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Atypical age of onset of the small bowel adenocarcinoma – case report

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Abstract

Background: Small bowel adenocarcinoma is a rare malignant tumor, with the typical age of diagnosis being 60 years old. In the field of risk factors, we can distinguish, between genetic disorders, inflammatory bowel diseases, smoking, and alcohol abuse. Guidelines for the screening methods are very wide, hence it may be difficult to make the right diagnosis at the early stages of cancer. Additional difficulties can be caused by similarities to inflammatory processes in the gastrointestinal tract.

Case report: We present a case of a 22-year-old male patient, with the symptoms of weight loss, stool retention, abdominal pain, and vomiting. The patient was initially misdiagnosed with inflammatory bowel disease and received the first dose of anti-inflammatory treatment. At that point, the diagnostic process and the workflow of medical care were delayed due to the COVID-19 pandemic. After receiving the x-ray and computed tomography, the obstruction of the ileum was found, and an urgent operation was performed. Tissue samples obtained during surgery revealed the proper diagnosis, a low-differentiated adenocarcinoma (G3) of the small intestine, stage T4Nx.

Conclusion: Taking into consideration the patient's condition, age, and symptoms, we should always think about the probable malignant process. Inflammatory diseases are known for increasing cancer risk and should always be the indication of this disease. The age of onset of the disease is very unusual, however, we must be aware of such cases in our clinical practice.

Keywords: Small bowel adenocarcinoma; ileus; covid-19

Introduction:

Tumors of the small intestine are rare neoplasms of the digestive system, accounting for less than 5% of all gastrointestinal cancers. However, they are characterized by an increase in incidence, probably associated with lifestyle changes in world population and urbanization. The probability of cancer occurrence along the intestine seems to decrease with increasing distance from the stomach, as most tumor cases (55-82%) are in the duodenum, 11-25% in the jejunum and 7-17% in the ileum [1]. Small bowel carcinomas exist in numerous histological variants. The two main histotypes of them are adenocarcinoma, covering around 30% to 40% of cases and neuroendocrine tumors accounting for 35-42%. Other gastrointestinal tumors - sarcomas, and lymphomas - consist of 20-25% of the small intestine cancers [2].

Small bowel adenocarcinoma (SBA) provides a rare problem including less than 3% of gastrointestinal tumors and about 0.5% of all malignant tumors. It localizes mainly in the duodenum reaching about 55% of cases, in the jejunum (29%) and less frequently in the ileum (10%) [3, 4]. This kind of neoplasm may be both primary (more commonly in elderly patients), and metastatic. Metastases are typically from the lung, breasts, and kidneys [5]. The median age at diagnosis is in the sixth decade of life, with the incidence increasing after the age of 40. It affects men with a frequency of 2.6 per 100,000 and women with a frequency of 2.0 per 100,000 [6,7]. Risk factors for SBA include genetic burden, especially inflammatory bowel diseases (IBD), such as Crohn's disease, Lynch syndrome, familial *adenomatous polyposis*, Peutz-Jeghers syndrome and lifestyle factors, like smoking, alcohol abuse, and dietary factors [7]. Strong evidence seems to justify the opinion that small bowel carcinoma is significantly more frequently observed in patients with Crohn's disease, regardless of their age and gender, this background may lead to the occurrence of specific histological types [8]. According to the study by Bojesen et al. based on patients with small intestine carcinoma, two histological types - adenocarcinoma and neuroendocrine tumors - are more frequent in patients with Crohn's disease than in patients with small bowel carcinoma resulting from prior non-inflammatory bowel disease (non-IBD) [9]. Most patients had moderate-to-severe Crohn's disease involving the small intestine and the upper gastrointestinal tract [10]. Contrary to the non-IBD population, SBA localizes more often in the ileum because of the inflammatory culmination in this part of the small intestine. These factors predispose to carcinogenesis. The risk of SBA in those patients arises at younger ages and ranges from 17 to 41 [1]. Another study shows that the cumulative risk of SBA is estimated at 0,2% after 10 years of Crohn's disease and increases to 2.2% after 25 years [1, 11].

Difficulties and delays in diagnosis may often be caused by non-specific symptoms and few guidelines for SBA screening methods [12]. Different diagnostic methods are used to detect the disease, including contrast tests of the small intestine, computer tomography, magnetic

resonance imaging (MRI), and endoscopic methods, including endoscope-body capsule, arteriography, and scintigraphy. In case of genetic and Crohn's diseases, a full small bowel exploration should be done to detect related tumors. In the case of advanced disease, it is also beneficial to employ baseline plasmatic carcinoembryonic antigen (CEA) and carbohydrate antigen (CA) 19.9 as biomarkers with a good prognostic value. [13, 14].

Case report:

A 22-year-old man was admitted to the gastroenterology department due to persistent abdominal pain, gas, and stool retention, as well as vomiting for two days. The patient's body weight decreased by 20 kg in two months (to body mass index – BMI = 19.7), while the first symptoms appeared 5 months before the admission. The patient periodically had abdominal pain and distension, nausea, vomiting and diarrhea in previous months. One month before the admission, the patient fell ill with Covid-19, which delayed the diagnostic process.

In the preliminary examination, inflammatory disease of the digestive system and partial obstruction of the digestive tract were suspected. It should be indicated that the epidemic situation related to Covid-19 harmed the workflow of the department and the availability of imaging and histopathological diagnostics, limiting the availability of tests that should be performed immediately. Laboratory tests performed described hemoglobin (HGB) = 11.3 g/dl (mild anemia), C-Reactive Protein (CRP) = 41 mg/l (far above normal), total protein deficiency in serum, extended Activated Partial Thromboplastin Time (APTT) = 40.5 (upper normal range), increased D-dimers and γ -Glutamyltranspeptidase (GGTP) = 119 (noticeably above normal), deficiencies of folic acid, iron, and potassium in serum. Considering the patient's condition and persistent complaints, the gastroenterological team had decided to introduce the first line of anti-inflammatory treatment, prescribing mesalazine and hydrocortisone, before the results of histopathological and imaging examinations were obtained.

Supportive treatment had also been implemented, involving metoclopramide, pain medications, iron, folic acid, and parenteral nutrition. In the abdominal X-ray review fluid levels (Figure 1) in the umbilical region and short levels in the left hypogastric region (with a feature of subileus) were shown. Computed tomography (CT) scan of the abdomen (Figure 2) described a significant volume of fluid in the peritoneal cavity, a transverse colon with mural thickening, and lumen restriction (about 95mm in length). Small intestine loops with strongly strengthening thickened walls with fluid levels were also described. The lesions were accompanied by widening of the mesenteric veins, strictures of the inferior vena cava compression, and flattening of the portal vein drainage.



Figure 1. The X-ray of the Abdomen with visible fluid levels.

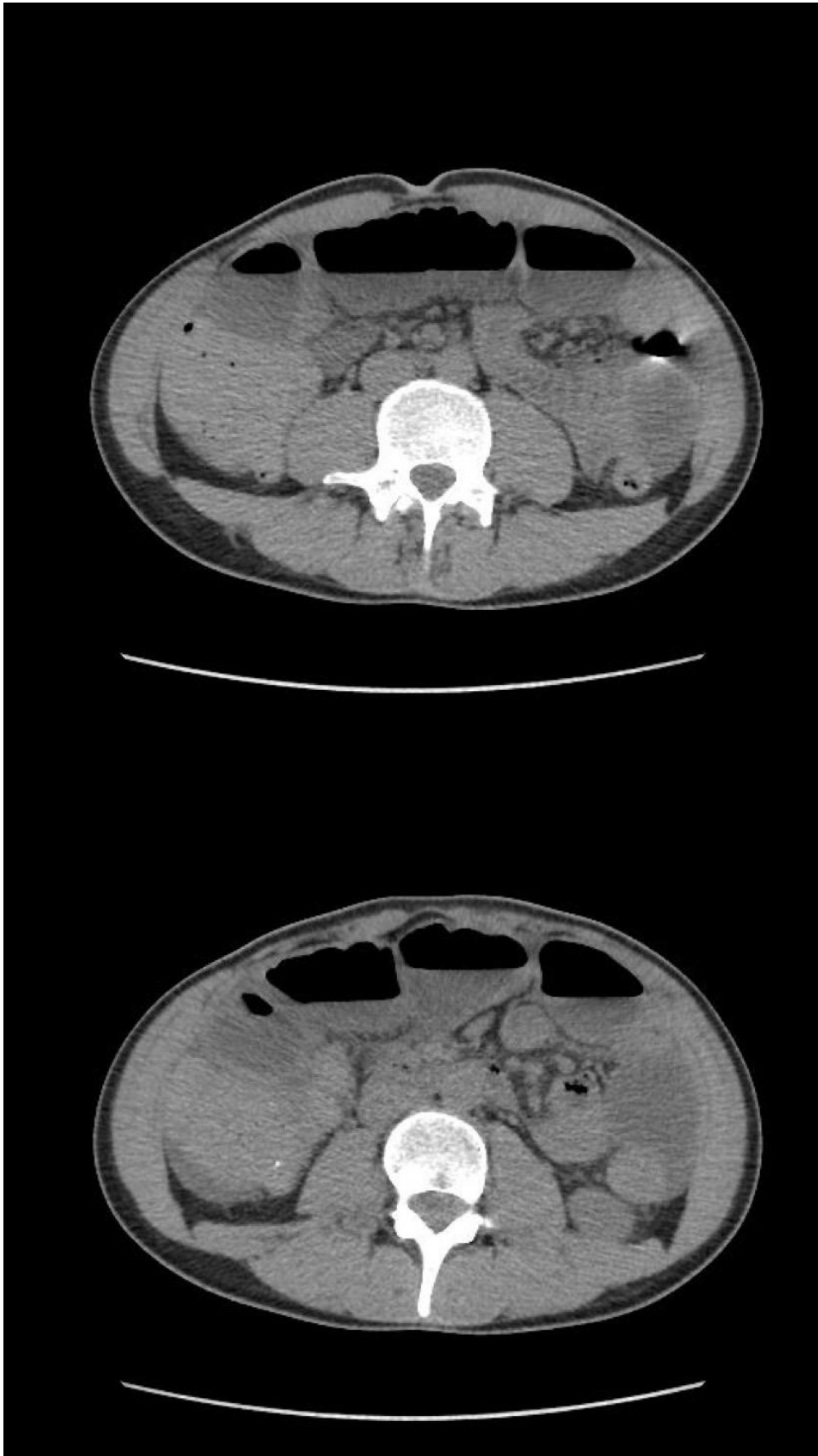


Figure 2 - Computed tomography of the abdomen. Fluid levels and the thickening of the transverse colon are visible.

The presented image justified the assumption that the patient had a persistent intestinal obstruction. Therefore, it was decided to transfer the patient to the Department of General

Surgery and Nutritional Treatment at the Medical University of Lublin, for urgent surgical treatment. Partial resection of the small intestine was performed with 2000 ml of fluid aspirated. A section of the net, a fragment of the small intestine with infiltration, and a fluid were taken for histopathological and bacteriological examination. A low-differentiated adenocarcinoma (G3) of the small intestine was diagnosed (rendered on surgical material), and the pathological stage was described on T4Nx. A multidisciplinary meeting of the therapeutic team was convened to establish a treatment plan for a malignant neoplasm exceeding the limits of the small intestine. It was decided that an oncological treatment would take place after stabilization of the patient's condition due to his general condition, a short time after COVID-19, infection, numerous inflammatory changes in the lungs and intestines, and the constant risk of complete obstruction of the gastrointestinal tract.

Discussion:

Tumors of the small intestine are not common neoplasms. Symptoms seen in patients are not specific and may be confused with infectious or inflammatory symptoms of the gastrointestinal tract. Diagnosis in the early stages of cancer will not always be effective unless a detailed radiological diagnosis is performed. Early diagnosis of small bowel tumors is however a serious diagnostic challenge for both clinicians and radiologists. It has long been noted that small intestine neoplasm often shows overlapping imaging features, however, their imaging features may be seen in specific locations [15]. If possible, based on the radiological image, the probable histotype, indications and probable course of surgery, including the subject to surgical resection, should be assessed. Mesenteric mass with features of calcifications and spiculations may suggest carcinoid tumor, homogeneous mass with thickened, distended walls and lumen dilatation in the ileum suggest lymphoma, while lymphoma, metastases and Gastrointestinal Stromal Tumors can often be described as large masses [15]. Adenocarcinoma is primarily characterized by the presence of a soft-tissue polypoid or constricting annular mass in the duodenum (ulceration is also strongly suggestive of adenocarcinoma). Other common features may be concentric or asymmetric thickening or annular narrowing or ulcerated mass [15, 16, 17]. In the case in question, even the X-ray image, despite the serious risk to the patient, was not specific, as it could be considered typical of inflammatory and obstructive processes, with noticeably high levels of fluid in the serosa cavities. The indication for the procedure was the risk of obstruction, and not the cell mass, indicative of cancer. This may suggest that CT or MRI may be necessary for the diagnosis of suspected small intestine cancer. It should be noted, however, that, as the description of the CT examination results shows, even this test did not provide data enabling the diagnosis of neoplasm. Hence it seems to be highly justified to employ other diagnostic methods including contrast scans, endoscope-body capsule, arteriography, and scintigraphy.

Conclusions:

The presented case proves that the diagnosis can be made late, here not earlier than in such an extreme condition as the gastrointestinal obstruction. It should be noted that the symptoms of a neoplastic disease may coincide with those of inflammatory disease, based on which neoplasm may develop. For this reason, patients with chronic inflammatory diseases of the small intestine, those with Crohn's disease, should - as it seems - be systematically monitored through laboratory tests and appropriately planned diagnostic imaging intervals. Also, a full small bowel exploration in case of genetic burden and Crohn's disease recommended by some guidelines as well as using CA, and CEA as biomarkers may be highly beneficial for early diagnosis of cancer [13, 14]. Nevertheless, particularly disturbing symptoms are (apart from lower abdominal pain) weight loss, especially if it is fast enough to suggest cancer, as well as gastrointestinal bleeding or vomiting. As patients with inflammatory intestine diseases are at higher risk of neoplasm, any diagnosis of an inflammatory small intestine disease should be followed by an assessment of cancer risk.

Further research to develop new effective biomarkers for early diagnosis may be very important to allow the practitioners to rapidly assess risk and further appropriately manage diagnostics and treatment.

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