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Small Bowel Adenocarcinoma in Celiac Disease

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1. Abstract

Celiac disease is defined as a chronic immune disorder triggered by gluten ingestion. It results in damage of intestinal lining and causes diarrhea, fatigue, weight loss, bloating and anemia. It is estimated to affect 1 in 100 people worldwide (9) and incidence of small bowel carcinoma (SBA) is extremely rare (5). It can occur in a sporadic form or can be associated with a number of predisposing conditions such as hereditary syndromes and immune-mediated intestinal disorders, e.g., celiac disease (CD). However, the features of SBA in the context of CD remain not well understood. Herein we are reporting a case of CD with small bowel adenocarcinoma 41- year-old female patient whom diagnosed with celiac disease for 2 years. Followed over this period with the gastroenterology with progressive symptoms of abdominal pain, nausea and vomiting. Investigations showed segment of bowel with the mass and elements of partial bowel obstruction. The biopsy revealed invasive adenocarcinoma which she underwent surgical resection.

2. Introduction

Celiac disease is defined as a chronic immune disorder triggered by gluten ingestion. It results in damage to the intestinal lining and causes diarrhea, fatigue, weight loss, bloating and anemia. It is estimated to affect 1 in 100 people worldwide (3) and overall small bowel adenocarcinoma is very rare with an incidence of 5.7 cases per million people. [1] Incidence of Small bowel adenocarcinoma (SBA) is extremely rare [1]. It can occur in a sporadic form or can be associated with several predisposing conditions such as hereditary syndromes and immune- mediated intestinal disorders, e.g., celiac disease (CD). [2] Small bowel adenocarcinoma (SBA) accounts for around 40% of all cancers of the small bowel [1]. Small-intestinal adenocarcinoma is diagnosed before surgery in only about 50% of cases and often occurs in conjunction with small bowel obstruction [3] However, the features of SBA in the context of CD remain not well understood. The association between CD and SBA was first described more than 40 years ago and, in contrast to the general population, CD patients have a markedly increased risk of developing SBA [1]. The duodenum is the most common localization for this neoplasia accounting for 55–82% of cases, followed by jejunum (11–25%) and ileum (7–17%).(1) There is an increased risk of small bowel adenocarcinoma in patients with coeliac disease compared with the normal population.(14)

3. Case Presentation

We are reporting a case of celiac disease (CD) with small bowel adenocarcinoma in a 41-year- old female who is diagnosed with celiac disease 2 years ago, presented to the gastroenterology clinic with a history of nausea, vomiting, early satiety and abdominal distention. Work up including Computed Tomography showed dilated segment loop of jejunum with a maximum diameter of 5.4 cm secondary to a short segment of 1.4 cm stricture that is 22cm from duodenojejunal junction. Upper endoscopy showed duodenum bulb fissuring and at 45 cm from pylorus, circumferential friable mass causing luminal narrowing which was biopsied (Figure 1). The histopathology report showed invasive adenocarcinoma. Accordingly, the patient was referred to general surgery clinic. On examination, the patient looked well, not dehydrated, not cachectic. Vital signs are all within normal range. Abdomen was mildly distended with no tenderness. Initial laboratory investigations revealed a white blood cell count (WBC) of 4.5/uL (4000-11000 uL), hemoglobin 12.3 g/dL (10-15 g/dL), and carbohydrate antigen 19.9 was 4 KU/L (0-34 KU/L) And carcinoembryonic antigen CEA was 0.9 (ng/mL). CT of the abdomen and pelvis with intravenous contrast revealed 3.5 CM focal stricture at the level of the proximal jejunum with interval reduction in the diameter of the upstream jejunal loop dilation was 5.3, the stricture demonstrates irregular mucosal enhancement with no extraluminal extension or

locoregional lymph nodes enlargement no other bowel stricture, there is no lymphadenopathy, peritoneal disease or ascites (Figure 2). She was admitted under the care of the general surgery ward, and underwent exploratory laparotomy found mass away from D.J 35 cm which was resected using GIA and side to side anastomosis (Figure 3) then drain was inserted near the anastomosis site and nasogastric tube (NGT) was inserted and the abdomen was closed (Figure 3) The patient was transferred to the general surgery ward patient was started on TPN and NGT was removed after upper gastrgraffin study was performed which showed no leak, NGT and drain were removed and the patient started orally, on 5th-day post-op patient was discharged home with good condition patient follow in OPD after 2 weeks tolerated orally and passing bowel motion and her symports were improved abdomen wound healthy and clips were removed histopathology showed invasive poorly differentiated carcinoma consistent with adenocarcinoma and tumor size 1cm in greatest dimension and the tumor invades into muscularis propria with all margins are free of tumor, no lymph vascular invasion and seventeen lymph node present all negative for tumor with pathologic stage (pTNM) pT2,pN0, PMX patient was referred to oncology for further follow-up. They recommend that no need for adjuvant chemotherapy at this stage and they will follow up the patient.

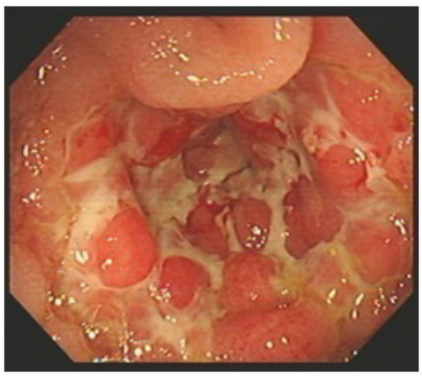

Figure 1. Endoscopic view of upper GI tract showing at 45cm from pylorus, circumferential friable mass causing luminal narrowing. Multiple biopsies were taken.

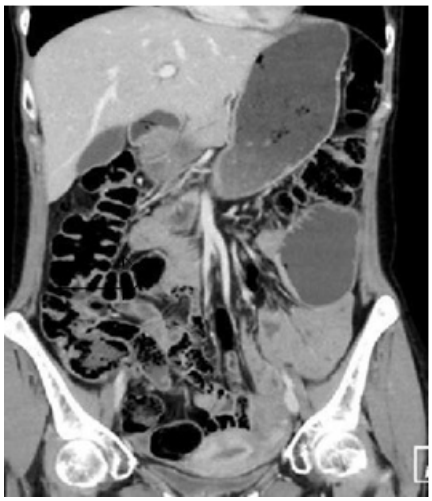

Figure 2. Coronal view of the CT abdomen showing is dilated segmental part of jejunal loop with maximum diameter of 5.4 cm secondary to short segment of 1.4 cm stricture. The strictureis about 22 cm from the dudojeujenal junction.

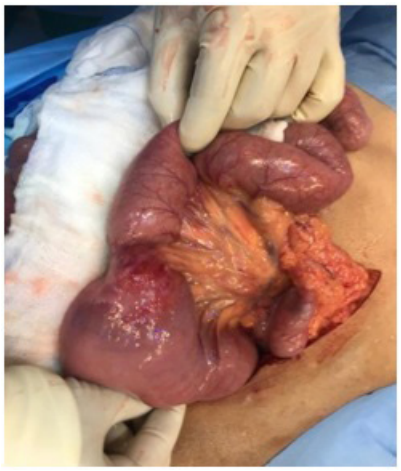

Figure 3. Exploratory laparotomy found mass away from D.J 35 cm which was resected usingGIA and side to side anastomosis.

4. Discussion

Small bowel adenocarcinoma (SBA) is a rare neoplasm, which can occur in a sporadic form or can be associated with several predisposing conditions such as hereditary syndromes and immune-mediated intestinal disorders, e.g., celiac disease (CD).(1) Patients with coeliac disease are at greater risk than the general population for the development of malignant neoplasms, particularly lymphomas.(9) In recent years, a growing number of studies reported a 6- to 9-fold higher incidence of enteropathy-associated T-cell lymphoma and non-Hodgkin neoplasm among CD patients compared with the general population. About gastrointestinal (GI) neoplasms, it was noted that CD patients have a higher risk of developing small bowel adenocarcinoma. (5) In CD, SBC risk has been estimated to be 14-fold higher than that of the general population. (7). Despite significant radiological and endoscopic progress. Still, early diagnosis of small bowel adenocarcinoma remains difficult (2). Accordingly, among all SBC, 13% are associated with a diagnosis of CD. (10) The first case of SBC in CD was described in 1972 and, from that on, many other reports followed up [13] Of nearly 50,000 celiac patients, 64% were diagnosed with some type of cancer since 2000. After a median follow-up of 11.5 years, the incidence of cancer was 6.5 and 5.7 per 1000 person-years in celiac disease patients and controls, respectively. (7) The risk of cancer rose overall, but it was most sharply elevated in the first year after celiac disease diagnosis, and not later on, although the risks of hematologic, lymphoproliferative, hepatobiliary, and pancreatic cancers remained [11]. Patients with celiac disease have a higher risk of developing small intestinal malignancy including adenocarcinoma and lymphoma [12]. Howdle et al. found in their study of 395 cases in the UK that celiac disease was associated with 39% of small bowel lymphomas and 13% of small bowel adenocarcinomas. The results of their study also emphasize the importance of early detection and diagnosis of these cancers in celiac disease patients to improve overall survival [11] Risk levels were highest for people diagnosed with celiac disease after age 60 years of age,

while those diagnosed before age 40 faced no such increase. Lastly, the cancer risk was similar among those diagnosed with celiac disease before or after the year 2000. The team's data showed an overall rise in cancer risk for celiac disease patients, even in recent years. However, the risk increase is only for those diagnosed with celiac disease after age 40, and then mostly within the first year of diagnosis [10]. There are no specific symptoms of these patients with CD from whom have cancer. But certain alarming symptoms may include weight loss, abdominal pain, enlarged lymph nodes, fatigue, and fever. Theoretically, the risk of cancer varies in those with celiac disease; those who have had more intestinal healing appear to be at lower risk than those who continue to have persistent damage. Following a strict gluten-free diet to help promote intestinal healing can lower the risk [10]. The pathophysiology and the mechanism of carcinogenesis in the underlying of celiac disease are not well understood. Hypothetically, chronic inflammation, the release of proinflammatory cytokines, malabsorption of protective nutrients, chronic antigenic stimulation, and impaired immune surveillance have all been proposed as possible contributors [12]. Patients who are older at the time of diagnosis may be at increased risk because the intestine may heal more slowly. The risk of developing cancer may increase if a person with celiac disease has remained undiagnosed for a long period. A metanalysis study involving 79,000 CD patients estimated an odds ratio of 14.41 for developing SBA [9] In another case series study published by Caio et al. in 2019, they concluded that a younger age of onset, a higher prevalence in the female gender, and better overall survival compared to sporadic, Crohn- and hereditary syndrome-related SBA [14-15].

5. Conclusion

Herein we presented a case of a very rare malignancy in the setting of a relatively rare disease. With the best available data from the literature adults with CD have an overall increase in the incidence and the risk for developing intestinal lymphoma and SBC as compared to the general population. Early diagnoses and close observation are the cornerstones to detecting the early conversion to the malignancy.

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