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Intestinal malrotation in patients with situs anomaly: Implication of the relative positions of the superior mesenteric artery and vein

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ABSTRACT

Purpose: To assess the usefulness of the relative position of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) in diagnosing intestinal malrotation in situs anomaly.
Materials and methods: From January 2004 to April 2015, 33 patients with situs anomalies were enrolled in this study who underwent abdominal USG, CT or MRI as well as upper gastrointestinal series (UGIS) or surgery: situs inversus (n = 16), left isomerism (n = 10), and right isomerism (n = 7); age 21.2 ± 23.2 years (mean ± standard deviation), range 0–72 years. The intestinal malrotation was confirmed with UGIS and/or operation in 16 patients. Relative positions of the SMV to the SMA were classified into four groups by reviewing abdominal USG, CT, or MRI: right sided, left sided, ventral sided, and dorsal sided. The incidence of malrotation was analyzed for each group.
Results: In 16 patients with situs inversus, there was reversed SMA-SMV relationship: left sided (n = 11) or ventral sided (n = 5). One situs inversus patient with ventral sided SMV had intestinal malrotation (6.25%). 17 patients with situs ambiguus showed various SMA-SMV relationships (ventral sided, n = 7; left sided, n = 5; right sided, n = 4; dorsal sided, n = 1). Among them, 15 patients (88.2%) had intestinal malrotation. Two patients with normal rotation had either right sided or dorsal sided SMV.
Conclusion: Situs ambiguus was commonly associated with intestinal malrotation with a variable SMA-SMV relationship. Reversal of the mesenteric vascular relationship was observed in situs inversus with normal rotation, not excluding the possibility of intestinal malrotation.

1. Introduction

It is well-established that an abnormal relative position of the superior mesenteric vein and artery can be present in patients with intestinal malrotation [1]. Intestinal malrotation is a potentially life threatening condition when complicated with midgut volvulus or associated with intestinal obstruction due to the presence of Ladd's bands.

Situs refers to the arrangement of organs, such as the lungs, liver, spleen, and atria in reference to the midline, i.e., the right-left relationship of the organ arrangement. Situs inversus refers to the reversed right-left relationship of the organ arrangement. Situs ambiguus, i.e. heterotaxy refers to the arrangement of the organs that is neither situs solitus nor situs inversus. Heterotaxy is characterized by symmetric arrangement of some body organs such as airway, branch pulmonary arteries and atrial appendages. Therefore, heterotaxy is divided into heterotaxy with right isomerism and heterotaxy with left isomerism, where isomerism is defined as symmetrical arrangement of organs that are normally asymmetrical [2,3]. Although situs ambiguus is not a common congenital anomaly (incidence of 0.01%) [4–6], it is known to be frequently associated with intestinal malrotation (40–90%) [2,4,7]. Therefore, it is important to identify radiological clues to diagnose intestinal malrotation in patients with situs anomaly. Thorough evaluation for a complete retroperitoneal course of the duodenum and the lower abdominal positioning of the cecum producing a

Abbreviations: USG, ultrasonography; CT, computed tomography; MRI, magnetic resonance imaging; UGIS, upper gastrointestinal series; SMA, superior mesenteric artery; SMV, superior mesenteric vein.

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sufficiently broad mesenteric base is required in order to safely exclude intestinal malrotation [8]. However, such evaluation can be difficult and require additional studies such as fluoroscopic studies or even surgery. In clinical practice, the relationship between the superior mesenteric artery and vein has been used as a useful initial clue to evaluate abnormalities of intestinal rotation in patients with normal situs because it is easy to evaluate on USG or other tomographic studies, although false positive or false negative cases can be present [1,9]. To our knowledge, there has been no comprehensive study about the relative position of the superior mesenteric artery and vein as well as the association between intestinal malrotation and the relative position of the mesenteric vessels in patients with abnormal situs. Therefore, the purpose of this retrospective study was to assess the usefulness of the relative relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) for predicting the intestinal malrotation in patients with situs anomalies.

2. Materials and methods

The Institutional Review Board of Seoul National University Hospital approved this retrospective observational study and waived the requirement for patient informed consent.

2.1. Patients

In order to identify patients with situs anomaly, we searched the radiology reports database from January 2004 to April 2015. USG, CT or MRI examinations with any of the following keywords in their final reports were investigated; "situs"; "isomerism"; "heterotaxia"; "polysplenia"; "asplenia"; and "inferior vena cava interruption". From this search; 1918 patients were found. Among them; only 39 patients underwent gastrointestinal series (UGIS); which is a fluoroscopic examination of upper gastrointestinal tract; and/or abdominal surgery; from which intestinal rotation status could be confirmed. Among 39 patients; 6 patients were excluded from the study; as the relative positions of the SMA and the SMV could not be adequately evaluated on USG; CT or MR images. Finally; 33 eligible patients comprised the study cohort (20 male and 13 female; mean age 21.2 ± 23.2 years; range 0–72 years).

2.2. Situs anomaly

Two radiologists (C.K.S and C.Y.H) identified the type of situs anomaly for each of the 33 patients, in consensus, by reviewing the findings of all available imaging studies, including chest radiography, USG, CT and MRI. Situs anomaly was divided into two groups: situs inversus, and situs ambiguus. Situs inversus was defined when the normal organs such as lungs, liver, spleen, and atria were positioned reversed (i.e. reversed arrangement of organs) [2]. Situs ambiguus was subdivided into two groups: right-sided isomerism (asplenia) and left-sided isomerism (polysplenia), where isomerism indicates the symmetrical arrangement of organs that are typically asymmetrical. Right isomerism was defined when bilateral right-sidedness, including bilateral 'right' lungs and atria, and absence of the spleen was noted. Left isomerism was defined by a tendency towards bilateral left-sidedness with bilateral 'left' lungs and atria, and multiple spleens were noted [3].

2.3. Intestinal malrotation

The presence of intestinal malrotation was identified by evaluating findings from fluoroscopic studies and/or surgery. In 26 patients, UGIS was performed in the standardized way, with both lateral and AP views on each patient, using barium as contrast media. The same two radiologists reviewed UGIS retrospectively to determine the intestinal rotation status of each patient. Intestinal rotation was considered normal when the duodenojejunal junction (ligament of Treitz) was located laterally to the ipsilateral vertebral pedicle with the stomach and at the level of the duodenal bulb and the cecum was located in the lower abdomen contralateral to the duoduenojejunal junction. In all other cases, intestinal arrangement was considered malrotated [2]. In 10 patients, surgery was performed and the intestinal rotation status determined based on the surgical findings.

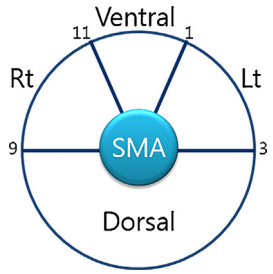

Fig. 1. Classification diagram for the relative position of the SMV to SMA.

2.4. Relative position of the SMV and SMA

Normal relative position of the SMV to SMA was defined when the SMV is located at 9–11 o'clock relative to the SMA [1]. The relative position of the SMV to the SMA was determined at the level of the proximal SMA and SMV by reviewing the images from the abdominal USG (n = 10), CT (n = 22) and/or MR (n = 1). The relative position of the SMV to SMA was divided into four groups: [1] ventral, when the SMV is located at 11–1 o'clock relative to the SMA; [2] right sided, when the SMV is located at 9–11 o'clock relative to the SMA; [3] left sided, when the SMV is located at 1–3 o'clock relative to the SMA; and [4] dorsal, when SMV is located at 3–9 o'clock relative to the SMA (Fig. 1). We evaluated the association between the relative position of the SMA to the SMV with the presence of intestinal malrotation in patients with situs anomaly.

2.5. Statistical analysis

Fisher's exact test was performed to analyze association between intestinal malrotation and situs anomalies (i.e. situs inversus and ambiguus). All statistical analyses were performed with commercially available software, SPSS 21.0 for Windows (SPSS Inc, Chicago, IL, USA). $P < 0.05$ was considered to indicate a significant difference.

3. Results

3.1. Types of situs anomaly

In 33 patients with situs anomaly, 16 patients exhibited situs inversus (Fig. 2), with the other 17 patients having situs ambiguus (Fig. 3). Among those exhibiting situs ambiguus, 7 patients had right isomerism, and the other 10 patients had left isomerism.

3.2. Association of malrotation with situs anomaly

Intestinal malrotation was confirmed in 16 out of 33 patients with a situs anomaly. In 6, the diagnosis was based on UGIS alone, in 3 on the combination of UGIS and surgical findings while the remaining 7 patients were operated without UGIS and the diagnosis was surgically confirmed.

Among the 16 patients with situs inversus, 15 patients had normal rotation (93.75%), and only 1 patient (6.25%) exhibited mal-