

Management of Ileocecal Valve Adenocarcinoma Presenting as Intestinal Obstruction: Report of A Case and Literature Review

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1. Abstract

1.1. Introduction

Ileocecal valve adenocarcinoma is a rare cause of colorectal cancer arising at the transitional zone where ileal and colonic mucosa merge. Diagnosis of this cancer type is difficult since the patients are usually asymptomatic until complications occur.

1.2. Case Report

In our surgical department, we encountered a case of a 55-year-old female patient who presented with lower abdominal pain and signs of partial bowel obstruction. Further investigation was carried out and revealed metastatic colorectal cancer due to ileocecal valve mass. A laparoscopic right hemicolectomy was performed, and the patient was quickly mobilized and discharged on the fourth post-operative day without any complications.

Discussion: The ileocecal junction has a high susceptibility to carcinogenesis due to the distinct features of the mucosa between the terminal ileum and colon. Nonetheless, it is a scarce cancer type with nonstandard clinical presentation until complications appear. A thorough investigation is necessary to identify the cause and implement an effective treatment plan.

1.3. Conclusion

The diagnosis and management of ileocecal valve tumor is demanding, commonly due to the delayed presentation. Patients usually present with signs and symptoms of bowel obstruction, and proper investigation is needed to identify the cause. Laparoscopic surgery can also be a choice if the patient is hemodynamically stable and there are no signs of peritonitis rather than emergency laparotomy.

2. Introduction

Transition zones are distinct anatomical sites in the body where two different types of epithelia meet, resulting in unique functional properties [1]. Several of these regions are linked to an increased risk of cancer susceptibility. However, the underlying pathophysiological mechanism is not clearly understood. Some of these studied regions include the endocervix, cornea-conjunctiva, and gastro-oesophageal junctions. In addition to the above, the ileocecal junction (ICJ) is an example of a transitional zone and is an exceptionally

rare site for the development of primary gastrointestinal malignancies, with only a limited number of cases reported in the literature to date [2,3]. ICJ is an epithelial transition zone between the terminal ileum and the ascending colon. Although both sites are lined by glandular epithelia, their morphology and function differ significantly. In the terminal ileum, several Paneth cells are present, and lymphoid tissue is more prominent, in contrast to the colonic mucosa. Additionally, the luminal environment varies, as the mucosa in the terminal ileum receives large amounts of bile acids, whereas the mucosa in the ascending colon is exposed to a different set of bacteria due to the distinct colonic flora. All of the above increase the risk of cancer development at the ICJ [2]. These tumors present with non-specific clinical features such as lower abdominal discomfort and, in some cases, may be accompanied by weight loss and changes in bowel habits. As a result, ICV tumors are often diagnosed only incidentally or when complications such as bowel obstruction or bleeding arise, contributing to delayed detection. Some predisposing factors include female gender, inflammatory bowel disease (IBD), and familial adenomatous polyposis (FAP). A significant challenge in understanding ICV carcinogenesis stems from the lack of distinct classification in most colorectal cancer studies. Historically, tumors arising in the cecum, ascending colon, and ICV have been collectively grouped under the term ileocecal tumors, thereby obscuring the true incidence, pathogenesis, and clinical behavior of ICV-specific carcinomas. This article presents a rare case of metastatic ICV adenocarcinoma in a relatively young patient who was asymptomatic and presented with signs and symptoms suggestive of partial bowel obstruction. A comprehensive investigation was conducted using laboratory and imaging tests. Despite the presence of ileus, the patient's nutritional needs were met to achieve an anabolic state prior to surgery. The patient underwent a laparoscopic right hemicolectomy seven days after admission without any complications and was discharged early.

3. Case Summary

3.1. Case Presentation

A 55-year-old, non-smoker female patient presented to the ER with progressive lower abdominal pain, bloating, and constipation for five days. There was no reported nausea or vomiting, and they denied any fever or weight loss.

3.2. Clinical Examination and Medical History

The patient was hemodynamically stable, with a blood pressure of 130/85 mmHg, a heart rate of 92 bpm, a respiratory rate of 18 breaths per minute, oxygen saturation of 98% in room air, and afebrile. Upon clinical examination, the abdomen was distended, with mild tenderness in the lower quadrants, but no signs of peritonitis were observed. Bowel sounds were present, and a digital rectal examination did not reveal any palpable masses or gross blood. The patient's past medical and surgical history was clear, and no medications were being taken. Penicillin allergy was reported. The patient had not performed any imaging studies in the past, neither colonoscopy nor gastroscopy.

3.3. Blood Tests and Imaging Studies

Laboratory tests revealed no leukocytosis, with C-reactive protein and electrolyte levels within the normal range. An abdominal X-ray demonstrated dilated small bowel loops without air-fluid levels. An abdominal contrast-enhanced CT scan was then performed, and a 3.8 cm solid mass was identified at the ileocecal valve (ICV). It was extending 7-8 cm into the terminal ileum, causing partial bowel obstruction (Figure 1a). A transition point was present, and enlarged mesenteric lymph nodes (0.5 cm) were also noted near the ICV. Multiple small hepatic lesions were identified in the CT scan. The patient was admitted to the surgical ward for monitoring and further investigation.

3.4. Management

The patient was admitted for bowel rest, intravenous fluids, and further evaluation. Over 48 hours, symptoms were improved with conservative management. A clear-fluid diet was initiated 48 hours after admission without causing any intolerance, and the patient was able to pass flatus. Solid food was avoided in order not to induce complete bowel obstruction. Additionally, high-protein oral nutritional supplements were added to the patient's diet to support their nutritional needs. A colonoscopy was also conducted six days after admission without any bowel preparation in order to avoid causing complete bowel obstruction. The purpose of colonoscopy was to confirm the presence of solid mass at the ICV, obtain biopsies, and exclude any synchronous colorectal pathology. However, due to the lack of preparation, the colonoscope reached up to 70 cm from the anal verge with no pathological findings. Regarding imaging studies, a chest CT scan was also performed as part of the staging protocol (Figure 1). Finally, abdominal magnetic resonance imaging (MRI) was performed to investigate the hepatic lesions further. As it is demonstrated, the abdominal MRI confirmed that the small hepatic lesions were suggestive of secondary metastases (Figure 1b). A laparoscopy was performed seven days after admission in order to identify the location of the lesion and remove the cause of bowel obstruction-pre-operative patient optimization followed with a high-protein diet, light bowel preparation, and respiratory physiotherapy. During laparoscopy, a solid and firm mass was identified at the ICV. Therefore, a laparoscopic right hemicolectomy was performed following oncological surgical protocol. The resected specimen (Figure 2) was sent for pathological examination and further investigation. The post-operative recovery was uneventful, with the nasogastric tube and urine catheter removed intra-operatively, and no drainage tube was placed. The patient returned to a regular

diet on the third day and was discharged on the fourth post-operative day with instructions for further oncology follow-up. The histological examination revealed an ulcerative, mucus-producing adenocarcinoma of the ICV, measuring 2x1.5x1.5 cm with full-thickness infiltration. Perilymphatic nodes (7/20), perineural (PnI) and vascular (LVI: 4) invasion, and pericolic fat tumor deposits were also noted (Figure 3). According to the TNM/AJCC 2017 staging system, a T4aN2bM1 stage was reported.

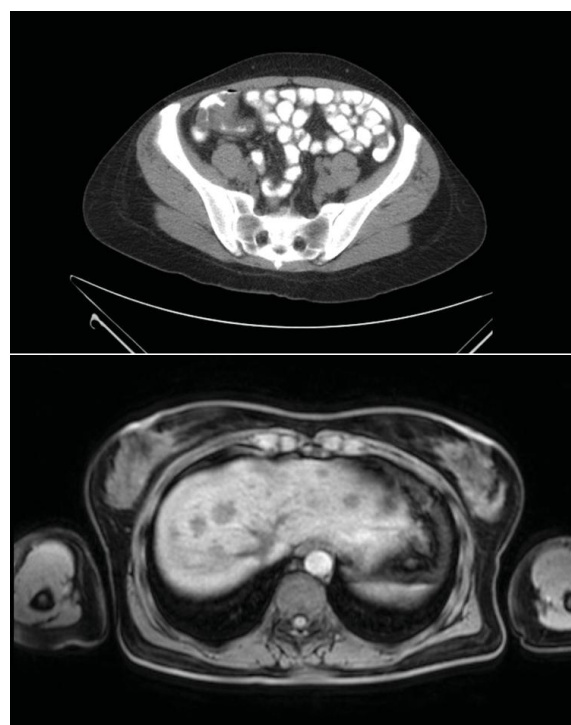


Figure 1a,b: Contrast-enhanced CT abdomen/pelvis showing a solid mass at the ileocecal valve, extending into the terminal ileum b) Abdominal MRI revealing multiple hepatic metastatic lesions.



Figure 2: Resected specimen of the laparoscopic right hemicolectomy.

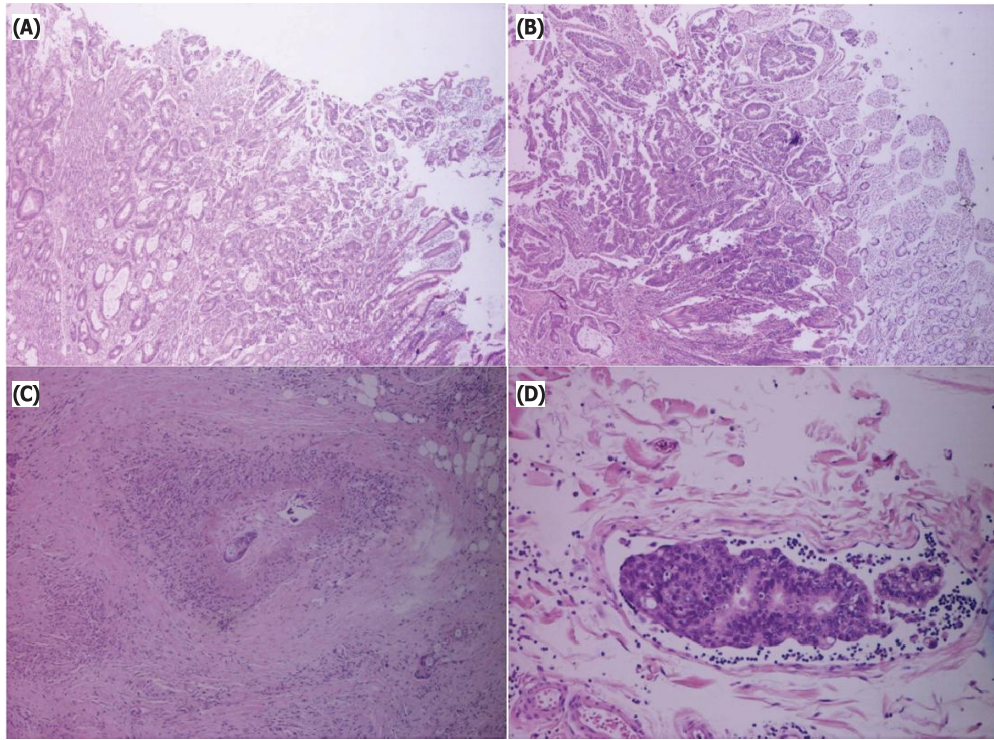


Figure 3a,b,c,d: Histological findings of resected specimen, a)Adenocarcinomaextendingtothe cecum (HEX40), b) Adenocarcinoma extending to terminal ileum (HEX40), c) Lymphovascular emboli in the subserosa (HEX200), d) Neoplastic emboli in a small arterial branch of the subserosa (HEX100).

4. Review of The Literature

4.1. Epidemiology

Demographics analysis reveals that ileocecal junction carcinomas (ICJ-CAs) present more frequently in female patients (56.3%) compared to left-sided (42.9%) and rectal (37.9%) carcinomas. Furthermore, ICJ-CAs are more prevalent in elderly populations, with a mean age of 71.8 ± 12.7 years at diagnosis– higher than that noticed in patients with appendiceal, left-sided, or rectal cancers. These findings suggest that ICJ-CAs may exhibit a distinct clinical and biological subtype of colorectal malignancies. ICJ-CAs are associated with an increased risk in individuals with predisposing conditions such as IBD, FAP, and hereditary nonpolyposis colorectal cancer (HNPCC).[2]

4.2. Clinical Presentation

The clinical presentation of ICV carcinomas ranges from asymptomatic to acute surgical emergencies. In the early stages, patients may exhibit nonspecific symptoms, such as fatigue and generalized weakness, resulting from iron deficiency anemia caused by chronic gastrointestinal blood loss. In more advanced cases, symptoms become more localized and suggestive of gastrointestinal compromise. These may include abdominal pain, nausea, and changes in bowel habits. A firm, palpable mass in the right lower quadrant is a common finding on physical examination in patients with significant tumor burden. In acute cases, patients may present with signs and symptoms of complications, such as bowel obstruction caused by tumor-induced luminal narrowing of the ICV. More specifically, abdominal distension, tinkling bowel sounds accompanied by lower abdominal pain, tenderness, nausea, vomiting, and inability to pass flatus are signs of bowel obstruction. Signs of peritonitis due to bowel perforation can

also be presented on rare occasions. [3,4]

4.3. Diagnosis

Diagnosis of ICJ-CAs depends on the clinical presentation and can differ between acute and non-acute cases, as described above. Under all circumstances, physical examination and laboratory tests are required. Laboratory values may be within the normal range but most commonly include leukocytosis, C-reactive protein elevation, and electrolyte disturbances, accompanied by increased lactate levels, which is likely due to bowel obstruction and ischemia. Additionally, iron deficiency anemia may present due to blood loss. In acute cases, such as those presenting with signs of intestinal obstruction or peritonitis, imaging studies are also conducted in order to identify the cause. Abdominal and chest X-rays are the first modalities that are usually performed, which may reveal air-fluid levels suggestive of ileus or subdiaphragmatic free air indicative of gastrointestinal perforation. A computed Tomography scan of the abdomen is a vital diagnostic tool in such scenarios, offering detailed visualization of the gastrointestinal tract, mass lesions, or complications like perforation and bowel obstruction. In specific scenarios, diagnosis is made intraoperatively during laparoscopic or open exploratory surgery. In chronic or subclinical presentations, particularly in patients with undetermined iron deficiency anemia, colonoscopy is the diagnostic tool of choice. It allows direct visualization of the ileocecal region, biopsy of suspicious lesions, and assessment of the extent of mucosal involvement [4].

4.4. Anatomy, Histopathology and Grading

The ICV is an anatomical structure made up of two mucosal folds–the upper and lower lips– located at the junction of the ileum and the cecum. The upper lip is typically longer than

the lower, with a mean diameter of 2.5 to 3.5cm, with mucosal protrusions into the cecal lumen measuring approximately 0.6cm. Histologically, the ICJ serves as a transitional zone where the confluence of ileal and colonic mucosa is mainly located at the tip of the valve flap. Similar to other epithelial transitional zones, such as the gastroesophageal and anorectal junctions, this region exhibits increased susceptibility to malignant transformation. Primary neoplasms of the ICV include adenocarcinoma (most prevalent), lymphoma, melanoma, sarcoma, neuroendocrine tumors, and granular cell tumors. There are reports of metastatic involvement from breast and gallbladder cancers as well. Ileocecal carcinomas (ICJ-CAs) are typically characterized by a higher frequency of advanced nodal disease (N-stage), a greater proportion of T4 tumors, and an increased prevalence of mucinous, signet-ring cell, and neuroendocrine characteristics, which may imply a higher incidence of microsatellite instability (MSI). Moreover, these tumors are not typically related to sessile-serrated adenomas and more frequently demonstrate low histological grade compared to right-sided colonic carcinomas (79.9% vs. 77.1%). While the American Joint Committee on Cancer (AJCC) TNM system remains the standard for staging [5], Virgilio et al. have proposed a tumor classification based on anatomical proximity to ICV: Type I (within the valve), Type II (1-5mm from the valve), Type III (6-10mm), Type IV (1.1-5cm), Type V (greater than 5cm, involving the ascending colon). This system provides more detailed information about tumor location, which may assist in surgical decision-making and prognostic evaluation [2,6].

4.5. Surgical Treatment

Surgical resection with right hemicolectomy is the first-line treatment for ICV carcinoma, performed by established oncologic principles, including en bloc resection with adequate margins and regional lymphadenectomy to ensure accurate pathological staging and minimize the risk of recurrence. For early-stage malignancies and benign lesions limited to the mucosa or superficial submucosa, endoscopic techniques such as endoscopic mucosal resection (EMR) and endoscopic submucosal dissection (ESD) offer effective, minimally invasive alternatives, particularly in selected patients with favorable characteristics or high surgical risk. In the case of primary lymphomas involving the ICV, most commonly non-Hodgkin B-cell lymphomas, cytotoxic chemotherapy remains the cornerstone of treatment. Surgical intervention is typically reserved for diagnostic purposes or in cases of complications such as obstruction, bleeding, or perforation. The most widely used chemotherapeutic regimen is CHOP therapy, which consists of Cyclophosphamide, Doxorubicin, Vincristine, and Prednisone. This combination is often used in conjunction with rituximab (R-CHOP) for CD20 (+) B-cell lymphomas and has demonstrated high response rates and improved survival outcomes, particularly in intermediate- and high-grade lymphomas. Neuroendocrine tumors (NETs) of the ICV are managed based on tumor grade and extent. Surgical resection remains the primary approach, particularly for localized tumors. High-grade NETs are typically treated with systemic platinum-based chemotherapy, while well-differentiated or metastatic NETs may benefit from somatostatin analogs, liver-directed therapies, or peptide receptor radionuclide therapy (PRRT), depending on the expression of somatostatin receptors and the tumor burden [7-9].

4.6. Prognosis

Interestingly, when defined broadly as tumors with their epicenter within 5cm of the ileocecal valve, ICJ carcinomas do not exhibit a significantly different overall or disease-specific survival compared to other right-sided colon cancers. However, applying a narrower definition—limiting ICJ-CAs to those that directly involve ICV—reveals a prognostic difference. Specifically, patients with these more strictly defined tumors tend to experience poorer outcomes during the initial two years after diagnosis, indicating a potentially more aggressive clinical course in cases with direct ICV involvement [2].

5. Discussion

Ileocecal junction carcinoma is a distinct anatomical site with unique structural and functional features resulting from the differences in the mucosa between the terminal ileum and the ascending colon, which makes it susceptible to carcinogenesis. ICV-CA is a rare entity with a higher prevalence in older female patients. The clinical manifestations of people with ICV-CA are non-specific, and the majority remain asymptomatic until complications arise. Patients may experience lower abdominal pain, fatigue, weakness, weight loss, and changes in bowel habits. However, when complications arise, such as obstruction, symptoms become more prominent, and patients usually seek medical advice. In acute cases, patients usually experience lower abdominal pain, tenderness, nausea, vomiting, and inability to pass stool or flatus. As can be inferred from the above, diagnosis is challenging, especially in non-acute settings. In acute cases, when signs and symptoms of bowel obstruction arise, several laboratory and imaging tests should be conducted in order to identify the cause of ileus. X-ray and CT scans are of vital importance in order to confirm the presence of ileus and its most likely cause. Colonoscopy is typically performed in non-acute cases, particularly when blood is present in the stool or an iron deficiency anemia is suspected. However, in this patient, an endoscopy was also conducted without bowel preparation to avoid causing complete bowel obstruction in order to exclude synchronous malignancy, hence altering the management plan. As a result, colonoscopy also has its role in acute cases of partial bowel obstruction. In this case, the patient presented with signs and symptoms of partial bowel obstruction; however, a clear fluid diet and protein-rich supplements were given 48 hours after admission in order to keep the patient in an anabolic status. The patient was investigated correctly to identify the most likely cause of partial ileus prior to surgery. As can be derived from the above, patients with incomplete bowel obstruction due to a narrowing cause at the region of ICV should be carefully investigated and try to meet their nutritional needs prior to the surgery. Finally, laparoscopic surgery should also be an option in cases where the patient is hemodynamically stable and there are no signs of peritonitis instead of emergency laparotomy since it is associated with fewer post-operative complications, earlier mobilization, and less need for analgesia.

6. Conclusion

Diagnosis of ICV cancer is commonly challenging due to the asymptomatic nature of the disease and the late presentation when complications arise. Patients usually present with signs and symptoms of bowel obstruction due to narrowing of the bowel lumen. Imaging modalities are of vital importance in acute cases in order to identify the cause of obstruction.

Patients who are hemodynamically stable without signs of peritonitis should be carefully investigated prior to surgery, and a colonoscopy can also be performed with caution. It is of vital importance to support the patient's nutritional needs to reduce the risk of post-operative complications. Finally, patients with partial ileus due to ICV-CA may undergo laparoscopy in order to confirm the presence of mass at the ICV region. In cases where the surgeon is experienced enough, a laparoscopic right hemicolectomy can be performed, as it is associated with fewer post-operative complications compared to the open surgical method.

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