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Case Report

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Diagnosis and management of pediatric intestinal pseudo-obstruction: A multidisciplinary approach through a case of perforated appendicitis

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Abstract

Pediatric intestinal pseudo-obstruction (PIPO) is a rare clinical condition characterized by signs of bowel obstruction in the absence of any mechanical narrowing or occlusion of the intestinal lumen, resulting from gastrointestinal motility dysfunction. In approximately 80% of cases, symptoms emerge within the first year of life. The etiology of PIPO may involve genetic predisposition, as well as triggering factors such as infections, surgical procedures, anesthesia, stress, and malnutrition. In this case, PIPO was diagnosed in a patient with prolonged diagnostic delay due to persistent ileus episodes following perforated appendicitis, thanks to a multidisciplinary approach. The patient responded well to neostigmine therapy. In conclusion, this case highlights the late-onset form of PIPO and emphasizes the challenges in the diagnosis and treatment process.

Keywords: Gastrointestinal motility, ileus, neostigmine, pediatric pseudo-obstruction, postoperative complication.

Pediatric intestinal pseudo-obstruction (PIPO) is a rare and heterogeneous disorder characterized by severe gastrointestinal motility impairment, leading to signs of obstruction without any anatomical blockage. The clinical picture typically includes recurrent ileus episodes, abdominal distension, and chronic constipation. While most cases present within the first year of life, late-onset cases have also been reported.[1-3]

The etiology of PIPO is highly complex. Neuromuscular, immunological, and environmental factors contribute to its pathogenesis. Triggers such as infections, fever, general anesthesia, surgical interventions, psychosocial stress, and malnutrition can precipitate symptoms.[4,5] Moreover, genetic mutations have been identified in some cases. In particular, mutations in the Smooth Muscle (ACTG2) gene are frequently observed in hereditary PIPO with autosomal dominant inheritance.[6]

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In this article, we report a pediatric patient who developed recurrent ileus episodes in the postoperative period and was diagnosed with PIPO through a multidisciplinary evaluation and discuss the diagnostic and therapeutic process in the light of literature data.

CASE REPORT

A 14-year-old female patient presented with severe epigastric pain lasting for five days, right lower quadrant tenderness, and recurrent bilious vomiting. Although there was no history of significant gastrointestinal motility disorders, she had occasional constipation. It was also learned that her sibling was followed during the neonatal period for unexplained feeding intolerance. She had no history of prior surgeries. On physical

examination, diffuse abdominal distension, marked tenderness in the right lower quadrant, rebound, and guarding were noted. Laboratory evaluation revealed leukocytosis (22,500/mm^3) and elevated C-reactive protein (148 mg/L). Abdominal upright X-ray showed multiple air-fluid levels (Figure 1), and ultrasound revealed no visualization of the appendix and the presence of free fluid in the abdomen and pelvis. Based on these findings, the patient underwent emergency laparotomy with a preliminary diagnosis of acute abdomen.

Intraoperatively, a perforated appendix was detected, and appendectomy and peritoneal debridement were performed. However, on postoperative day three, persistent abdominal distension, absence of fecal and gas discharge, and continued bilious vomiting suggested adhesive ileus. Despite two subsequent adhesiolysis operations, the clinical course did not improve, and a diverting ileostomy was created. During surgery, rectal and ileostomy segment biopsies were obtained to rule out neuronal intestinal dysplasia (NID) and Hirschsprung disease. Histopathological examination showed the presence of ganglion cells and no abnormal proliferation in the enteric nervous system (Figure 2).

Due to the continued non-functioning ileostomy and persistent air-fluid levels in the postoperative period, a multidisciplinary council was formed for further assessment. Computed tomography (CT) angiography ruled out median arcuate ligament syndrome and superior mesenteric artery syndrome (Figure 3). Pathological, biochemical, and genetic evaluations for scleroderma, hyperimmunoglobulinemia D syndrome, cystic fibrosis, and mitochondrial neurogastrointestinal encephalomyopathy were all normal.

Due to ongoing motility dysfunction, nonfunctional ileostomy, and bilious vomiting, a diagnosis of PIPO was considered. Partial response was observed with parenteral nutrition and domperidone therapy. Intravenous neostigmine was, then, initiated, resulting in activation of the ileostomy and clinical improvement. Oral intake was gradually increased, and the patient transitioned to enteral nutrition. The patient is

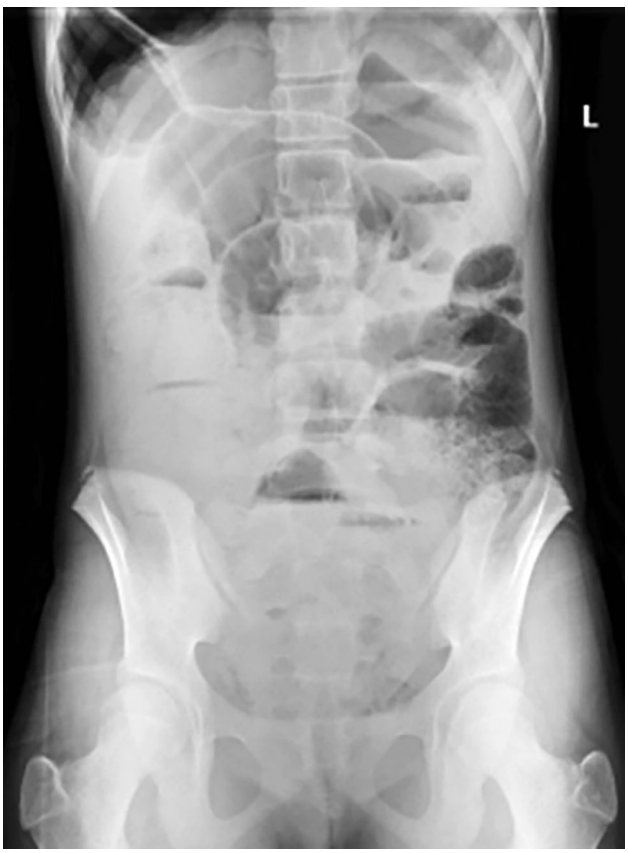

Figure 1. Radiographic features indicating intestinal obstruction. Upright abdominal radiograph on admission showing multiple air-fluid levels distributed in dilated bowel loops. These findings are suggestive of a mechanical intestinal obstruction. Statistical tests were not applicable for radiographic interpretation.

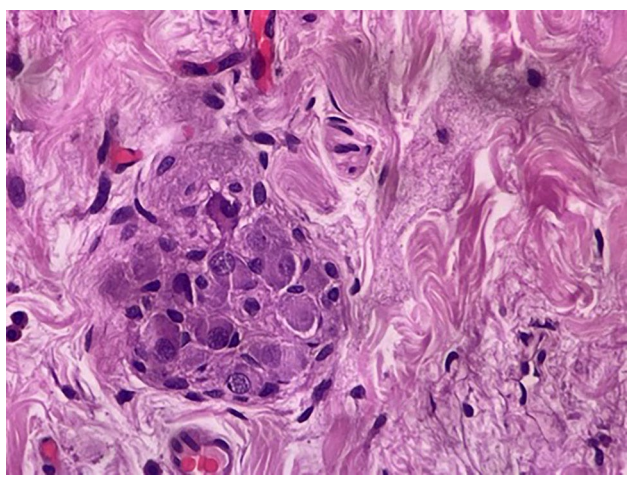

Figure 2. Presence of ganglion cells in rectal tissue. Histopathological image of rectal biopsy stained with hematoxylin and eosin, showing ganglion cells within the submucosal layer, indicating normal innervation (H&E, ×200). No significant pathology was noted. Statistical analysis was not required.

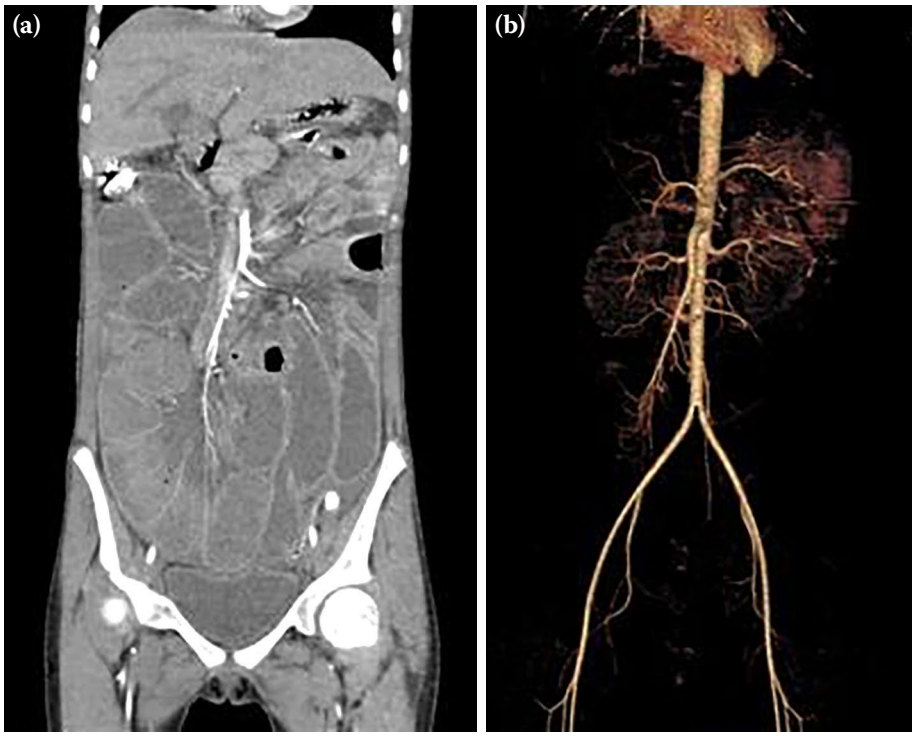

Figure 3. Normal vascular configuration on CT angiography. **(a)** Coronal CT angiography view showing no signs of vascular occlusion or abnormality. **(b)** 3D reconstruction image further confirming a normal anatomical and vascular configuration, effectively ruling out any vascular pathology. No statistical comparisons were performed.
CT, computed tomography; 3D, three-dimensional.

currently being followed under individualized care with a functioning ileostomy. Written informed consent was obtained from the parent of the patient.

DISCUSSION

Pediatric intestinal pseudo-obstruction is a rare group of disorders characterized by severe bowel motility impairment in the absence of surgically demonstrable mechanical obstruction. Although it typically presents in the neonatal or infantile period, some cases may first manifest during late childhood or adolescence. The present case represents a rare instance of adolescent-onset PIPO, initially suggestive of mechanical obstruction.[7]

Diagnosis of PIPO requires a comprehensive approach including clinical, radiological, histopathological, and genetic evaluation. Differential diagnosis with conditions such as Hirschsprung disease, NID, metabolic syndromes, and autoimmune enteropathies can be particularly challenging. While the presence of ganglion cells in rectal biopsy is critical for ruling out Hirschsprung disease, genetic testing becomes essential in cases where neuropathological analyses are inconclusive. In this case, persistent postoperative ileus led to rectal biopsy which excluded Hirschsprung disease, but the continued dysmotility warranted further investigation for PIPO.[8,9]

Pediatric intestinal pseudo-obstruction is often misdiagnosed as postoperative non-mechanical ileus, leading to unnecessary surgical interventions. In the present case, repeated laparotomies were performed under the impression of adhesive ileus following perforated appendicitis, yet intestinal function did not improve even after ileostomy. This highlights the importance of reevaluating the diagnosis in patients unresponsive to surgery. According to literature, more than half of patients with delayed PIPO diagnosis undergo unwarranted surgeries.[10,11]

Genetically, mutations in genes such as Actin Gamma 2, ACTG2, Filamin A (FLNA), Myosin Heavy Chain 11 (MYH11), and Thymidine Phosphorylase (TYMP) have been implicated in PIPO pathogenesis. Of note, ACTG2 mutations, associated with visceral myopathy, are the most common and follow an autosomal dominant inheritance pattern. However, the absence of mutations in many patients suggests a broader, yet undiscovered genetic heterogeneity. The lack of identifiable mutations in our case supports this hypothesis.[12,13]

The main goal of treatment is symptom control and support of intestinal motility. Prokinetic agents including domperidone, erythromycin, and neostigmine play important roles in the therapeutic protocol. Neostigmine, as an acetylcholinesterase inhibitor, enhances bowel peristalsis via the enteric nervous system. In our case, intravenous neostigmine reactivated the ileostomy, indicating therapeutic efficacy. For long-term management, nutritional status must be addressed through total parenteral and enteral nutrition and intestinal rehabilitation programs.[14,15]

In conclusion, although PIPO is rare, it should always be considered in the differential diagnosis of persistent ileus, particularly in the postoperative setting. Early diagnosis and multidisciplinary evaluation can prevent unnecessary surgical interventions and facilitate appropriate treatment, positively influencing patient prognosis. In long-term management, individualized therapeutic approaches, intestinal rehabilitation, and consideration of potential transplantation strategies emerge as key components to improving patients' quality of life.

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