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Dear editor:

We would like to thank the reviewers for their valuable suggestions. Our manuscript has been revised based on every comment from the reviewers (see the next pages for details). We have also asked MedSci colleagues of native English speakers to proof read the manuscript and they have already gave us the certification of English editing. We sincerely hope that the paper is now suitable for publication on World Journal of Clinical Cases. If you have any questions, do not hesitate to contact me.

Regards,

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Reviewer 1:**Question:**

This paper only discussed the clinical feature of 5 patients of PJS who were diagnosed by clinical criteria only. The importance of PJS in clinical practice lies on its increased risk of associated malignancy. The authors did not included on these data

Answer:

1. The data of malignant tumor risk in PJS pointed out by the reviewer have been added in the paper.

(1) **In “Material and Methods” part, we add this paragraph:** The present study retrospectively analyzed 46 cases PJS patients, between June 2014 and January 2017, diagnosed by biopsy, endoscopic resection and postoperative pathology. Among these patients, there are five cases from different pedigrees were diagnosed as malignant change secondary to PJS.

(2) **In “Discussion” part, we add this data:** The results show that^[6] the median age for first diagnosed cancer is about 45 years old. With the coming of age, the cumulative cancer risk rate of PJS patients significantly increase from 20% to 76%, especially higher in young female patients^[6]. Another research^[7] analyzed 336 PJS patients show that 52 patients (15.5%) diagnosed cancer, the cancer relative risk and the cumulative risk were 63.8 and 55%, respectively. The median age for first diagnosing malignant tumor was about 41 years old. In our study, there are 46 patients diagnosed PJS. Among these patients, five people occurred malignant tumor secondary to PJS, the cancer risk rate was 10.9% and the median age at first diagnosis was 43.6 years (range from 29 years to 67 years). Our result is consistent with above studie^{s[6-7]}. Interestingly, gene mutation in PJS patients also increase cancer risk rate.

(3) **In “Discussion” part, we add another research data for gene mutation:** An Italian multicenter research reported that^[10] there are 29 patients occurred gene mutation among 119 patients. The overall cumulative cancer risk rate were 20%,43%,71%,89% at age of 40,50,60 and 65years old respectively. Therefore, we need to perform the gene detection and it can be a promising way^[11]. Meanwhile, for young female patients with PJS, long-term combined chronic abdominal pain, diarrhea and gastrointestinal bleeding with no improvement after conservative treatment should take into consideration of malignant tumors and take positive measures.

Reviewer 2:

The second reviewer think that our paper is well-documented and adequately written in detail.