

Heterotaxy and tumor of the rectum - operative difficulty (a case report)

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Abstract: *Heterotaxy (From Grec “ heteros “ meaning different, and “ taxos ” meaning positioning), or lateralization defect are two terms used to designate any anomaly in the positioning of one or more thoraco-abdominal organs along the right/left axis . The association of rectal cancer and situs ambiguous is particularly rare. We report the case of a 59 year old female patient with no previous clinical history, suffering from a rectal adenocarcinoma whose CT scan (initially performed for the extension evaluation) showed the situs ambiguous : with a median ectopic spleen under the liver, a short pancreas and a horseshoe-shaped kidney. Candidate for surgical treatment, our concern was to plan her surgical procedure given the difficult anatomical conditions exposing to more intraoperative risks related to the possible anatomical and vascular abnormalities in this patient.*

INTRODUCTION

The association of Heterotaxy and rectal cancer is rarely reported in the literature (6). Through this clinical case, we will discuss the anatomical specificities of this association, as well as the importance of imaging in establishing an accurate mapping of anatomical variations in order to guide surgical treatment.

OBSERVATION

Our 59-year-old patient, with no previous history, Presenting hematochezia evolving for 5 months, the digital rectal exam found a non-stenosing ulcerative mass 6 cm from the anal margin.

Colonoscopy revealed an earlobe-like process at 7 cm from the anal margin that could be crossed and extended over 5 cm. A biopsy was performed and found a moderately differentiated adenocarcinoma.

The TAP scan (thoracic-abdominal-pelvic) showed an irregular thickening of the middle rectum with a median ectopic spleen under the liver, a short pancreas and horseshoe-shaped kidneys. (Img 1-3)

The patient received a fractionated radiochemotherapy CAP50 (50Gy and Capecitabine) After eight weeks, she underwent an anterior resection of the rectum (total resection of the mesorectum) associated to inferior mesenteric lymph nodes dissection followed by a coloanal anastomosis protected by a temporary ileostomy.

The anatomopathological study confirmed a moderately differentiated adenocarcinoma infiltrating the rectum with a respected resection limit, tumor classified as ypT3N0.

Three months later, the patient underwent an ostomy reversal, followed by a clinical and radiological control. Six months following her tumoral resection, the patient had no functional complaints or complications.

DISCUSSION :

The etiologic nature of heterotaxy is unclear(1).

This condition is usually associated with a normal life expectancy unless a gastrointestinal or cardiac abnormality is present (2,3). Situs ambiguus is usually detected by abnormal physical or radiological examinations: vascular abnormalities variation of the celiac trunk, superior mesenteric artery, inferior mesenteric artery (4)....Gastrointestinal system abnormalities (complete common mesentery, polysplenia/asplenia, annular pancreas, short pancreas, diaphragmatic hernia, horseshoe kidneys(5).

Transposition of internal organs is not easy to identify on a physical examination. Palpation of a transverse hepatomegaly can be used as a clue to the diagnosis.

CT scan is the most reliable examination to identify the different digestive and vascular anomalies.

Fourteen cases associating situs ambiguus and adenocarcinoma have been described in the literature. 64% of the cases were women of 63 years old

Sixty percent of the cases had a colonic location, with 21% in the rectum.

Evaluation of gastrointestinal and vascular abnormalities and orientation of internal organs is important. Various imaging techniques should be used to define the abnormalities and determine the appropriate surgical treatment to reduce the difficulty and duration of the procedure so that unsuitable surgical incisions, and complications are avoided. (7,8)

Rectal cancer is a surgical challenge. The goal is to achieve complete tumor removal to minimize local recurrence and prolong survival while preserving quality of life.

Age, preoperative radiotherapy, abdominoperineal amputation surgery and removal outside the anatomical level of the mesorectum are the four identified risk factors for postoperative urinary and sexual complications. In the large retrospective series by Hendren et al. published in 2005, the authors isolated patients (25% of women and 40% of men) for whom surgical treatment was responsible for a deterioration of sexual life (vaginal dryness, dyspareunia, erectile dysfunction...) (9)

Nerve damage, the sympathetic and parasympathetic, may occur with different symptoms. Nerve damage may result from tumor invasion, radiotherapy or surgery.

Ligation of the inferior mesenteric artery (as part of lymph node dissection) at its origin may damage the hypogastric plexus. To avoid this, the section should be performed one centimeter from the origin of the inferior mesenteric artery, avoiding the nerve fibers surrounding the artery.

Sexual and urinary disorders are thought to be caused by trauma to the superior hypogastric plexus or the hypogastric nerves prior the confluence of the parasympathetic fibers in the inferior hypogastric plexus, and may therefore occur regardless of the height of the rectal resection (abdominoperineal amputation [APA], high or low anterior resection) (11,12).

In addition to the factors described above, there are anatomical variations (13, 14) In our patient with horseshoe kidneys, renal vascularisation was provided by two arteries, one main artery coming directly from the aorta and an accessory artery coming from the internal right iliac artery. The isthmus joining the two lower poles was at the L4-L5 disc level, the aorta bifurcates slightly higher than normal and the inferior mesenteric artery was situated at the upper edge of the isthmus, making intraoperative location more difficult.

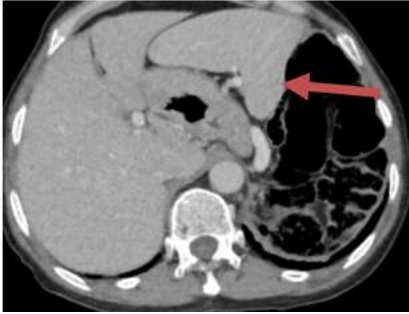
The course of the left ureter was also modified. Its origin was medial and it was taut, forming a curvilinear path with a posterior concavity. Early intraoperative identification was necessary to avoid injury.

Conclusion :

The association of rectal cancer with heterotaxy is rare.

The anatomical variations present must be clearly defined, firstly by imaging and then by intraoperative exploration if surgery is indicated, in order to avoid damage to the abnormally located anatomical structures.

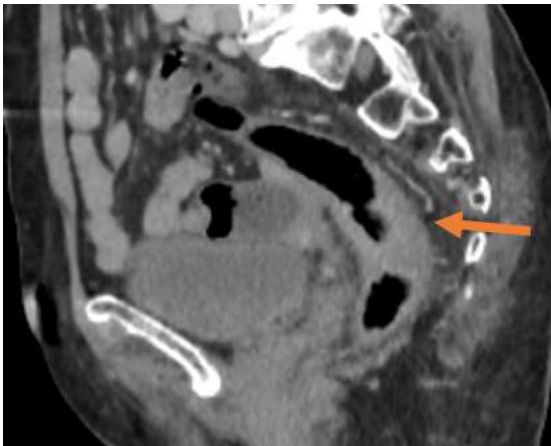
figures:



"Img. 1: Injected CT scan showing median subhepatic ectopic spleen. "



"Img.2: Injected CT scan showing a horseshoe kidney"



"Img.3 Injected CT scan showing Circumferential, asymmetric, irregular and short digestive parietal thickening of the middle rectum".

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13. Laparoscopic abdominal perineal rectal resection for rectal cancer with a horseshoe kidney using preoperative 3D-CT angiography: a case report Jun-ichi Yoshizawa^{1,2*}, Kuniyuki Gomi¹, Arano Makino¹, Ryo Hisamune¹, Sinsuke Sugeno¹, Kou Shimada¹, Kiyotomi Maruyama¹, Motohiro Mihara¹ and Shoji Kajikawa¹.
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