

Mar 3, 2014

Dear Editor,

We are really grateful for the three referees' comments and revised the manuscript accordingly. We hope that the manuscript has improved significantly.

Title: Intraductal papillary neoplasm of the bile duct developing in a patient with primary sclerosing cholangitis.

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The following are our responses to the specific comments.

Responses to Reviewer #1 69920

Comment 1.

It is the first case report of intraductal papillary neoplasm of the bile duct (IPNB) from PSC. As a case report, it is accepted.

The language must be revised before publishing.

Our Response 1.

Thank you for your acceptance.

We already depend on a proofreading company and proofread it.

Responses to Reviewer #2 2444990

Comment 1

Very detailed description of the clinical case and may be interesting the association with Behcet's syndrome and IBDu.

Our Response 1

The patient was intestinal Behçet's disease. Both PSC and Behçet's disease show a bowel lesion. To our knowledge, there was no case report in which Behçet's disease and PSC

complicated.

Comment 2

Provide abstract In case report it would be useful to describe what kind of follow-up was performed and, if available, add some images of the various imaging modalities used.

Our Response 2

Reviewer#2 is right. We performed postoperative observation using CT, but no recurrence has been detected 3 years after surgery. We were not able to mention it by number of words in abstract.

Comment 3

In the discussion it would be very useful include a more detailed description of clinical-diagnostic-therapeutic process of this disease

Our Response 3

Because it was case report and the existence of several reports, we omitted description of clinical-diagnostic-therapeutic process. However, we added some sentences.

- It is difficult to make an accurate preoperative diagnosis because of IPNB's low incidence and the lack of specificity in its clinical manifestation.
- US, CT, MRI, ERC, cholangioscopy and IDUS are all useful for diagnosing tumor extension, involvement of bile duct and the presence of mucin.

Responses to Reviewer #3 1076129

Comment 1

Although definite conclusions cannot be drawn without longitudinal follow-up, IPNB is suggested to be a part of the carcinogenesis in patients with primary sclerotizing cholangitis.

Our Response 1

Because I had not observed the PSC patient for a long term, we wrote it in consultation with other report.

It has been suggested that PSC may have some involvement in stepwise progression to CCA via the metaplasia-dysplasia-carcinoma sequence. (Reference7)

Comment 2

The description of symptoms, imaging and laboratory data does not represent unique

features for IPNB, but depict obstructive jaundice and cholangitis irrespective of cause.

Our Response 2

Reviewer#3 is right. The laboratory data which we showed is data of preoperation.

Table1 deleted it and mentioned data in sentence.

We changed the data at the time of the hospitalization that developed obstructive jaundice.

Finally thank you very much again for very helpful comments and discussion. We believe that the revised manuscript would be valuable information for readers of *World Journal of Gastroenterology*.

Sincerely yours,

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