Distal intestinal obstruction syndrome

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A 31-year-old woman with cystic fibrosis (CF) presents with a right-sided obstructive pyeloureteral junction stone complicated by urosepsis. Following an emergency treatment with ureteral stent placement, she is transferred to the intensive care unit for symptoms of septic shock and hemodynamic instability. Her clinical history reveals an exocrine pancreatic insufficiency and bilateral lung transplantation, both related to CF.

One week after admission, she starts vomiting and develops a distended, painful abdomen. A plain abdominal radiograph shows dilated small bowel loops in absence of a distended colon (Fig. A). This finding is new compared to the abdominal radiograph taken on admission (figure not shown). Despite treatment with nasogastric tube placement and parenteral nutrition, the pain persists and contrast-enhanced computed tomography (CT) of the abdomen is performed. CT confirms serious distension of the small bowel loops with exception of the terminal ileum, with the lumen of the latter filled with dense content (Fig. B and C). Similar dense material is seen in the non-distended caecal lumen. No other cause of mechanical intestinal obstruction (e.g. adhesions, internal herniation, volvulus, extrinsic compression...) is present. Based on the CT-findings the diagnosis of a distal intestinal obstruction syndrome (DIOS) is made. Conservative treatment is continued with complete regression of the abdominal complaints and restoration of the transit the following days.

Comment

DIOS is a syndrome unique to CF. It is defined as an acute, complete or incomplete, fecal/mucus obstruction in the ileocaecum. Previously the disease was called ‘meconium ileus equivalent’, referring to the meconium plugs that can be seen in neonates with CF. DIOS occurs in 16% of adults with CF, with the highest prevalence in the 2nd and 3rd decade, an age-group that is becoming significantly larger thanks to better treatment of CF. Pre-disposing factors for the development of DIOS include dehydration, pancreatic insufficiency or pancreatic enzyme supplement cessation and recent lung transplantation. The tentative diagnosis of DIOS is based on the clinical history, physical examination and a plain abdominal radiograph. CT is the imaging modality of choice to confirm the diagnosis of DIOS, to rule out other causes of intestinal obstruction and to prevent unnecessary surgical exploration. A small bowel obstruction in CF patients in combination with hyperdense mucofaeculent material in the terminal ileum/caecum is pathognomonic for DIOS. Conservative treatment is the rule.

Reference


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