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CASE REPORT

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Acute enteric eosinophilic myenteric ganglionitis: A challenging and uncommon case of lung adenocarcinoma

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Abstract

Chronic intestinal pseudo-obstruction (CIPO) is caused by metabolic and endocrine factors, but it can also be caused by the cancer's paraneoplastic effect. The cancers most associated with paraneoplastic syndromes are lung cancers and are generally associated with small cell lung cancers. Lung adenocarcinomas may rarely cause paraneoplastic syndromes, however, they mostly cause general effects such as cachexia and leukocytosis. The goal of this case report was to provide a case of non-mechanical obstruction due to lung adenocarcinoma as a very unusual cause of intestinal pseudo-obstruction, together with clinical, laboratory, and imaging results.

Keywords: Chronic intestinal pseudo-obstruction, lung adenocarcinoma, eosinophilic myenteric ganglionitis

Introduction

CIPO (chronic intestinal pseudo-obstruction) is an uncommon condition caused by abnormalities in the underlying Cajal cells, which can be neuropathic, myopathic, or interstitial. CIPO is a serious disorder marked by an inability to move the contents of the intestine normally [1]. Abdominal pain, vomiting, bloating, constipation, and diarrhea are all symptoms of this illness. The ENS neurons are divided into two types of ganglia: myenteric (Auerbach's) and submucosal (Meissner's) plexuses [2]. CIPO can be classified as either primary, secondary, or idiopathic [3]. According to the predominant involvement of interstitial cells, smooth muscle cells, or enteric neurons, primary CIPO can be categorized into three basic types of gastrointestinal neuromuscular disorders (GINMD): mesenchymopathies, myopathies, and neuropathies [1,3]. The enteric neuropathies that cause CIPO can be classified into two groups: (a) degenerative neuropathies with no visible inflammatory response, and (b) inflammatory neuropathies, also known as myenteric ganglionitis [2]. Eosinophilic myenteric ganglionitis is a condition in which eosinophils infiltrate the Auerbach myenteric plexus [2]. It's been linked to bowel dysmotility, and there have been a few reports of intestinal pseudo-obstructive syndrome. These changes could be idiopathic or related to another illness. In around half of the cases, neurological, paraneoplastic, autoimmune, metabolic/ endocrine, and viral illnesses play a significant role in the etiology [1,3,4]. CIPO is realized, most commonly due to small cell lung cancers, carcinoid tumors and paraneoplastic syndrome reported in malignant thymoma [4]. In these patients, serological panel of paraneoplastic syndrome is often positive. Paraneoplastic CIPO usually affects gastrointestinal motility and occurs with signs and symptoms mimicking mechanical bowel obstruction. Most of these

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patients do not respond to medical treatment and the non-functional bowel segment needs to be resected surgically [4]. To the best of our knowledge, we have not encountered CIPO as a paraneoplastic syndrome due to pulmonary adenocarcinoma in the literature. The first patient with eosinophilic myenteric ganglionitis underlying CIPO as a harbinger of paraneoplastic syndrome related to lung cancer is presented in this case report.

Case report

A 63-year-old male patient was admitted to the hospital with complaints of loss of appetite, weight loss, delayed stool and gas passage, inability to defecate for the past 10 days, abdominal pain, and bloating. He was hospitalized for diagnosis, monitoring and treatment. He had not a previous history of any health problems. Radiographic air-fluid levels on the abdominal X-ray, diffuse small bowel dilatations on abdominal CT, and diffuse fecal retention, especially on the right colon, were present. sigmoid colon was found to be very long (dolichocolon) and diffuse thickening of its wall (Figure 1).

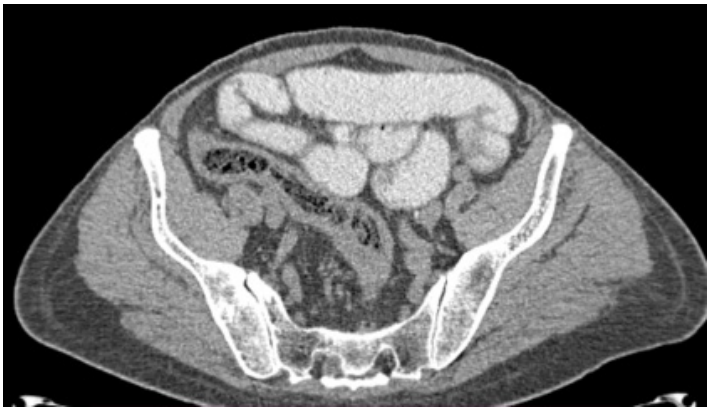

Figure 1. In the December 2017 abdominal CT examination of the patient who presented with symptoms of intestinal obstruction, diffuse wall thickening (white arrow-head) in the sigmoid colon, dolicolone, and dilatation in the intestinal loops (black arrow-head) were detected

However, no tumoral formation was detected to suggest colon cancer. Supportive treatment and antibiotic therapy with broad-spectrum antibiotics were started. Colonoscopy was not performed as the general condition began to deteriorate and surgical resection was performed. Subtotal colectomy with terminal ileectomy were performed by surgeons. Chronic inflammation findings caused by diffusely infiltrating eosinophils, lymphocytes, and plasma cells in the internal and external layers of the muscularis propria were observed during the examination of the patient's surgical specimen, and it was diagnosed as CIPO secondary to mixed type ganglionitis that also affected the deep submucosal and myenteric plexuses (Figure 2). All possible causes were investigated. The patient was checked for paraneoplastic syndromes, infectious causes, endocrine and metabolic causes, malignancies, autoimmune causes by serological and biochemical investigations and using all imaging modalities. All relevant diciplines were consulted. All neuronal antibodies were negative on serological examination. Only Anti-Smooth muscle antibody (ASMA) tested positive in 1/640 titer and steroid treatment was started because the reason could be autoimmune and had been mentioned in the literature. The patient's steroid treatment was reduced and discontinued upon continued complaints. The patient was reevaluated and his thoracic CT scans, which were normal before, developed multiple pathological lymphadenopathies in the mediastinum after the third month (Figure 3). The patient was diagnosed with lung adenocarcinoma on histological examination of the biopsy taken from the lymph nodes and is currently receiving chemotherapy and radiotherapy. Informed consent was obtained from the patient for the endoscopic procedure. Ethics committee approval was not required in this case report.

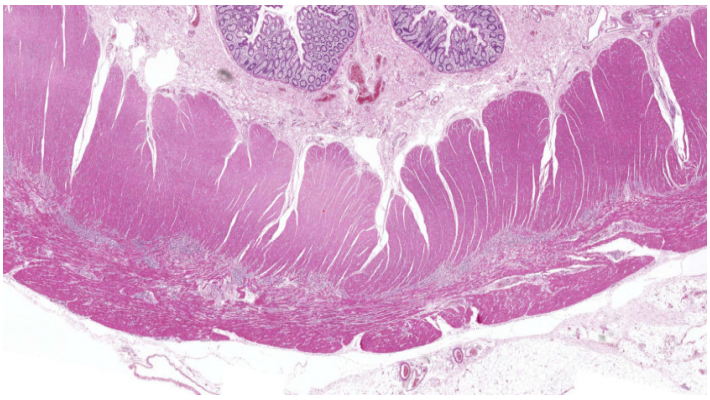

Figure 2. Pathological examination of the surgical specimen reveals diffuse (ileum+colon+appendix vermiformis) mixed ganglionitis. There is inflammation of myenteric and deep submucosal plexuses, consisting of lymphocytes, eosinophils or plasma cells. It was evaluated as CIPO secondary to mixed type ganglionitis that also affects muscularis propria

Figure 2a. Diffuse inflammation of myenteric and deep submucosal plexuses, (H&E, x2)

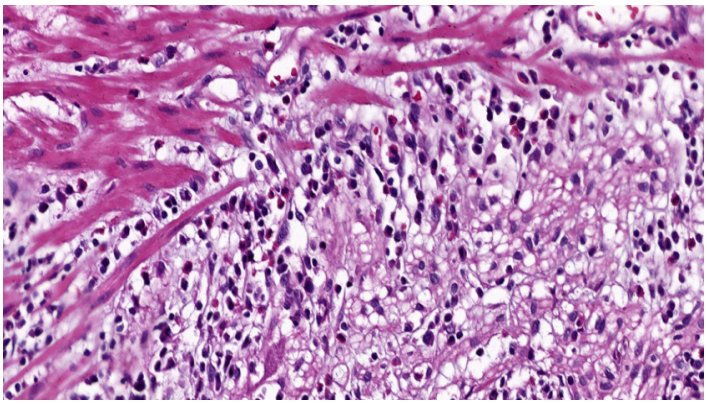

Figure 2b. Myenteric ganglionitis showing severe eosinophilic and lymphocytic infiltration of plexuses, (H&E, x42)

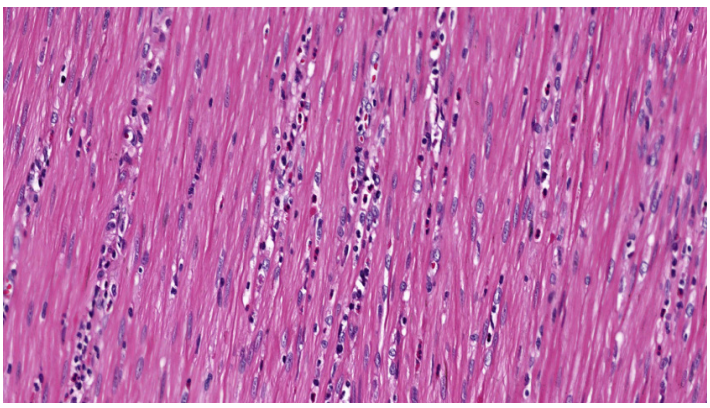

Figure 2c. İnflammatory cell infiltration of muscularis propria, (H&E, x42).

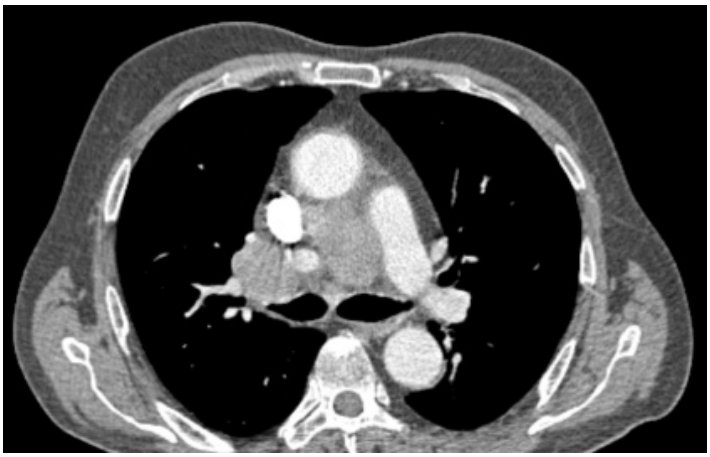

Figure 3. Contrast-enhanced thorax CT examination dated June 2018 revealed mediastinal multiple lymphadenopathies (white arrow-head) in the patient whose previous thorax CT examinations were normal

Discussion

In the absence of any lesion occluding the gut lumen, CIPO is an uncommon and very morbid condition characterized by reduced gastrointestinal propulsion, as well as symptoms and signs of bowel obstruction [5]. CIPO can be categorized into three major types based on histologic examination: neuropathies, mesenchymopathies, and myopathies, depending on whether enteric neurons, ICCs, or smooth muscle cells are involved [5]. The two primary ganglionic plexuses are characterized by a dense infiltrate of lymphocytes and plasma cells, although the myenteric ganglia and associated nerve axons are more frequently damaged (hence the term myenteric ganglionitis) [6]. In pediatric and adult patients with CIPO, non-lymphocytic inflammatory neuropathies such as eosinophilic or mast cell ganglionitis have been observed [3,6]. In 15% of juvenile CIPO patients and roughly 20% of adult CIPO patients, CIPO is a major cause of chronic intestinal failure [7]. While certain cases of CIPO are caused by a variety of known pathological illnesses, such as myxedema, Duchenne muscular dystrophy, hypothyroidism, hypoparathyroidism, celiac disease, Chagas disease, and mitochondrial disorders, the majority of cases have no known cause [7-9].

Inflammatory and degenerative histopathologic abnormalities are present in CIPO linked with intestinal neuropathies. T-lymphocytes infiltrate the enteric neurons in the two ganglionated plexuses of the ENS, causing inflammatory neuropathy. The inflammatory infiltration most usually targets the myenteric plexus, hence the clinicopathologic term "myenteric ganglionitis" [7]. In pediatric and adult patients with CIPO, non-lymphocytic inflammatory neuropathies such as eosinophilic or mast cell ganglionitis have been observed [3,6]. Because non-lymphocytic forms of inflammatory neuropathy are so infrequent, their clinicopathological characteristics are still unknown. In our case; the cause of CIPO was inflammatory neuropathy. Histologic examination of surgical specimen of the patient; eosinophilic myenteric ganglionitis was detected. It was seen mixed inflammatory infiltration in the myenteric plexus which predominate cells are eosinophils. We decided that our patient has myenteric eosinophilic ganglionitis . After his evaulation by clinical , serological and imagine modalities at this time, we didn't demonstrate any etiologic cause of this problem. But later, when lung adenocarcinoma was determined in histologic examination of lymph nodes of the mediastinum, we thought that this clinical picture was depended to this malignancy. Paraneoplastic syndromes are predominantly the precursors of cancer. CIPO is one of these paraneoplastic syndromes. To the best of our knowledge, we have not encountered CIPO as a paraneoplastic syndrome due to pulmonary adenocarcinoma in the literature. In the literature, it is reported that much small cell lung cancer plays a role in etiology. Therefore, we aimed to share this rare and difficult phenomenon.

Patient informed consent

Patients were not required to give their informed consent for inclusion in this retrospective case study, because we used anonymized clinical data and individuals cannot be recognized based on the data available.

Conflict of interests

The authors declare that they have no competing interests.

Financial Disclosure

All authors declare no financial support.

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