



Situs Inversus Totalis: Does Colon Cancer Sidedness Matter? A Rare Case Report

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ABSTRACT

Situs inversus totalis is a rare congenital abnormality characterized by a mirror-image transposition of both the abdominal and the thoracic organs. While this anomaly is known since the ancient times, practicing doctors do not have much experience with it. Laterality is established early in development, and any failure in that process might lead to a wide variety of disorders which may be partial or complete. Situs solitus describes the normal anatomy, situs inversus is the complete reversal, and situs ambiguous is used for any other abnormality of left-right development.

Keywords: Situs inversus totalis, Colon cancer, Sidedness, Cetuximab, Anti EGFR

INTRODUCTION

Situs Inversus Totalis (SIT) is a rare congenital condition, characterized by complete transposition of the thoracic and abdominal viscera. It is associated with normal life expectancy unless other anomalies present [1]. Few case reports of SIT found in association with colon cancer however no available data on response to specific biologic therapy [2,3].

CASE PRESENTATION

In May 2020, a 40-year-old male patient, soldier, with no history of chronic illnesses, no family history of malignancy and he is nonsmoker. He presented with two months history of right hypochondrial pain associated with low grade fever at night, fatigue, and generalized bone pain with significant loss of weight. Clinical examination: Eastern cooperative oncology group performance status 1. Abdomen slightly distended soft and non-tender, hepatomegaly with liver span 18 cm, no other significant clinical findings.

Complete blood counts normal, alanine transaminase: 40 units per liter (u/L), aspartate transaminase: 146 u/L, lactate dehydrogenase: 1549 u/L, alkaline phosphatase: 595 u/L, calcium: 2.47 millimole per liter, Carcinoembryonic Antigen (CEA): 1501.8 nanograms per milliliter (ng /ml).

Baseline Chest, Abdominal and Pelvic (CAP) Computed Tomography (CT) scan showed: SIT, mural thickening of the sigmoid, peri-colic necrotic soft tissue mass, small pelvic mass, mesenteric lymph node metastasis, liver is enlarged with multiple hypodense (non-enhancing) spherical nodules through hepatic parenchyma, largest 2.5 cm, numerous bone lesions

Bone scan: Multiple spots of active trace uptake at skull, rib, cervical, dorsal, lumbo-sacral, pelvic, both scapular, clavicular, and femoral bones.

Lower gastro-intestinal endoscopy showed recto-sigmoid mass at 19 cm from the anal verge extended to the whole circumference of the lumen with normal appearance of other parts of the colon and biopsy confirmed moderate differentiated adenocarcinoma.

He was started on systemic chemotherapy in the form of leucovorin calcium, fluorouracil, and oxaliplatin (FOLFOX). He was also given Denosumab 120 mg for bone metastasis. Upon presentation for second cycle, he showed modest clinical improvement and at that time we got the result of RAS-Wild type then cetuximab was added. During evaluation for third cycle of chemotherapy, he noticed clinically significant improvement of his symptoms. He had grade I cetuximab-induced acne-like rash on his face. CEA decreased to 979 ng/ml.

CAP CT scan post week 8 of systemic therapy reported a remarkable regression of rectal thickening, peri-rectal mass, mesenteric and pelvic lymph nodes and hepatic lesions.

RESULTS AND DISCUSSION

Cetuximab is an Epidermal Growth Factor Receptor (EGFR) inhibitor used for the treatment of metastatic colorectal cancer. It is approved for RAS wild-type metastatic colorectal cancer [4]. We are reporting a rare case of un-resectable stage IV colon cancer in a SIT and if sidedness of colon cancer matter to decide options of selecting initial antineoplastic.

The primary tumor side has the strongest predictive value for EGFR inhibitor's response in the first-line treatment of patients. The phase III CALGB/SWOG 80405 trial showed that all RAS wild-type left-sided primary tumors (splenic flexure to rectum) had longer OS with 6 month improvement if treated with cetuximab than if treated with bevacizumab, while OS is shorter in the right-sided colonic primary group [5]. Genomic and molecular differences between right and left side may result in a significant survival impact when they have been treated with EGFR inhibitors.

Looking to this case, we decided to add cetuximab to his chemotherapy backbone FOLFOX as left side colon cancer. Baseline CT showed irregular exophytic mass in the descending colon in right side. However, the mass was located at 19 cm from anal verge. This patient showed excellent response to treatment combination. To our knowledge this is the first case of un-resectable stage IV colon cancer in SIT where the use of Anti-EGFR with chemotherapy showed significant effect.

We would like to highlight the importance of colonoscopy findings for mass localization and tumor sidedness to decide the optimum therapy in a case of SIT with colon cancer (Figure 1).

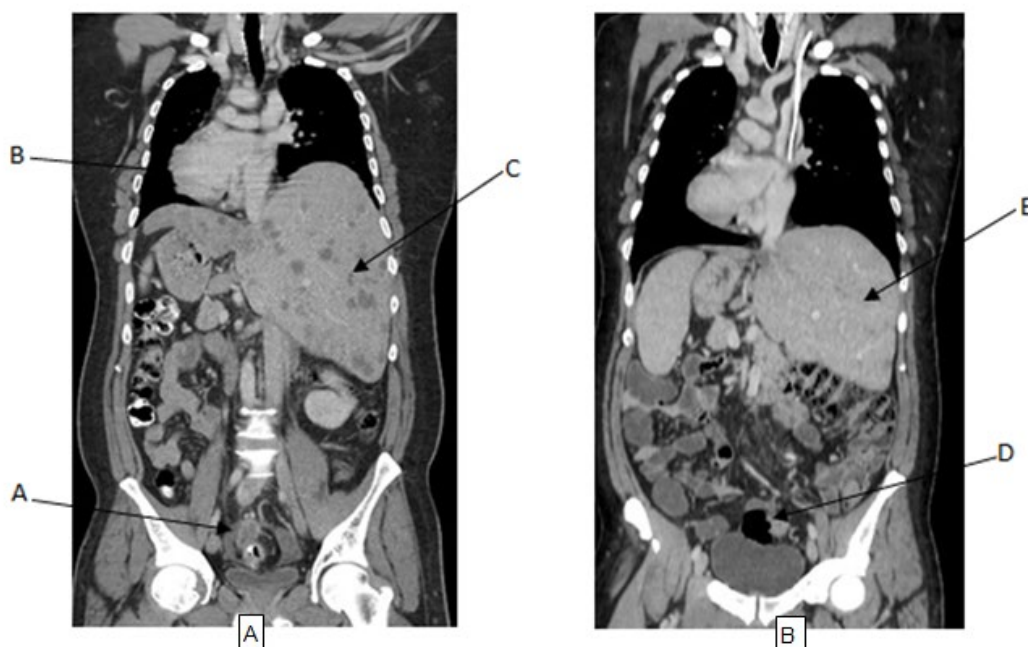


Figure 1: (A) Coronal reformatted image pre-treatment demonstrate exophytic sigmoid colon mass (arrow A), dextrocardia (arrow B) and liver is located in the upper abdomen contains multiple metastatic lesions (arrow C). (B) Coronal reformatted image post-treatment demonstrates left side port-A-cath, significant reduction of sigmoid colon mass (arrow D) and reduction in size of multiple hepatic metastatic lesion denoting good disease response (arrow E).

CONCLUSION

We report a rare case of advanced colon cancer in a patient with Situs Inversus Totalis (SIT). Despite the complexity of the condition, the patient responded well to a combination of chemotherapy and Cetuximab, highlighting the importance of tailored treatment strategies. This case underscores the significance of thorough evaluations, including colonoscopy, in guiding treatment decisions for patients with SIT-associated malignancies. Further research is needed to better understand optimal therapeutic approaches in such cases.

AUTHOR CONTRIBUTIONS

All authors shared the idea, contributed significantly and approved final draft of the manuscript

DISCLOSURE

The authors have none to disclose.

CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest to declare.

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SUPPLEMENTARY MATERIAL

None.

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