Situs Inversus Totalis with Malignant Lymphoma of the Stomach: Report of a Case

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Abstract

Situs inversus totalis is a rare congenital anomaly that often occurs concomitantly with other disorders. We report a case of situs inversus totalis with malignant lymphoma of the stomach, which was successfully treated by surgery followed by chemotherapy and irradiation. The patient was a 51-year-old woman who presented with colicky pain in the left upper quadrant of her abdomen. Chest X-ray showed a right-sided heart, and ultrasonography and computed tomography (CT) of the abdomen showed a situs inversus totalis with multiple gallstones in the gallbladder. Tree-dimensional reconstructed CT of the abdomen showed no other malformations coexisting with situs inversus totalis, but a barium upper gastrointestinal series found an inverted stomach and an elevated tumor with ulceration in the center, localized in the antrum of the stomach. First, we performed a cholecystectomy, followed by a total gastrectomy with dissection of the lymph nodes and splenectomy, and Roux-en-Y reconstruction. Histopathological examination confirmed a diagnosis of malignant lymphoma of the stomach (diffuse large B-cell type) with metastasis to the regional lymph nodes. Chemotherapy using the CHOP regimen was given three times, starting 1 month postoperatively. A follow-up CT scan showed enlargement of one lymph node around the abdominal aorta and irradiation was delivered to the area of the inverted Y in the abdomen. At the time of writing, 10 months after surgery, the patient is well with no signs of recurrence and leading a normal life. Careful preoperative assessment is very important for determining the most appropriate surgical procedure in patients with situs inversus totalis associated with a malignancy.

Key words Malignant lymphoma · Stomach · Situs inversus totalis

Introduction

Situs inversus totalis is a congenital anomaly, occurring at an incidence of 1 in 10000–50000 live births.1 It is defined as a complete mirror image of the thoracic and abdominal viscera, although this does not seem to affect normal health or life expectancy, and it is not considered to be premalignant. However, a few cases of primary gastrointestinal malignancy developing concomitantly with situs inversus have been reported, although there is no known association. Most of these cases are carcinoma (epithelial malignancy), whereas nonepithelial malignancy is very rare. We recently treated a patient with situs inversus totalis and malignant lymphoma of the stomach. This case is reported because of its rarity, and the surgical implications are discussed in relation to the relevant literature.

Case Report

A 51-year-old woman was referred to our department for investigation and treatment of colicky pain localized in the left upper quadrant. Chest X-ray revealed a right-sided heart, and a barium upper gastrointestinal series showed an inverted stomach and an elevated tumor with ulceration in the center, localized in the antrum of the stomach (Fig. 1). Endoscopic examination showed the same lesion in the stomach, as well as a small elevated lesion in the upper part of the stomach. Biopsies of both lesions confirmed malignant lymphoma of the stomach of the diffuse large cell type. Ultrasonography and computed tomography (CT) of the abdomen showed situs inversus totalis with multiple stones in the
gallbladder. CT also showed enlarged lymph nodes around the celiac artery, probably indicating lymph node involvement by malignant lymphoma of the stomach. Three-dimensional reconstructed CT (3D-CT) of the abdomen showed no malformations concomitant with situs inversus totalis (Fig. 2). According to the Ann Arbor\(^2\) and Naqvi\(^3\) classifications, the lymphoma was evaluated to be stage II. Under the diagnosis of malignant lymphoma of the stomach and cholelithiasis with situs inversus totalis, we performed a laparotomy. Abdominal exploration confirmed situs inversus totalis (Fig. 3). First, a cholecystectomy was done, followed by total gastrectomy with dissection of the lymph nodes (D2\(^4\)) and splenectomy, and finally, Roux-en-Y reconstruction. Histopathological examination confirmed a diagnosis of malignant lymphoma of the stomach of diffuse large cell type (Fig. 4), with metastasis to the regional lymph nodes (nos. 5, 6, and 7).\(^4\) Immunohistochemical staining showed that these cells were positive for CD20 and CD79a, indicating diffuse large B-cell lymphoma (LSG classification\(^5\)). She was commenced on chemotherapy using the CHOP regimen (cyclophosphamide 1200 mg, adriamycin 50 mg, vincristine 80 mg, and prednisone 65 mg) 1 month postoperatively, which was given three times. A follow-up CT scan 3 months postoperatively revealed an enlarged lymph node about 2 cm in diameter, around the abdominal aorta. Therefore, 1.8-Gy doses of irradiation (total 36 Gy) were delivered to the area of the inverted Y in the abdomen, resulting in disappearance of this swollen lymph node.

The patient is currently well and leading a normal life with no sign of recurrence, 10 months after her operation.

**Discussion**

Situs inversus totalis is a rare congenital anomaly that occurs once in every 10 000 to 50 000 births.\(^1\) Several abnormalities are associated with situs inversus totalis, including cardiac malformation, bronchiectasis (Kartagener’s syndrome),\(^6\) polysplenia, genitourinary anomalies, and so on.\(^1,7,8\) Situs inversus is considered to have a genetic predisposition that is autosomal recessive. Brueckner et al. recognized a mutation on chromosome 12 in mice with inversus viscerum,\(^9\) and Kato et al. reported the case of a Japanese girl with heterotaxia associated with a de novo balanced translocation (6;18)

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![Fig. 1. Upper gastrointestinal tract series showed a tumor with central ulceration in the antrum with situs inversus (arrow)](image1)

![Fig. 2. A,B. Three-dimensional computed tomography findings. A Multiple-planar reformation view demonstrated situs inversus totalis. B Shaded surface display showed no anomalies of the arteries)](image2)