Endoscopic features for early detection of superior mesenteric artery syndrome in children

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Research Article

Keywords: Superior mesenteric artery syndrome, Children, Endoscopy, Duodenum

Posted Date: January 5th, 2021

DOI: https://doi.org/10.21203/rs.3.rs-133222/v1

Abstract

Background

Superior mesenteric artery syndrome (SMA) syndrome is a rare cause of duodenal obstruction. Diagnostic delay of SMA syndrome is common due to its rarity and lack of index of clinical suspicion. Present study aims to explore the endoscopic features for early decision to evaluate SMA syndrome in children.

Methods

In case controlled observation study, we identified three endoscopic findings in enrolled cases like as, finding I: a pulsating band like luminal narrowing of the third part of the duodenum without no expansion over one third during air insufflation for at least 15 seconds, finding II: marked dilatation of the duodenal second part during air insufflation at the duodenal third part, and finding III: bile lake in the stomach. SMA syndrome was confirmed with upper GI series in patients with endoscopic finding I or I plus more. We analyzed positive endoscopic findings related with SMA syndrome.

Results

Twenty-nine patients consisted of 18 (62.1%) cases with SMA syndrome and 11 (37.9%) cases without SMA syndrome. The 3 most common presenting symptoms were postprandial discomfort, abdominal pain, and early satiety. The initial impressions before endoscopy were functional dyspepsia (37.9%), gastritis or gastric ulcer (31.0%), and SMA syndrome (24.1%). Sixteen patients (55.2%) had the constellation of three endoscopic findings (finding I + II + III). Of them, 72.2% confirmed with SMA syndrome, and 27.3% had no SMA syndrome in upper GI series ($P = 0.027$).

Conclusions

Endoscopic examination down to the third part of duodenum can provide a clue making a decision to evaluate SMA syndrome, which includes a feature composed of three findings like as a pulsating band like luminal narrowing of duodenal third part, a marked expansion of duodenal second part, and a bile lake in the stomach.

Background

Superior mesenteric artery (SMA) syndrome is a symptom complex condition caused by the external compression of the third part of the duodenum between the aorta and SMA [1]. It commonly manifests nonspecific gastrointestinal (GI) symptoms such as upper abdominal pain, postprandial fullness, and chronic food intolerance with easy satiety, anorexia, nausea and vomiting.

Making the early diagnosis of SMA syndrome is challenging. Diagnostic delay commonly occurs due to its rarity, nonspecific symptoms, and lack of high indices of suspicion [2, 3]. Early decision to perform the optimal diagnostic tests is essential to avoid diagnostic delay of SMA syndrome. However, it is often diagnosed by incidentally identifying external compression of the duodenal third part during investigative process of the exclusion approach. The external duodenal compression can be demonstrated with upper GI series, contrast enhanced computed tomography (CECT) scan or magnetic resonance angiography (MRA). These radiologic studies can show inflation of proximal duodenum with or without gastric expansion and evidence of abrupt vertical or oblique compression of the duodenal third part by SMA [4, 5]. Furthermore, CECT scan and MRA can make measurements of the angel and distance between superior mesenteric artery and aorta.

Most patients with SMA syndrome present with upper GI symptoms. So most may commonly undergo esophagogastroduodenoscopy (EGD) for initial investigative process. Till now, it has been known that EGD hardly affords an information to make a suspicion or diagnosis of SMA syndrome. We hypothesized that upper GI endoscopy can provide the evidences reflecting the external compression of the duodenal third part in patient with SMA syndrome. We aimed to search for endoscopic features related with external compression of the third part of the duodenum by SMA through the analysis of endoscopic findings of patients who confirmed SMA syndrome.

Methods

Patient selection, study design, and data analysis

We retrospectively or prospectively had collected data on patients who underwent EGD and upper GI series with or without CECT scan because of upper GI symptoms at the Department of Pediatrics, Chungnam National University Hospital since 2007. EGD performed by the single same pediatric gastroenterologist.

The recruitment in our study was limited to patients who had endoscopic findings showing the following features: a pulsating vertical or oblique band like luminal narrowing with partial luminal opening less than one-third during air insufflation more than 15 seconds at the duodenal third part (finding I), with or without an over expansion of duodenal second part during air insufflation at the duodenal third part (finding II) with or without a large amount of bile mixed fluid in the stomach (finding III) (Fig. 1). We classified endoscopic findings into 4 features as followings: feature A by finding I, feature B by finding I and II, feature C by finding I and III, and feature D by the constellation of finding I, II and III.

All enrolled cases underwent upper GI series with or without CECT scan to confirm SMA syndrome. A pediatric radiologist performed and interpreted the upper GI series. Positive findings of upper GI series consistent with SMA syndrome included findings of (i) dilatation of the first and second parts of the duodenum, (ii) abrupt vertical or oblique compression of the duodenal third part, (iii) antiperistaltic to and

pro waves of contrast from the proximal to the obstruction, and (iv) significant delays in gastroduodenal transit [4, 5] (Fig. 2).

All enrolled cases were divided into 2 groups according to the results of upper GI series: group I included patients with SMA syndrome and group II included control patients without SMA syndrome. The medical records, including endoscopic findings, demographics, clinical presentations, growth status with weight, weight-for-age Z score, body mass index (BMI), BMI-for-age-Z score, and weight-for-height Z score were analyzed. The Z scores were assessed according to the stature percentiles of the Korean National Growth Charts drafted by the Korea Centers for Disease Control and Prevention.

Statistical analysis

Statistical analysis on the data was performed by using SPSS version 19.0 (SPSS Inc. Chicago, IL, USA). Demographics, duration of illness and growth status were assessed by the Mann-Whitney *U* test. Clinical manifestations and endoscopic findings were assayed using the Fisher's exact test. A value of $p < 0.05$ was considered significant.

Results

The total number of patients available for this study was 29; group I included 18 patients with SMA syndrome and group II included 11 patients without SMA syndrome. The median ages of group I and II were 11.5 years (range, 7.8–16.2 years) and 13.2 years (range, 10.5–15.8 years) and male to female ratios were 0.8:1 in group I and 0.6:1 in group II, respectively. The median duration of symptoms before diagnosis was 68 days (range, 5-760 days) and 30 days (range, 5-1825 days) in each of the groups. The three most common presenting symptoms were postprandial discomfort (61.1% in group I and 54.5% in group II), abdominal pain (55.6% in group I and 90.9% in group II), and early satiety (50.0% in group I and 18.2% in group II). Less common symptoms included anorexia (33.3% in group I and 9.1% in group II), vomiting (27.8% in group I and 18.2% in group II), and nausea (16.7% in group I and 9.1% in group II). Weight loss was noted in 38.9% and 18.2% in group I and II, respectively. These presenting symptoms and signs were not statistically different between the groups (Table 1).

Table 1
Demographic Data and Clinical Features in Both Groups

Variable	Group I, n = 18(%)	Group II, n = 11(%)
Age (years)*	11.5 (7.8–16.2)	13.2 (10.5–15.8)
Sex: male/female (ratio)	8/10 (0.80)	4/7 (0.57)
Duration of illness (days)	68 (5-760)	30 (5-1825)
Presentation		
Postprandial discomfort	11 (61.1)	6 (54.5)
Abdominal pain	10 (55.6)	10 (90.9)
Early satiety	9 (50.0)	2 (18.2)
Anorexia	6 (33.3)	1 (9.1)
Vomiting	5 (27.8)	2 (18.2)
Nausea	3 (16.7)	1 (9.1)
Weight loss	7 (38.9)	2 (18.2)
SMA, superior mesenteric artery		
Results of age and duration of illness expressed as median (range)		
*P = 0.041		

Table 2 shows growth status including the mean body weight, weight-for-age Z score, BMI, BMI-for-age Z score, and weight-for-height Z score. These parameters were not significantly different in both groups.

Table 2
Comparison of Growth Status in Both Groups

Variable	Group I (n = 18)	Group II (n = 11)
Weight, kg	36.4 ± 10.9	42.1 ± 9.7
Weight-for-age Z score	-0.69 ± 0.92	-0.68 ± 1.00
BMI, kg/m^2	15.7 ± 2.0	17.3 ± 2.3
BMI-for-age Z score	-1.37 ± 1.02	-0.95 ± 1.10
Weight-for-height Z score	-1.50 ± 1.52	-0.77 ± 1.59
SMA, superior mesenteric artery		
Statistically no significance between two groups		

The initial impression and final diagnosis of 29 cases are shown in Table 3. The three most common initial impressions before EGD were functional dyspepsia (37.9%), gastritis or gastric ulcer (31%), and SMAS (24.1%). Other impressions were reflux esophagitis (10.3%), functional abdominal pain (6.9%), small bowel obstruction (3.4%), and Crohn's disease (3.4%). All cases underwent EGD and upper GI series with or without CECT scan. SMA syndrome was confirmed in 62.1% of the enrolled cases.

Table 3
Initial Impression and Diagnosis of 29 Patients

Variable	Impression (%)	Finial diagnosis (%)
Functional dyspepsia	11 (37.9)	5 (17.2)
Gastritis or gastric ulcer	9 (31.0)	5 (24.1)
Superior mesenteric artery syndrome	7 (24.1)	18 (62.1)
Reflux esophagitis	3 (10.3)	1 (3.4)
Functional abdominal pain	2 (6.9)	1 (3.4)
Small bowel obstruction	1 (3.4)	0 (0.0)
Crohn's disease	1 (3.4)	0 (0.0)
Bile reflux gastropathy	0 (0.0)	4 (13.8)
Some patients had more than one disease at initial impression and diagnosis.		

The results of EGD were as below: one case for feature A, three cases for feature B, nine cases for feature C, and sixteen cases for feature D. The cases of SMA syndrome confirmed by upper GI series according to each EGD feature were as follows: none (0%) of one case with feature A, one (33.3%) of three cases with feature B, four (44.4%) of nine cases with feature C, and thirteen (81.3%) of sixteen cases with feature D. Figure 3 shows the evaluation flow of the total enrolled cases and the results.

The most common endoscopic finding associated with SMA syndrome was feature D, which was documented in 72.2% in group I and 27.3% in group II, respectively ($P = 0.027$) (Table 4). There were no statistically significant in feature A, B, and C between the groups.

Table 4
Comparison of Endoscopic Findings in Both Groups

Variable	Group I, n = 18 (%)	Group II, n = 11 (%)	*P* value
Feature A (Finding I)	0 (0.0)	1 (9.1)	0.379
Feature B (Finding I + II)	4 (22.2)	5 (45.4)	0.114
Feature C (Finding I + III)	1 (5.6)	2 (18.2)	0.539
Feature D (Finding I + II + III)	13 (72.2)	3 (27.3)	0.027

Fining I, a vertical pulsatile compression and opening less than one third of the duodenal third part with air insufflation during endoscopy; Finding II, a marked duodenal second part expansion and/or gastric distension; Finding III, a large amount of bile mixed fluid (bile lake) in the stomach; SMAS, superior mesenteric artery syndrome

Discussion

The aim of the present study was to evaluate whether the EGD provides clues on early decision to perform a diagnostic test for SMA syndrome. To the best of our knowledge, no study investigating on the endoscopic features associated with SMA syndrome has yet been conducted. Our study suggests that endoscopic observation up to the third part of the duodenum can give a significant information in determine to perform diagnostic tests of SMA syndrome. The endoscopic features in our study were the three constellations of pulsating vertical or oblique band like luminal narrowing of the third part of the duodenum with luminal expansion no more than one third during air insufflation over 15 seconds, proximal duodenal over distension during air insufflation in the duodenal third part, and bile lakes in the stomach.

SMA syndrome is a disease caused by external compression of the duodenal third part between the aorta itself and the SMA branching from the aorta at the level of the first lumbar vertebra. Although the exact pathogenesis of this syndrome is unknown, it is often associated with rapid linear growth without or small weight gain, rapid weight loss, external cast compression, an abnormally high position of the ligament of Treitz, and unusually a low origin of the SMA [4–8]. Rapid weight loss and certain metabolic states cause depletion of the mesenteric and retroperitoneal fat, subsequently decreasing the aortomesenteric distance (AMD) (1). However, weight loss and low BMI do not necessarily relate with SMA syndrome. Indeed, no substantial difference were observed in the weight and BMI between the SMA syndrome and non-SMA syndrome groups in our study.

SMA syndrome often presents with non-specific upper GI symptoms such as nausea, vomiting, early satiety, anorexia, postprandial discomfort, and abdominal pain [9, 10]. In our study, postprandial discomfort, abdominal pain, and early satiety were the major symptoms. Symptoms can be acute or chronic. Making a diagnosis of SMA syndrome is challenge and diagnosis may commonly be delayed for a period of time after symptoms appear. The median duration of symptoms before diagnosis varies from 5 to 30 days and up to 18 months according to the literature [9, 11, 12]. In our study, the median was 68

days (range: 5 to 760 days) until diagnosis. The reasons for varying duration of diagnostic delays may be due to rarity, nonspecific symptoms, and a lack of high suspicion of clinical index of SMA syndrome. Many clinicians remain unaware of this syndrome. So this syndrome may commonly be diagnosed in the process of excluding other suspected conditions [2, 3]. And many patients may suffer from upper GI symptoms and related comorbidity such as weight loss, malnutrition, and poor quality of life until a diagnosis was made.

A confirm diagnosis of SMA syndrome is made mainly through the conventional upper GI series. CECT scan is also known to assist a diagnosis of SMA syndrome with findings of proximal duodenal dilatation and decreased AMD and aortomesenteric angle (AMA). An AMA of less than 22–25° and an AMD of less than 8 mm correlated well with the development of symptoms of SMA syndrome in adults [13–16]. Sinagra et al. [17] reported that AMD seems to be more accurate rather than AMA to diagnose SMA syndrome. And these are consequently used as cutoff values for diagnosis of SMA syndrome in adults. However, there is no cutoff value for diagnosis in children and the normal ranges of these are variable in this age. So, interpretation should be performed with caution when using CECT scan for diagnosis of SMA syndrome in children [18, 19]. Ultrasound color Doppler imaging or MRA has recently been proposed as alternative modalities rather than CECT scan for the diagnosis in cases of suspected SMA syndrome [20].

In the practice, SMA syndrome can be linked to inadequate investigation and ineffective treatment due to its rarity and unawareness by clinicians, which may lead to electrolyte imbalance, dehydration, weight loss, malnutrition, and even death [3, 11]. So, early investigations and detection of SMA syndrome is important to avoid these problems. Most patients with SMA syndrome present nonspecific upper GI symptoms. Thus EGD is usually first performed to differentiate the upper GI diseases such as peptic diseases, functional dyspepsia, and other disease but, it is not possible to make a diagnosis of SMA syndrome with EGD. Lippl et al. [5] suggested that endoscopic findings such as duodenal dilatation, liquid stasis, and antiperistaltic waves may indicate SMA syndrome. Meanwhile, Sundaram et al. [20] stated that although the Lippl's suggested endoscopic findings may indicate duodenal obstruction, a diagnosis of SMA syndrome cannot always be made with certainty with those findings. They recommended using both endoscopic ultrasound and endoscopy for the diagnosis of SMA syndrome. Unlike this, Sinagra et al. [17] suggested that pulsatile extrinsic compression in the third part of the duodenum by EGD is the most reliable finding to suspect SMA syndrome. Cappell et al. [21] reported a case with SMA syndrome with an endoscopic photograph showing pulsatile band-like luminal narrowing of the third part of duodenum which was gradually, partially opened by moderate air insufflation. We observed that diagnostic yield of SMA syndrome was significantly higher in patients with endoscopic feature D compared to those without that. The sensitivity and the positive predictive value having SMA syndrome in cases with endoscopic type D were 72.2% and 81.3%, respectively. So we suggest that EGD can be a useful in deciding to investigate SMA syndrome when endoscopic feature D is documented through examination up to the duodenal third part.

Conclusion

In conclusion, clinical symptomatology, physical examination, and endoscopic information are important for early suspicion of SMA syndrome. Clinicians should consider as differential diagnosis of SMA syndrome in patients with symptoms such as postprandial distress, abdominal pain, early satiety, anorexia with or without weight loss. Even though our study has some limitations due to the small number of enrolled patients and lack of endoscopic follow-up after improvement of symptoms, we recommend to examine up to the duodenal third part when thick bile stained fluid retention (bile lake) is noted in the stomach during EGD in patient with upper GI symptoms like as postprandial discomfort, abdominal pain, and early satiety. We suggest endoscopic feature D would be a substantial clue to reach an early suspicion and make a decision for evaluation of SMA syndrome. Further clarification with future large scale case studies are warranted.

Abbreviations

SMA: Superior mesenteric artery; GI: gastrointestinal; CECT: contrast enhanced computed tomography; MRA: magnetic resonance angiography; AMD: aortomesenteric distance; AMA: aortomesenteric angle; EGD: esophagogastroduodenoscopy

Declarations

Ethics approval and consent to participate

Present study was conducted with the approval of Ethics Committee/institutional review board of Chungnam National University Hospital (IRB No. 2013-06-014-001). All procedures in this study involving human participants was performed in accordance with the declaration of Helsinki. Written informed consent was obtained from all participant's parents or legal guardians for participants. All data published here are under the consent for publication.

Consent for publication

Not Applicable.

Availability of data and materials

The datasets generated and analyzed during the present study are available

from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

Funding

Not applicable.

Authors' contributions

JYK designed and performed most of the investigation, data analysis and wrote the most of manuscript; MSS and SHL contributed to write some part of discussion. All the authors have read and approved the manuscript for publication

Acknowledgements

Not Applicable.

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