Author's response to reviews

Title: Gorlin syndrome associated with small bowel carcinoma and mesenchymal proliferation of the gastrointestinal tract: Case report and review of literature

Authors:

Peter M. Prodinger (p_prodinger@hotmail.com)
Mario Sarbia (sarbia@pathologie-muenchen.de)
Jörg Maßmann (massmann@pathologie-muenchen.de)
Christian Straka (christian.straka@schoen-kliniken.de)
Günther Meyer (Dr.Meyer@chkmb.de)
Ortrud K. Steinlein (Ortrud.Steinlein@med.uni-muenchen.de)

Version: 5 Date: 5 May 2010

Author's response to reviews: see over
Re-submission of manuscript MS: 1040713522326744

Dear Dr. Shipley,

Please find attached the second revision of our manuscript that we would like to re-submit to *BMC Cancer*.

We are again very grateful for the reviewer’s helpful comments and, according to them, made the following changes:

**Reviewer 1: Mariana Cajaiba**

We agree with reviewer 1 that the term *neuroleiomyomatosis* can not be fully characterized by a single case report. However, the sole use of a descriptive term such as “mesenchymal proliferation” would hide one of the most unique observations in this case and thus prevent the scientific discussion of this point. We (also the reviewing pathologists in Basel/Switzerland) are convinced that this is a probably not yet described proliferation and some unusual kind of ganglioneuromatosis. We therefore would like to keep the term neuroleiomyomatosis as provisional diagnostic entity, but also added the description suggested by reviewer 1: “mesenchymal proliferation with neural and smooth muscle components”.

1. Both methods showed the presence of neurons, indicating that a neuronal component was part of the mesenchymal proliferation. However, the application of the term ganglioneuromatosis would not be correct due to the proliferation of the smooth muscle component, which was not separable from the neuronal component (see Results page 4/paragraph 2).

2. We cannot entirely exclude that the smooth muscle component was a reactive process along with a neural neoplasm, however, we can either not exclude that the neural proliferation was a reactive process to a smooth muscle proliferation. The arguments against both interpretations are that there were numerous such proliferations and all of these consisted of an intimate proliferation of both components that could not be separated by light microscopy.

3. Thanks!
Additional points:
1. The following information was added to the results paragraph: This mutation has not been described before.

2. Page 3: The corrected sentence here now reads: Macroscopic examination revealed numerous....

3. Node was substituted by nodules.

4. We deleted the misleading question and substituted it with the following sentence: “Are both the mesenchymal proliferation and the small bowel carcinoma pathogenetically linked to the PTCH stop codon mutation?” We also deleted the sentence If the adenocarcinoma in our patient is indeed pathogenetically connected with the mesenchymal proliferation, the malignant potential of the later is probably too low to raise a red flag in a syndrome this rare from the conclusions paragraph.

Reviewer 2: Vesna Musani

The spelling and grammatical errors have been corrected.

We hope that we were able to answer all questions raised by the referees and would again like to thank them for their time and most valuable comments.

Best regards,

Prof. Dr. Ortrud Steinlein