Double cancer of the stomach and oesophagus with situs ambiguous with polysplenia: The importance of preoperative evaluation

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Abstract
A rare case of double cancer with situs ambiguous with polysplenia is presented. A 58-year-old patient was initially diagnosed with an early gastric cancer. On evaluation, the computed tomography of the abdomen demonstrated situs ambiguous with polysplenia. We performed a subtotal gastrectomy with the stomach being reconstructed in a Billroth-II fashion. Three months after the operation, he again visited our department complaining nausea and dysphagia. Examinations confirmed the other oesophageal malignancy with advanced stage. Because of unfamiliarity to situs anomaly and rarity of double cancer, we missed another coexistent cancer. This is the first case presentation of a double carcinoma occurring in a patient with situs ambiguous with polysplenia. The literature is reviewed and the importance of preoperative evaluation is discussed.

Keywords: Double cancer; Polysplenia; Situs ambiguous

1. Introduction
Situs ambiguous, or heterotaxy, implies a disordered organ arrangement in the chest or abdomen. This abnormal arrangement of body organs is different from the orderly arrangement seen in situs solitus or situs inversus. Polysplenia is a type of situs ambiguous characterised by left isomerism (bilateral left sidedness) [1,2–4]. This syndrome has been called situs ambiguous with polysplenia (SAP) or polysplenia syndrome.
The occurrence rate of multiple primary malignant neoplasms in patients with malignant disease is reported to be about 6.4% [3]. For the patients with gastric cancer, the relative risk of a second cancer is twice as high as the incidence of cancer in the general population, especially in the alimentary organs, and second malignancies have been revealed to be the first cause of death [4].

We recently encountered a patient having SAP and gastric cancer who was finally diagnosed as having another oesophageal cancer during the postoperative period. We report this case because of its rarity, and we wish to emphasise the need for a detailed preoperative evaluation of the alimentary tract in patients with a situs anomaly.

2. Case report
A 58-year-old man visited a local hospital and he presented with epigastric pain. Endoscopy showed no pathological finding in oesophagus but showed a small depressed lesion on the greater curvature of the gastric antrum, and the biopsy revealed a histological diagnosis of a moderately differentiated adenocarcinoma. He was referred to our hospital in January 2003 for further evaluation and surgical treatment. Chest X-ray and electrocardiography showed the normal thoracic organs (Fig. 1). Computed tomography (CT) of the abdomen revealed SAP (Fig. 2). All the other laboratory data including
the tumour markers (carcinoembryonic antigen (CEA) and cancer antigen (CA 19-9)) were within normal limits. The patient underwent a subtotal gastrectomy with the stomach being reconstructed in a Billroth-II fashion. During the operation, the stomach was located in the right hypochondrium with the duodenum to the left of the midline. Polysplenia was present in the right upper quadrant and the liver was situated in the left hypochondrium. The colon showed a mirror image of a 180° malrotation, with the caecum and appendix in the left hypochondrium (Fig. 3). Gross appearance of the resected stomach showed a depressed type carcinoma that was 0.5 cm × 0.5 cm. Histological examination of the specimen demonstrated that the cancer was confined to the mucosal layer without lymph nodes metastasis. His postoperative course was uneventful and he was discharged on the 15th postoperative day. Three months after the operation, he again visited our department complaining nausea and dysphagia. An endoscopic examination demonstrated an oesophageal mass and an oesophagogram showed diffuse oesophageal wall thickening in the distal oesophagus suggesting oesophageal malignancy (Fig. 4). Biopsies of the lesion confirmed squamous cell carcinoma of the oesophagus. He refused the recommended operation and had an oesophageal stent inserted, and he received chemo-radiation therapy.
3. Discussion

There are three types of situs dictated by the arrangement of organs that are normally asymmetrical. Situs solitus occurs when these organs are in the normal position and situs inversus occurs when this arrangement is reversed. Situs ambiguous (heterotaxy) is a third and abnormal type of situs in which the relationship of the atria and the viscera is inconsistent [2,5]. Situs ambiguous can be divided into two broad groups: right-sided isomerism (asplenia) and left-sided isomerism (polysplenia); isomerism describes the symmetrical arrangement of the organs that are normally asymmetrical. In polysplenia syndrome, however, there is no single anomaly that is pathognomonic for this condition. The affected patients have a lower prevalence of congenital heart disease (50–90%) and less severe defects than those with situs ambiguous with asplenia [1,2]. The majority of patients with polysplenia syndrome die by the age of 5 years. This high mortality rate is mainly due to the severe cardiac anomalies. In cases with an absence of severe cardiac or visceral abnormalities, as was observed in the current case, asymptomatic survival into adulthood is possible [5,6]. The anomaly is often incidentally discovered on computed tomography or magnetic resonance (MR) imaging when these patients are evaluated for other medical reasons.

The incidence of situs inversus totalis ranges between 1:1000 and 1:20,000 depending on the population surveyed [7–9]. In 1959, Varano et al. [9] reviewed the literature since 1946, and they found that there were 17 cases (2.3%) of partial situs inversus among 722 cases of situs inversus. Thus, we could estimate that there is a very low incidence of situs ambiguous and furthermore, like the presented case, double cancer with situs anomaly is extremely rare. The occurrence of multiple primary malignancies in the same individuals is also an interesting subject for investigation, Yoshino et al. [4] have reported on 22,163 gastric cancer patients from 66 hospitals, and this revealed 445 patients (2%) with multiple primary malignancies. Hundred and thirty-seven (30.8%) of the patients had synchronous tumours and the most frequent site was the colon and rectum followed by the uterus, the oesophagus and the breast. Although this anomaly is not considered to be a premalignant entity, several malignant neoplasms have been sporadically reported in association with situs anomaly [10–13]. The combination of congenital anomaly and neoplasm has generally been considered to be coincidental rather than a reflection of a common pathogenesis. There has been no previous documentation on a double carcinoma occurring in a patient with SAP, and to the best of our knowledge, this is the first such case in the English literature.

The diagnosis of a coincident malignancy is not difficult to make when bearing this congenital anomaly in mind. However, we did not consider the possibility of a second carcinoma at that time and unfortunately, the routine preoperative G-I tract examination missed the unrelated oesophageal carcinoma. According to the data of malignant cases registered to Central Cancer Registry Centre in Korea, gastric cancer is the most common malignant neoplasm in Korean population, but oesophageal cancer is relatively rare one (11th rank). The frequencies of gastric cancer and oesophageal cancer in malignant cases were 20.9 and 2.0%, respectively [14]. Thus most of Korean gastroenterologists and radiologists may tend to concentrate more on detecting gastric lesion than oesophageal lesion. Furthermore, striking CT findings of the patient attracted our attention to combined congenital anomaly.

From the surgical point of view, Sands et al. [15] have offered a multisystemic prescreen evaluation approach, and the careful preoperative anatomic assessment before an operation is very important because of the multiplicity of abnormalities that are possible with this anomaly and especially in cases that are complicated by polysplenia syndrome. We believe it is necessary to screen patients with situs ambiguous for malignancies as well as for malformations or anomalies of other organs. When a patient with situs inversus or when a splenic anomaly is encountered, the anomalies should be elucidated by using various imaging technologies to determine an appropriate surgical treatment. We think that if a thorough anatomical mapping is taken into consideration, the condition of situs ambiguous with polysplenia should not hinder the surgical procedure. Yamaguchi et al. [16] have recently reported a successful laparoscope-assisted distal gastrectomy for early gastric cancer in a patient with situs inversus totalis,
and in fact, we also experienced no anatomical problems when performing a curative gastric resection with D2 lymph node dissection for our patient.

In summary, this report demonstrates the possibility, even though it may be rare, that multiple primary malignancies can occur in cases of SAP. The careful assessment of abnormalities by the preoperative evaluation was very important for the surgical treatment of this patient to rule out another malignancy. Although a busy surgeon may expect to encounter this anomaly only once or twice in a lifetime, the surgeon is responsible for avoiding embarrassing and potentially fatal errors by familiarising himself with this anomaly.

Conflict of interest statement
None declared.

References