Author’s response to reviews

Title: Superior Mesenteric Artery Syndrome coexists with Nutcracker Syndrome in a female: a case report

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Author’s response to reviews:

Point by point response

Editor Comments:

In addition to the referee comments, please address the following editorial points:

1. Please further copy edit your manuscript. See below for more information.

2. Some overlap still exists in the Case Presentation section with the study by Michael PG et al (your reference 5). Please see the attached and make further minor edits to reduce overlap in the highlighted sentences.

Thank you very much. Please find the revised manuscript in Abstract section on Line 44-46, Page 1-2, and in Case presentation section on Line 90-91, 95-97, 102-103, 118-119, 120-122, 128-129, Page 3.
Reviewer reports:

Weiwei Ding (Reviewer 1): This case reports a female patient with SMA syndrome with Nutcracker phenomenon. The authors highlight the importance of the combination therapy of long-term nutritional supporting and promotility agents.

I have several concerns:

1. As a national GI referral center, SMA syndrome with Nutcracker phenomenon is not a rare case in my department. Even in the literatures, there has been lots of reports about the treatments and diagnosis of SMA syndrome with Nutcracker phenomenon.

   Thank you, Professor. The prevalence of SMA syndrome has been reported at around 0.0024–0.3% (J Gastrointest Surg 2018, J Gastrointest Surg 2009, Dig Surg 2007). The low prevalence is generally defined as “fewer than 1 in 2,000 people”. Overall, SMA syndrome is a rare disease. Please find the revised manuscript on Line 64-65, Page 2. However, it is no doubt that, as a national GI referral center, SMA syndrome with Nutcracker syndrome may be not a rare case in your department. We supposed that those data of SMA syndrome are valuable and probably could be probably summarized. If possible, we sincerely hope to have an opportunity to cooperate with you.

2. The treatment in this case added nothing new to our knowledge about this disease.

   Thank you, Professor. Medical treatment may consist of nasogastric intubation for gastroduodenal decompression, reversal or removal of the precipitating factor and nutritional supplementation with hyperalimentation by jejunal feeding tube. Here, we highlight the importance of the combination therapy of long-term nutritional supporting and Pro-motility agents. Besides, we recommend the rehabilitating practice after discharge, such as swimming, which may be beneficial to enhance the abdominal wall and reduce the frequency of recurrence. However, it is just a case report. Clinical trials and much more cases still be needed to assess the effectivity. Please find the revised manuscript on Line 239-243, Page 6.

3. As a CASE REPORT, this manuscript lacks the results of a long-term follow up. How about the updated status after six-month of home nutritional treatment?
Thank you, Professor. The long-term follow up is indeed an essential part of case report. We have added the - Follow-up - section following the - Case presentation - section. Please find the revised manuscript on Line 134-149, Page 4. We also collected follow-up BMI data as Figure 3 and figure legends (Line 357-360, Page 9).

Jussi M Karkkainen, MD, PhD (Reviewer 2): The authors report a case of bowel obstruction caused by SMA syndrome. The acute symptoms resolved with decompression by nasogastric tube. The CT images of this case report are illustrative and conclusive.

It is unclear, whether the use of spasmolytic drugs was a predisposing factor for the acute symptoms. Was the patient still on that medication during hospitalisation? How often and for how long did she need the medication? Please be more precise. Use small first letters when captioning generic drug names. Avoid abbreviations such as po tid, not everyone knows what this means.

Because of the kidney stone history, this patient received oral drotaverine hydrochloride tablets 40mg three times per day for 2 weeks. It is no doubt that weight loss should be the predisposing factor for the acute symptoms. And meanwhile, spasmolytic drugs could relax gastrointestinal smooth. Thus, we supposed that spasmolytic drugs led to the disorder of the gastrointestinal motility, which aggravate the symptoms of duodenal obstruction and stomach retention. However, it is hard to clear the effect of spasmolytic drugs on SMA syndrome based on the case report. Probably, a clinical trial or a retrospective study could be designed to discuss the relationship between spasmolytic drugs and the acute symptoms of SMA syndrome. According to your suggestion, the manuscript has been revised as Line 89-91, Page 3, Line 164-165, 167, 169-175, Page 4.

No need to abbreviate AA, AMD.
Thank you. Abbreviations AA (aortomesenteric angle) and AMD have been removed in the manuscript. In the manuscript, abbreviated AA (abdominal aorta) has been kept to make the manuscript and figures easier to illustrate.

0.37mm must be a mistake. I suggest using mm instead of cm throughout the manuscript.

Thank you very much! 0.37mm was miswritten and has been revised as 3.7 mm. We have revised mm instead of cm throughout the manuscript on Line 103, 106, 113 and 115, Page 3.

The weakness of this case report is that there is no follow-up. How long was the patient followed? Is she still symptomatic? Did she gain weight? If this is information is not available in the medical records, maybe a simple phone call to the patient would clarify this.

Thank you, Professor. The long-term follow up is indeed an essential part of case report. We have added the - Follow-up - section following the - Case presentation - section. Please find the revised manuscript on Line 134-149, Page 4. We also collected follow-up BMI data as Figure 3 and figure legends (Line 357-360, Page 9).

The authors stress that this condition could be life-threatening and misdiagnosis is dangerous. What are the potential complications? Any reference in the literature of potentially lethal complications?

Mascolo M, et al. reported a case of severe gastric dilatation due to SMA syndrome with a history of anorexia nervosa. (Int J Eat Disord, 2015). Ugras M, et al. (Turk J Emerg Med, 2017) reported that an adolescent boy with SMA syndrome showed gastric dilatation without any previous history who was treated conservatively. Acute gastric dilation may result in severally life-threatening complications such as dehydration, metabolic alkalosis, gastric necrosis and systemic circulatory failure (J Trauma. 1987).
Please discuss any surgical treatment options with adequate references.

Thank you very much! Surgical treatment is an important part of the management. The surgical intervention will be required if medical treatment fails or the condition is severe. We have added the - Surgical treatment - section in the Discussion section. The manuscript was revised as following. (Line 233-244, Page 6)

Treatment of proximal intestinal obstruction caused by duodenum compression in SMA syndrome initially involves conservative management, especially for those rapid developed cases less than 4 weeks. However, for those patients who have persistent symptoms with nutritional support, surgical therapy could be chosen (Nutr Hosp. 2017). The alternative operations mainly included open duodenojejunostomy and laparoscopic duodenojejunostomy. Ganss A, et al. reported a prospective study in a single institution that thirty-nine SMA syndrome patients underwent duodenojejunostomy, and in thirty-two patients also performed a distal duodenum resection (J Gastrointest Surg, 2018). Laparoscopic duodenojejunostomy is considered to be feasible, safe, less morbid and effective for SMA syndrome as compared to open surgery (J Clin Diagn Res, 2017, Ann R Coll Surg Engl, 2017). Even for SMA syndrome combined with renal nutcracker syndrome due to severe weight reduction, weight gain and SMA syndrome was corrected by laparoscopic duodenojejunostomy (Korean J Gastroenterol, 2017).

Silvío Mazziotti (Reviewer 3): Although already described in literature, SMA and Nutcracker phenomenon are rare pathological conditions that sometimes can coexist. Radiologists and clinicians should always be aware of these conditions in young patients with an history of weight loss and abdominal pain.

The case is interesting and well written.

However, some points need to be clarified:
- In literature the SMA syndrome is also known as "Wilkie's syndrome". I suggest to add this denomination too.

Thank you very much! "Wilkie's syndrome" has been added in the revised manuscript. (Line 28-29, Page 1, Line 62-63, Page 2)

- The authors refer to "Nutcracker phenomenon" and not to "Nutcracker syndrome". Indeed, it is true that the patients did not suffer from hematuria, but anyway a left flank pain was detected at the abdominal examination.

Moreover, two different phenotypes of Nutcracker phenomenon/syndrome exist (anterior and posterior).

I would like that the authors could be clearer in the text about these two points.

This patient did not suffer from hematuria, but a left flank pain was detected at the abdominal examination for the first hospitalization. While, in the second hospitalization, a hematuria was found, still with a left flank pain. Therefore, we gave the diagnosis of Nutcracker syndrome instead of Nutcracker phenomenon. The revised manuscript in line 1, 35, page 1, line 53, 59, 74, page 2, line 111, 123, page 3, line 227, page 6, line 319, page 8.

- The authors report that the upper gastrointestinal double-contrast radiograph was performed after the CT-scan. Considering the age and the gender of the patient, was it really necessary to achieve a diagnosis? If yes, was that the right timing for its performance (after the CT-scan)?

In this sense, I would like that the authors could briefly describe in discussion the different imaging modalities available (especially for the "radiation-free" alternatives), the indications and the principle findings for each one.

Thank you very much! It is really an important part for the imaging diagnosis of SMA syndrome, especially for the female patient with childbearing age. Thus, according to your suggestion, we added - - Diagnosis - "Radiation-free" alternatives - - in the Discussion section. For the patient
we just reported, the CT-scan was performed to screen the cause of abdominal pain. Although she suffered from the radiation exposure, she still had a strong desire to confirm duodenal stasis and make a clear diagnosis. Thus, the upper gastrointestinal double-contrast radiograph was performed after then. In our hospital, the ultrasonography to diagnose SMA syndrome has not been conducted till now. However, the Nutcracker syndrome has been diagnosed by ultrasonographic examination, which we have added in the Case presentation section. The manuscript was revised on Line 203-218, Page 5.

Moreover, I would suggest these articles:


- In my opinion, the graphical overlay pointers of Fig. 1 c are a bit confusing. I think they should be put within the structure that they are labeling. Maybe a "zoomed" detail of the image could be easily understandable for the reader.

Thank you very much! The arrow and star icons were replaced within the structure. The zoomed structures showed in the lower left of figure 1c. Please find the revised Figure 1c and figure legends (Line 356-357, Page 9).
For the 'Availability of data and materials' section, please provide information about where the data supporting your findings can be found. We encourage authors to deposit their datasets in publicly available repositories (where available and appropriate), or to be presented within the manuscript and/or additional supporting files. Please note that identifying/confidential patient data should not be shared. Authors who do not wish to share their data must state that data will not be shared, and provide reasons for this in the manuscript text. For further guidance on how to format this section, please refer to BioMed Central's editorial policies page - http://www.biomedcentral.com/submissions/editorial-policies#availability+of+data+and+materials.

Declarations

- Ethics approval and consent to participate
- Consent to publish
- Availability of data and materials
- Competing interests
- Funding
- Authors' Contributions
- Acknowledgements
- Authors' Information

We have added the - Authors' Information – in Declarations section on Line 398-304, Page 7.