

Celiac Disease Complicated by a Rare, Ulcerative Duodenal Adenocarcinoma

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Abstract Celiac Disease (CD) is an autoimmune condition that is well-known to be associated with increased risk of malignancies, specifically gastro-intestinal neoplasms. We report a case of a young male patient with CD presenting with an ulcerative duodenal adenocarcinoma, successfully operated by cephalic pancreaticoduodenectomy.

Keywords: celiac disease, histopathology, small bowel adenocarcinoma

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1. Introduction

Celiac Disease (CD) is an emerging autoimmune disease and is well known to predispose to various malignancies. There is an important clinical heterogeneity in celiac patients, strengthening the need of screening for such neoplastic complications. [1,2,3]

We report a case of a young male patient with CD presenting with an ulcerative duodenal adenocarcinoma, successfully operated by cephalic pancreatico-duodenectomy.

2. Case Description

A 34 year old male celiac patient presented to the emergencies for acute abdominal (epigastric) pain.

A thorough investigation was ordered: ultrasounds retrieved a small bowel mass while abdominal CT and the abdominal MRi confirmed the duodenum localization of the tumor.

Upper gastrointestinal endoscopy found an ulcerative, semi-obstructive mass of third duodenum, and subsequent biopsies and pathology highlighted an adenocarcinoma.

The patient could benefited a curative pancreaticoduodenectomy with good post-operative results.

3. Discussion

Growing data, with meta-analyses, systematic reviews and population-based studies, confirm that patients with diagnosed CD are at increased risk of malignancy and mortality [4,5].

In fact, people with CD have a high risk of lymphoproliferative malignancies and digestive neoplasms. The peculiar enteropathy-associated T cell lymphoma (EATL) might also occur, particularly in patients with refractory CD [1,6].

Thus, a 2015 meta-analysis found that CD was associated with increased risk of all malignancies as well as gastro-intestinal (GI) malignancies, including esophageal neoplasm and small bowel carcinoma: the pooled Odds Ratio (OR) for risk of all malignancies was 1.25 in CD population, and the OR of GI malignancy was as high as 1.60.

Specifically, patients with CD have a great risk of esophageal cancer (OR = 3.72) and small bowel adenocarcinoma (OR = 14.41) [7].

This latter small bowel adenocarcinoma (SBA) is a rare neoplasm, appearing as a sporadic form or associated with a number of predisposing conditions such as immune-mediated intestinal disorders, e.g CD [8].

Celiac-related SBA cases are characterized by a younger age of onset, a higher prevalence in female gender and a better overall survival compared to sporadic, Crohn- and hereditary syndrome-related SBA [8].

A 2020-published Cohort Study of 347 Patients with newly diagnosed or recurrent SBA at 74 French centres found that the prevalence of a predisposing CD was as high as 1.7% [9].

Among these SBAs, primary carcinoma of the duodenum (like in our patient) is considered as rare: duodenal adenocarcinoma accounts for less than 0.5% of all gastrointestinal cancers and 30-45% of small intestinal cancers [10].

The role of celiac patients follow-up needs more evidence than actually available.

A recently published systematic review of 31 eligible studies found that persisting symptoms were more frequently associated with persisting villous atrophy and this persisting villous atrophy might be a risk factor of lymphomas (although a lot of symptom-free patients had persisting villous atrophy, and many symptomatic patients achieved a mucosal recovery) [11].

Unfortunately, a concomitant celiac serology for this patient (to uncover a causative refractory sprue, for example) could not be obtained.

Imaging is also crucial for such complicated disease. Particularly, magnetic resonance imaging (MRI) is an extremely useful tool for demonstrating duodenal lesions as well as anatomic relationships with adjacent intra-abdominal organs; and is considered as paramount in enabling to establish promptly a correct diagnosis and to plan appropriate surgical procedures [12].

Evidently, surgery is paramount in this condition. Surgical resection in order to achieve clear operative margins is the only available option for cure of this disease; either through pancreaticoduodenectomy or through pancreas-sparing duodenal segmental resection [10].

Our patient benefited a curative surgery by cephalic pancreaticoduodenectomy with good postoperative results.

Finally, in such SBA-resected tumors, the main prognostic factors for overall survival were grade and tumor stage [9].

4. Conclusion

Digestive neoplasms, like small bowel carcinoma, are a rare but real risk in celiac patients.

A long-life gluten free diet and a thorough follow-up are imperative; and a curative surgical approach is highly recommended.

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