

April 4, 2014

Dear Editor,

Please find enclosed the edited manuscript in Word format (file name: 9458-edited).

Title: Malignant gangliocytic paraganglioma of duodenum with distant metastasis and lethal course

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Name of Journal: World Journal of Gastroenterology

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The manuscript has been improved according to the suggestions of reviewers:

1 Format has been updated according to the suggestion of editor.

2 Revision has been made according to the suggestions of the reviewer (reviewer #1)

(1) I think there was a discrepancy between malignant clinical course and pathological findings. Therefore the author should speculate the reason in Discussion.

Answer: Many thanks. In revision, we have added comments to postulate the discrepancy between malignant clinical course and pathological findings in Discussion section.

(2) Please describe the detail of chemotherapy regimen and radiotherapy. Because this case was a rare, I think the detail of therapy is important for readers.

Answer: Many thanks. The details of chemotherapy regimen and radiotherapy have been described in the revised manuscript. The discussion of efficacy of adjuvant treatments for this tumor has also been added in revision. At present, there is no guideline for chemotherapy of duodenal gangliocytic paraganglioma due to the rarity of distant metastases. The systemic chemotherapy strategy in our case is conventional chemotherapeutic regimens for malignant paraganglioma. But the tumor seems to lack the response to these regimens. Although radiotherapy has been used to treat the regional lymph node metastasis, but the efficacy of treatment was difficult to ascertain.

(3) Please show the picture of MIB-1 (Ki-67) in immunohistochemical findings. Because this case showed malignant and lethal clinical course, but Ki-67 labeling index estimated less than 1% in both primary and metastatic foci. I think this point is interesting. In addition, please add the information about lymphovascular invasion identified by D2-40 and vascular invasion identified by CD31 or other markers.

Answer: Many thanks for your suggestion. Immunohistochemical staining for Ki-67 have been performed on the sections of primary and metastatic lesion, the Ki-67 index low. The figures of Ki-67 have been added into the revision. But Ki-67 index is lower in GP in our case and other previously reported cases, and seems not appropriate marker for malignancy of GP. [Okubo Y, et al. Duodenal gangliocytic paraganglioma showing lymph node metastasis: a rare case report. *Diagn Pathol* 2010; 5:27; Okubo Y, et al. Literature survey on epidemiology and pathology of gangliocytic paraganglioma. *BMC Cancer* 2011; 11:187]

In addition, the information of CD31 and D2-40 immunohistochemical staining has been added into the result section in revision.

3 Revision has been made according to the suggestions of the reviewer (reviewer #2)

(1) The presented case is an unusual case of Malignant gangliocytic paraganglioma of duodenum. This is a well documented case report and your suggestion of changing the tumor behavior code is acceptable.

Answer: [Many thanks for your comments.](#)

4 References and typesetting have been corrected according to the suggestions of editor. Thanks

Thank you again for publishing our manuscript in the World Journal of Gastroenterology.

Sincerely yours,

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