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CASE REPORT | COLON

Chronic Intestinal Pseudo-Obstruction Due to Incidentally Found Thymoma

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ABSTRACT

Chronic intestinal pseudo-obstruction is a severe gastrointestinal tract disorder mimicking a bowel obstruction with no mechanical causes. Our patient presented with bowel obstruction symptoms, and a thymoma was incidentally found during the diagnostic workup. After failing conservative management, the patient underwent exploratory laparotomy that showed negative results for a mechanical cause of obstruction. Laboratory workup was suggestive of paraneoplastic syndrome, and the patient was started on steroids and pyridostigmine course with symptom resolution. Thus, in patients with chronic intestinal pseudo-obstruction, paraneoplastic syndrome should be considered in the differential diagnosis.

INTRODUCTION

Chronic intestinal pseudo-obstruction (CIPO) and small bowel obstruction maybe indistinguishable from a clinical perspective. However, CIPO is caused by either myenteric plexus or intestinal smooth muscle dysfunction in the absence of mechanical obstruction. In rare cases, this syndrome can be associated with a paraneoplastic syndrome, such as small cell lung carcinoma. We present a patient with new-onset CIPO related to an incidentally found thymoma.

CASE REPORT

A 43-year-old woman with no significant medical or surgical history presented to the emergency department with a 2-day history of abdominal pain and distension, nausea, and persistent vomiting. She denied having bowel movements or flatus since the onset of symptoms and endorsed worsening constipation for the past 6 months. The patient reported blurred vision and worsening fatigue throughout the day for the same amount of time. She sought help in urgent care 3 days before and was prescribed 10 mg of dicyclomine as needed, without improvement. Vital signs on admission were unremarkable. An initial blood specimen workup was significant for 1.8 mg/dL of magnesium with normal blood cell count, liver enzymes, and electrolytes. An abdominal x-ray demonstrated multiple dilated small bowel loops suggestive of ileus vs small bowel obstruction.

An abdominal and pelvic computed tomography (CT) with intravenous (IV) contrast showed severe small bowel dilation with multiple air–fluid levels, without evidence of a mechanical obstruction (Figure 1). Notably, the upper sections of the CT scan showed a possible mediastinal mass. A thoracic CT chest showed a 7.5 × 3.8 × 9-cm eccentric lobulated homogeneous anterior mediastinal mass without evidence of serosal invasion (Figure 2). Despite conservative management with nasogastric tube and multiple tap water enemas, the patient failed to improve in her clinical and radiological evaluation after 7 days. Subsequently, the patient underwent diagnostic laparoscopy and was found to have diffuse small bowel and colonic dilation; no transition point or mechanical obstruction was observed on careful bowel inspection. Given the concern for paraneoplastic syndrome due to thymoma, the patient was started on 30 mg of empirical pyridostigmine twice a day and 40 mg of methylprednisolone once a day. The next day, the patient had bowel movements and flatus, with improvement in abdominal pain and distension severity. Workup for paraneoplastic syndrome came back with positive results for acetylcholine receptor antibody (AChR) (4.0 nmol/L) and anti-CV2 antibodies (titer 1:800). A subsequent biopsy of the mediastinal mass showed cores of tissue

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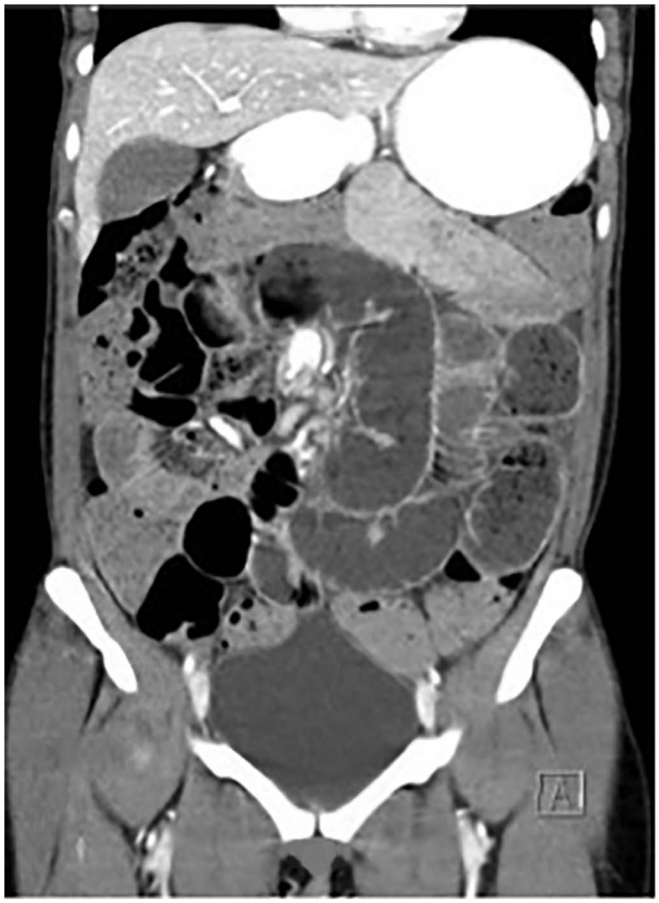

Figure 1. Abdominal computed tomography showing multiple air–fluid levels and small bowel dilation.

with expanded epithelial cell meshworks and rich lymphoid background consistent with a thymoma. The patient ultimately underwent outpatient thymoma excision.

DISCUSSION

Paraneoplastic neurological syndrome is an umbrella term for a group of neurological sequelae secondary to a paraneoplastic phenomenon, with an estimated prevalence of 1 in 10,000 cancer patients. Autoimmune paraneoplastic CIPO (AP-CIPO) is a subset of paraneoplastic neurological syndrome associated with a poor prognosis, with varying mortality depending on the underlying malignancy and response to oncologic therapy (small cell lung carcinoma, thymoma, breast cancer, or B-cell lymphoma), parenteral nutrition, and surgical complications.[1-3]

AP-CIPO arises after the myenteric plexus in the intestinal tract is damaged by antibodies produced by a pathologic interaction between neuronal antigens, malignant cells, and the immune system. The most common antibodies in AP-CIPO are antineuronal nuclear antibody type 1 (anti-Hu), Purkinje cell cytoplasmic type 1 (anti-Yo), and N-type voltage-gated calcium channel antibodies.[2] Other less frequent antibodies in AP-CIPO are type 2 anti-neuronal nuclear antibody (anti-Ri), amphiphysin antibody, PCA-2, and

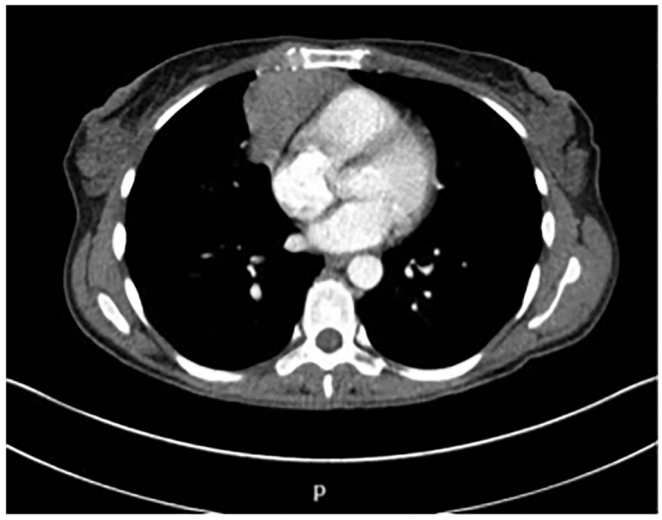

Figure 2. Chest computed tomography with incidental finding of mediastinal mass.

CRMP antibody.[4] Historically, CV2 antibodies were initially associated with encephalomyelitis and peripheral neuropathy.[5] Nevertheless, Paine et al recommended testing for CV2 antibodies in addition to anti-Hu and AChR antibodies in patients with chronic severe dysmotility to rule out an underlying malignancy.[6] In our patient, both CV2 and AChR antibodies were present, supporting an AP-CIPO diagnosis secondary to a thymoma. Interestingly, paraneoplastic syndromes often present before the clinical manifestations of the primary malignancy. For example, the discovery of the primary malignancy in our patient was performed after she presented with intestinal pseudo-obstruction.

Treatment options for AP-CIPO stem from various case reports and case series. A variety of treatment options have been attempted for AP-CIPO, with most of the current recommendations arising from case reports and case series. These treatments include 1 mg/kg of prednisone with subsequent taper, sometimes preceded by a short course of IV methylprednisolone or immunoglobulin.[3] Weinkauf et al highlighted the importance of immunomodulation with rituximab and cyclophosphamide in the treatment of patients with AP-CIPO, secondary to small cell lung cancer.[7] However, this drug combination carries a concern of tumor progression because of the associated immunosuppression.[8] Rakocevic et al reported a patient with thymoma, CIPO, and myasthenia gravis, who was treated with parenteral neostigmine and IV immune globulin for 5 days without improvement and subsequently died because of aspiration pneumonia and septic shock.[9] Musthafa et al also reported a patient with thymoma, AP-CIPO, and myasthenia gravis who responded to oral prednisone and pyridostigmine combination.[10] In addition, surgical options such as limited bowel resection guided by neurophysiological testing have also been used. Malhotra et al successfully treated a patient with thymoma and AP-CIPO with an extended right hemicolectomy when the patient failed to respond to transverse loop colostomy.[11] Our patient was treated with methylprednisolone, pyridostigmine, and a thymectomy, which led to symptom resolution. This approach has been recommended for thymoma-related AP-CIPO because it shares

myasthenic features for which combination therapy improves symptoms at the neuromuscular junction and, thus, the gastrointestinal motility.

Hence, patients with obstructive symptoms without a mechanical cause need to be evaluated for potentially reversible etiologies such as a paraneoplastic syndrome. A comprehensive evaluation can lead to early diagnosis of an occult malignancy. Several challenges exist in the treatment of these patients, with varying degrees of response to pharmacological or surgical treatment. Nevertheless, the main approach may vary based on the primary malignancy and its staging.

DISCLOSURES

Author contributions: D. Castaneda and R. Miret wrote the manuscript and reviewed the literature. R. Rajagopalan wrote and edited the manuscript and reviewed the literature. F. Castro revised the manuscript for intellectual content. D. Castaneda is the article guarantor.

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