo racemosa in a young woman before the completion of family planning

Version: 1 Date: 2 August 2013

Reviewer: Friedemann Paul

Reviewer's report:

This is an interesting case report that definitely contributes to the literature on this rare disease. Every new case is relevant so the paper deserves publication after some revisions, see below.

Major compulsory revisions:

There are some flaws regarding the case description, the literature cited and the discussion that need to be revised, in detail:

- abstract: the evidence on antibody-mediated damage of endothelial cells is still weak, one recent paper on that cited by the authors (nr 8 Magro) has severe methodological flaws. I suggest to be more cautious in this respect.

- introduction: a link is missing that leads to the case description, so the authors should add one or two sentences to the end of the introduction that makes clear why this case is presented subsequently.

- introduction: current and relevant literature is neglected and needs to be added and reference, for example on antibodies PMID:19643446, or PMID:23628737 so the number of 200 reported cases given by the authors is no longer correct, or PMID:22711711 on MRI

- case description: numerous open questions

-- CSF: what about OCB? pos or neg? this has to be mentioned

-- first MRI: it would be helpful to have this in the figure including the "strikingly atrophic" CC which is a relevant diagnostic and differential diagnostic feature of SS but not discussed at all by the authors (see discussion)

-- were FAG and/or OCT performed? If not, please mention this, this aspect would also contribute to the discussion, see for example PMID:22701702, PMID:20880549

-- MRI: what about meningeal contrast enhancement, see the Susac 2003 paper, this is quite typical of SS but rarely seen in MS

-- I do not understand the process of making the diagnosis of SS. The authors mention that they considered "a tentative diagnosis of MS" but do not report when and why they considered SS. Was it the clinical presentation or some typical MRI features like callosal atrophy? This needs to be described in more detail and should also be picked up in the discussion because it is a relevant
clinical challenge

-conclusion/discussion:
- approximately 200 patients: please correct and provide actual ref, see above
- the authors do mention correctly that unnecessary immunosuppression must be avoided. In this context it would be helpful to have some more thoughts on diagnostic work-up and differential diagnosis, in particular versus MS the probably most frequent misdiagnosis. For example, the interesting aspect of callosal atrophy the authors describe in their patient could be picked up, if this is impressively visible the authors should include this to the figure.

- references: far from up to date, needs to be revised, see above

Minor: please check affiliations, Dr. Kleffner has an email address in Münster but the affiliation on the paper is with the department of Gynecology in Tübingen

Level of interest: An article of importance in its field

Quality of written English: Acceptable

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:

no competing interests