Author's response to reviews

Title: Primary sclerosing cholangitis associated with CREST in an elderly woman: a case report.

Authors:

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Version: 6 Date: 13 September 2015

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Authors:

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Version: 2 Date: 13 September 2015

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The Biomed Central Editorial Team

Object: MS: 1401700523170479 - Primary sclerosing cholangitis associated with CREST in an elderly woman: a case report. Dr Alice Powell et al.

Thank you for your consideration of our manuscript for publication in your journal.

We have reviewed the above manuscript according to your reviewer’s comments:

Reviewer #1 (Dr Galati)

1. Does the Introduction explain the relevance of the case to the medical literature?: yes but the authors must refer to a similar case report between Primary Biliary Cirrhosis and CREST (Ryumachi. 1997 Feb;37(1):42-7. [Primary biliary cirrhosis (PBC)-CREST overlap syndrome complicated with Sjögren’s syndrome].

   - We have not specifically referenced this article but we have emphasised the association between PBC and CREST first reported in the 1970s by Murray-Lyon et al

2. I think that it is crucial to report the liver tests.

   - We understand this and the values have now been included

3. Although it is a different hepatopathy, the authors must refer to a possible association between CBP and CREST. The absence of a liver biopsy, although it is declared in the text, must be better supported. I think that it is important to report also the liver test at diagnosis.

   - We have now placed more emphasis on the relationship between PBC and CREST and provided justification for why a liver biopsy was not pursued. We have included the initial liver function test results.

Reviewer #2 (Dr Villani)

1. There is no mention of serum IgG4 level as the IgG4-SC should be considered in the differential diagnosis of primary sclerosing cholangitis.

   - This has been amended and the serum IgG4 level is now included

2. I think that it may be useful a more detailed discussion on differential diagnosis of the various type of primary and secondary sclerosing cholangitis also for therapeutic importance of steroid treatment in IgG4-SC

   - We have now included a new paragraph with a discussion of differential diagnoses of primary and secondary sclerosing cholangitis including IgG4 related disease. We have made note of the glucocorticoid responsiveness of IgG4 associated cholangitis
Reviewer #3 (Dr Guarascio)

1. *I think the work need only the approval of the patient*
   - We have obtained consent from the patient for publication of this case report

2. *Maybe I would not write the Indonesian background and the nationality; bringing instead the race (caucasian etc.)*
   - We understand this concern and have now amended the patient details to reflect her race rather than her nationality

3. *The diagnosis was properly made with the only MRCP without resorting to liver biopsy. However, this case is unusual since presents positive ANA with the alteration of liver enzymes. The ANA positivity and the alteration of liver enzymes, characteristic of AIH, suggest a possible overlap syndrome to be diagnosed histologically in order to prescribe the specific therapy. I think that it would be a considerable enrichment to know the histological features in PSC associated to CREST, never described so far in the medical literature.*
   - We recognise that a biopsy in this particular case may have been illuminating, although it is not usually necessary in the diagnosis of PSC and can be non-specific
   - The reason why a biopsy was not pursued was mainly, however, due to logistics and concern for the patient as she is elderly with multiple medical comorbidities and may not tolerate this procedure.
   - We determined that, on balance, she was unlikely to derive any individual additional benefit from having the biopsy with knowledge of the attendant risks involved/

4. *I would suggest to test the level of IgG4 which is a specific marker associated with a PSC variant.*
   - This information has now been included