Reviewer's report

Title: Life-threatening hypersplenism due to idiopathic portal hypertension in early childhood: Case report

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Reviewer: Henry Rice

Reviewer's report:

This case report by Dabritz et al is of an infant with idiopathic portal hypertension. The report is the clinical summary, novel pathologic findings, and management of this disease in a young infant. In itself, this is a rare condition in childhood and would by of interest to the readership of BMC Gastroenterology. However, in its present form, the report is limited by both grammatical and structural errors which would need substantial revisions before consideration for publication. My specific recommendations are as follows:

General

1) I find the language of the report difficult to read. It would be helpful to have a native English speaking editor review it and make appropriate changes. There are far too many specific examples to cite, but I will try to highlight a few.

2) I am unclear exactly what is the point of this report. It is simply the presentation of IPH in a young infant (which is rare and of interest in itself) or description of novel pathologic and laboratory findings suggestive of NO mediated disease (which is poor data but interesting). A hypothesis, even for a case report, would be helpful to direct the authors.

Abstract

1) Too wordy, long, and unclear. Again the lack of hypothesis shows in the structure of the abstract.

Background

1) I think the description of IPH is not directed appropriately, with many causes other than parenchymal atrophy of the liver. In children, the emphasis should be on portal vein thrombosis and non-thrombotic causes. Discussion along the lines such as in Sarin and Kapoor, J of Gastro and Hepatology, 17:526-534, 2002, which more clearly summarize the known etiology and classification of IPH would be helpful.

2) To follow-up, I would disagree that most references point to "increased splenic and portal vein blood flow." In contrast, most suggest either presinusoidal or extrahepatic vasculopathy which lead to portal hypertension.

Case Presentation
1) Too long and again, not directed. For example, the first paragraph on patients is almost itself not required, as this could be included in the case summary.

2) What were the findings of the open liver biopsy at 13 months? Most of the clinical background is not necessary, but this is a key data point that needs to be described.

3) Splenomegaly, not hepatosplenomegaly, is the hallmark clinical finding of IPH, along with variceal bleeding. Therefore, the diagnosis of IPH of extrahepatic origin is not clearly made.

4) I am not sure what sonogram findings of "highly normal blood flow" mean, as this is not common language.

5) Shorter, but clearer, description of the clinical problem of hypersplenism needs to be done. What was the "hemorrhagic diathesis."

6) Pathologic findings should be separate section. What is meant by "splenoadenoma?"

7) The rationale for choice of immunologic staining should be done. Why eNOS, VCAM-1, etc.

Splenectomy in IPH

1) Again too wordy. Is point of section simply to tell of previous case reports of splenectomy (which is standard care for IPH), reports of children, or association of autoimmune disease (which is well described)?

Role of Spleen in IPH pathogenesis

1) Again, too wordy and unclear what the point of this section is. The previous reports of overexpression of VCAM-1 and ET-1 are quite interesting, but I am not sure what the authors are trying to argue in terms of underlying pathogenesis of IPH. A figure describing their theory may be helpful here.

Conclusions

1) Needless to say, too wordy and poorly organized. Again a hypothesis would help organize this discussion.

2) In terms of treatment, there should be a notation of options other than splenectomy, including splenic embolization, etc.

Figures and Tables

1) Figure 1 clear, but as only one MRI, cannot state "progressive" enlargement. Not sure what "highly normal" portal vein flow means.

2) Figure 2 relatively clear and helpful.

3) Figure 3 does not contribute to the report and should be deleted.
4) Table 1 needs legend. Most of this information is in the text, and can probably be deleted in either the text or remove the table.

5) Table 2 needs legend. Relatively good in present form.

**Level of interest:** An article whose findings are important to those with closely related research interests

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

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

**Declaration of competing interests:**

I have no competing interests