Reviewer’s report

Title: A case report of adult-onset COQ8B nephropathy presenting focal segmental glomerulosclerosis with granular swollen podocytes

Version: 0 Date: 21 May 2020

Reviewer's report:

Dear authors:
The authors of this clinical case " A case report of adult-onset COQ8B nephropathy presenting focal segmental glomerulosclerosis with granular swollen podocytes" find some findings of abnormal podocytes in light microscopy that could be useful in the differential diagnosis of patients with FSGS. Although the findings found by the authors of this case report are novel and of great scientific interest, there are nevertheless some questions that would need to be answered to improve the scientific evidence on the paper.

Major revisions
1) First of all, it is striking that this patient has exclusively kidney involvement. As the authors will know, in a large percentage of cases there are extrarenal manifestations, mainly neurological. How could the exclusive renal involvement be explained in this clinical case?
2) The main question refers to the findings of the renal histology. Are these types of population abnormalities specific to this entity? Is there other scientific evidence that can corroborate these findings? These podocytic findings could not correspond to those found in the FSGS podocytes? It is essential to clarify these issues to give greater scientific relevance to this clinical case.

Please confirm that you have included your review in the ‘Comments to Author’ box?

As a minimum standard, please include a few sentences that outline what you think are the authors’ hypothesis/objectives, their main results, and the conclusions drawn. Your report should constructively instruct authors on how they can strengthen their paper to the point where it may be acceptable for publication, or provide detailed reasons as to why the manuscript does not fulfill our criteria for consideration. Please supply appropriate evidence using examples from the manuscript to substantiate your comments. Please break your comments into two bulleted or numbered sections: major and minor comments.

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NA

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- Are the statistical test(s) used in this study appropriate and well described?
- Is the exact sample size (n) reported for each experimental group/condition (as a number, not a range)?
- Are the description of any error bars and probability values appropriate?
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