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Case Report

Mixed Neuroendocrine-Non Neuroendocrine Neoplasms (MiNEN) of the Appendix Presenting with Signs and Symptoms of Small Bowel Obstruction and Gnarly Implants in the Peritoneal Cavity, but no Ascites

Geladari Eleni¹, Tsigani Elena¹, Georgakopoulou Nikolia¹, Papadimitropoulos Vasilios¹, Vallianou Natalia², Dourakis Spyros^{1*}

¹Department of Internal Medicine and Research Laboratory, Medical School, National and Kapodistrian University of Athens, "Hippokration" Hospital, Athens, Greece

²Department of Internal Medicine, Evangelismos General Hospital, Athens, Greece

*Corresponding author: Dourakis Spyros, Professor of Internal Medicine Department and Research Laboratory, Medical School, National and Kapodistrian University of Athens, "Hippokration" Hospital, Athens, Vasilissis Sofias 114, 11527, Greece

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Background

Mixed Neuroendocrine-Non Neuroendocrine Neoplasms (MiNEN) comprise a subcategory of uncommon neoplasms of the digestive tract [1]. They do amalgamate a neuroendocrine and a non-neuroendocrine component and arise in organs that contain neuroendocrine cells; pancreas, appendix, large intestine and to a lesser extent small intestine, where NENs do develop [2].

Objective

Mixed Neuroendocrine-non neuroendocrine neoplasm (MiNEN) of the appendix may have an indolent course that can mislead the clinicians, thus delaying diagnosis and treatment.

Case Report

A 41-year old man presented to the emergency department due to colicky abdominal pain. He also complained for flatulence and episodes of diarrhea alternating with constipation for the past two months. No blood or mucus within the stool was mentioned. On physical examination, his vital signs were within normal limits. Abdominal examination divulged a soft abdomen, mildly-distended and tender only in deep palpation. There was no rebound tenderness, while the bowel sounds were sparse. Abdominal X-ray showed air fluid levels (Figure 1).



Figure 1: Small bowel air fluid levels.

Past medical history included hypertension and a hospitalization for acute appendicitis 4 years ago that was treated conservatively. He referred recent travel to Middle East. A month ago, he visited a private practice clinician, who ordered laboratory and imaging studies. Specifically, the lab results revealed a ferritin level up to 480mg/ml and positive antibodies against Yersinia enterocolitica YopM (2a). Yersinia enterocolitica infection however was ruled out because there were no antibodies against YopD (4a), which need to be present in order to make the diagnosis of yersiniosis. Apart from that, there were no white blood cells

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in the stool. He underwent abdominal magnetic resonance tomography and MRI enteroclysis, which revealed dilated jejunum, accompanied by air-fluid levels and stenosis at the level of the terminal ileum. Endoscopy of the lower gastrointestinal tract showed edematous mucosa and the histology report stated that non-specific inflammatory type of colitis was present. He was placed on empiric treatment with mesalazine, metronidazole and ciprofloxacin with no clinical improvement. He was admitted for further evaluation. Abdominal computed tomography displayed dilatation of the small intestine, blurred mesenterium and reticular non-specific lesions at the right peritoneal cavity with no ascitic fluid (Figure 2). The helices of the small intestine were so deformed that the impression of an internal hernia was given. At that point, exploratory laparotomy was performed, where a dilated and edematous small intestine 75cm before the ileocecal valve was noted along with a large mass residing at the caecum and appendix as well as multiple white implantations at the serosa of the large and small intestine, peritoneum and mesenterium (Figure 3). The histology exhibited MiNEN of the appendix.



Figure 2: Nodular lesions of the peritoneum at the right, dilated small intestine helices, no peritoneal fluid.



Figure 3: Multiple white lesions representing nodular implants at the wall peritoneum of the right lateral groove and mesenterium.

Discussion

This case report is demonstrative of how a usual clinical presentation of small bowel obstruction may unveil an unusual tumor with a rare histological type arising on the appendix. Initially, having the positive antibodies against Yersinia along with the recent travel history made yersiniosis a likely scenario. However, the absence of fever and the negative inflammatory markers along with normal stool microscopy ruled out this diagnosis [3]. Another thought was inflammatory bowel disease, since there were imaging findings consistent with it; areas of bowel dilatation along with stenosis at the ileocecal valve. Nonetheless, colonoscopy did not display any abnormal macroscopic findings and the histology specimen exhibited no specific pathological lesions in favor of IBD. Granular diseases, tuberculosis, sarcoidosis or mesenteric panniculitis/sclerosing mesenteritis were also included in the differential list. However, the PPD, Quantiferon test, SACE and immunological profile came out negative and hence there were not suggestive of the aforementioned diagnostic thoughts [4]. Literally, it was concluded that the diagnosis would be established only by exploratory laparotomy, since this patient's clinical condition was deteriorating. Finally, histology revealed an intermediate grade of MiNEN. It has been acknowledged that appendicular MiNEN (formerly known as "adenocarcinoid tumors") comprise about 10% of all appendicular malignancies [5]. Histology unveiled both adenoma and well-differentiated neuroendocrine components, which accounted for the intermediate or low grade of malignancy. Most appendicular MiNEN are discovered at the systematic pathological examination of resected surgical specimens in patients operated on for an acute appendicitis, although 20-30% are identified incidentally. MiNENs' prognosis is closer to that of adenocarcinoma, depending on the rate of adenocarcinoma component and the presence of signet-ring adenocarcinoma [5]. Systemic chemotherapy with capecitabine plus oxaliplatin was initiated.

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