Case Report / Olgu Sunumu

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Distal Intestinal Obstruction Syndrome in Patients with Cystic Fibrosis: Two Separate Cases in the Pediatric Intensive Care Unit

Kistik Fibrosisli Hastalarda Distal İntestinal Obstrüksiyon Sendromu: Çocuk Yoğun Bakım Ünitesinde Takip Edilen İki Ayrı Olgu Yönetimi

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Abstract

Distal intestinal obstruction syndrome (DIOS), also defined as the equivalent of meconium ileus, is a sign of complete or partial ileocecal obstruction with intestinal contents in patients with cystic fibrosis. DIOS may occur because of darkened intestinal secretions, pancreatic insufficiency, undigested food residues and sticky stool stasis. Patients apply with abdominal swelling, constipation, severe abdominal pain in the form of recurrent cramps and vomiting. In direct abdominal radiographs, dilated small intestines, air-fluid levels or foamy appearances are observed in the ileocecal region. Obstruction developed in patients with cystic fibrosis is treated with medical and surgical methods with a multidisciplinary approach depending on the degree of severity and symptoms. In this paper, two critically ill children with cystic fibrosis were presented who were followed up in the pediatric intensive care unit with a diagnosis of DIOS, with one treated conservatively and the other surgically; the treatment methods were also highlighted.

Keywords: Cystic fibrosis, distal intestinal obstruction syndrome, surgery

Öz

Mekonyum ileusu eş değeri olarak da tanımlanan distal intestinal obstrüksiyon sendromu (DIOS), kistik fibrozisli hastalarda bağırsak içeriği ile tam veya parsiyel ileoçekal obstrüksiyon kliniğidir. DIOS, koyulaşmış intestinal sekresyonlar, pankreatik yetmezlik, sindirilmemiş gıda kalıntıları ve yapışkan gaita stazı sonucunda meydana gelmektedir. Batında şişlik, kabızlık, tekrarlayan kramp şeklinde şiddetli karın ağrıları ve kusma kliniği ile hastalar başvurmaktadır. Düz abdominal grafilerde dilate ince bağırsaklar, hava-sıvı seviyeleri ya da ileoçekal bölgede köpüksü görünüm izlenmektedir. Kistik fibrozis hastalarında gelişen obstrüksiyon; derecesine ve semptomlarına bağlı olarak, multidisipliner yaklaşımla medikal ve cerrahi yöntemlerle tedavi edilmektedir. Bu bildiride; çocuk yoğun bakım ünitesinde DIOS tanısı ile takip edilen; biri konservatif, diğeri ise cerrahi olarak tedavi edilen iki kistik fibrozisli kritik hasta çocuktan bahsedilerek tedavi yöntemlerine dikkat çekilmek istenmiştir.

Anahtar Kelimeler: Kistik fibrosis, distal intestinal obstrüksiyon sendromu, cerrahi

Introduction

Cystic fibrosis is an autosomal recessive inherited disease that may lead to various clinical manifestations as a result of the presence of dark and sticky secretions due to a mutation in the chloride channels in secretory cells. In patients with cystic fibrosis, multisystemic problems, including those of the digestive system, are a result of the inability to secrete enzymes or obstructions in the channels. The second most common complications after respiratory system issues were related to the gastrointestinal system in 65% of the patients. Pancreatic or liver involvement may be present, as well as clinical pictures leading to intestinal obstruction.[1] Invagination, meconium ileus, distal intestinal obstruction syndrome (DIOS) and volvulus are the conditions that may cause intestinal obstruction.[2]

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DIOS also called meconium ileus equivalent, occurs due to darkened intestinal secretions, pancreatic insufficiency, undigested food residues, and sticky stool stasis in patients with cystic fibrosis. Patients may present with abdominal swelling, constipation, severe abdominal pain in the form of recurrent cramps, and vomiting. Direct abdominal radiographs show dilated small intestines, air-fluid levels or a foamy appearance in the ileocecal region.[3]

Various treatment methods are applied according to the symptoms developed in the patients and the degree of obstruction. While the use of nutritional fiber supplements, stool softening agents and oral polyethene glycol solutions are included in the treatment in the chronic period, clinicians attempt to open obstructions with conservative treatment methods, such as hydration, laxatives, drugs that increase gastrointestinal peristalsis, pancreatic enzyme supplements, enema applications or n-acetylcysteine oral and rectal enema applications in the acute period. However, surgical treatment is applied if the obstruction is not fully opened, and complications develop.[2,3] In this case report, two critically ill children with cystic fibrosis who were followed up with a diagnosis of DIOS in the pediatric intensive care unit were presented, and attention was drawn to their treatment management.

Case Reports

Case 1: A 12-year-old male patient who presented with a homozygous Phe508del mutation in the *CFTR* gene, with pancreatic insufficiency, Type 1 diabetes mellitus and chronic lung disease reported severe widespread abdominal pain for five days, an inability to pass stools, a loss of appetite and vomiting for one day. On physical examination, abdominal distention, hypoactive bowel sounds and generalised sensitivity were present; other system examinations were normal. Laboratory tests were normal except for leukocytosis. While air-fluid levels and gas passage to the distal area were not observed in direct radiography (Figure 1A), distension presented significantly in the jejunum and ileum; on abdominal ultrasonography (US), the small intestines were more prominent than the colon and filled with stool, with no paralytic ileus, which was evaluated as mechanical ileus. With the present clinical and radiological findings, given the DIOS in patients with cystic fibrosis, the patient was hospitalised in the intensive care unit. It was determined that he had no history of meconium ileus and had no previous intestinal obstruction attack. Intraabdominal pressure follow-ups were between 12 and 15 mmHg. Enteral feeding of the patient, who also had intraabdominal hypertension, was discontinued and decompression was performed with a nasogastric catheter. A rectal enema was initiated for the patient who did not pass stools. Despite this, there was no defecation; however, vomiting increased on the 3rd day of hospitalisation, and abdominal computed tomography (CT) was performed on the patient, whose abdominal pain worsened. CT revealed that the intestines were distended and filled with stool; the bowel loop was 4.1 cm at the widest part. Both oral n-acetylcysteine and rectal enema with n-acetylcysteine and oral paraffin liquid were added to the patient's treatment. On the 5th day of hospitalisation, there was abundant defecation, and his intraabdominal pressures remained within normal limits. The patient's complaints, whose DIOS symptoms regressed with conservative treatment without the need for surgery and who tolerated oral nutrition on the 7th day of hospitalisation, did not recur. Patient consent was obtained for this report.

Case 2: A 17-year-old male patient who was homozygous for the Phe508del mutation in the *CFTR* gene showed pancreatic insufficiency, chronic lung disease and atopic dermatitis, and two days of severe abdominal pain, a loss of appetite, vomiting and inability to pass stool for five days. Physical examination revealed abdominal distension, hypoactive bowel sounds and widespread tenderness, especially in the right lower quadrant. Other system examinations were normal. Laboratory examinations were normal except for the elevation of acute-phase reactants. Air-fluid levels were found on direct abdominal radiography (Figure 1B). In abdominal tomography, diffuse free fluid, collapsed colonic loops, diffuse distension and air-fluid levels and faeces densities in the jejunal and proximal ileal loops were observed. While he was hospitalised with a diagnosis of DIOS with the current findings, it was determined that the patient had no history of meconium ileus but had a previous intestinal obstruction attack, and he recovered without the need for surgery.

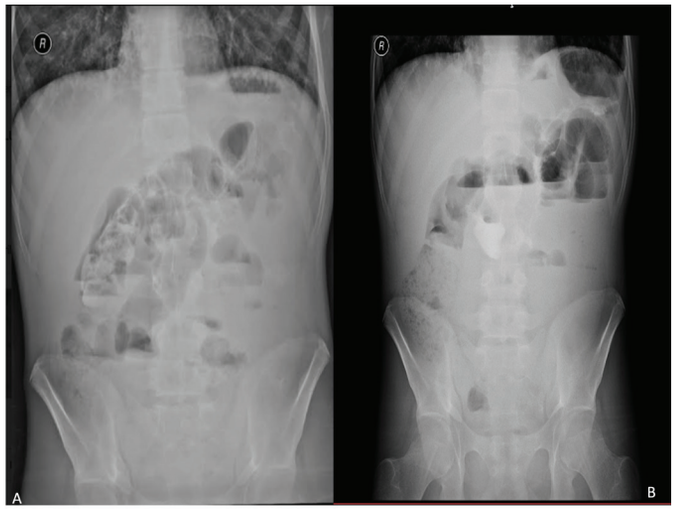

Figure 1. A) Direct abdominal radiography of a 10-year-old patient with distal intestinal obstruction syndrome diagnosis and **B)** Direct abdominal radiography of a 17-year-old patient with distal intestinal obstruction syndrome diagnosis before laparotomy

In his second DIOS attack, the oral intake of the patient was closed, and decompression was performed with a nasogastric catheter. Oral n-acetylcysteine and rectal enema were started as conservative treatments. In the follow-up, the patient had severe abdominal pain, and reported biliary vomiting followed by fecal vomiting; his complete intestinal obstruction symptoms did not regress, and laparotomy was performed by pediatric surgery. In the postoperative follow-up in the intensive care unit, intraabdominal pressures remained in the range of 6-9 mmHg; enemas with n-acetylcysteine were continued. The patient passed gas and stool two days after the operation; he did not have vomiting, tolerated oral feeding on the 5th day, and the acute abdomen symptoms did not recur. Patient consent was obtained for this report.

Discussion

Although there has been a decrease in mortality and morbidity in patients with cystic fibrosis with the advancement of new drugs, respiratory support strategies and approaches in disease exacerbations in recent years, severe complications are still seen. Therefore, complications that may require critical care can often be seen in patients with cystic fibrosis.[4] Patients with cystic fibrosis and distal intestinal obstruction syndrome need to be managed in a multidisciplinary manner, regardless of whether surgery or conservative treatment is applied. While this multidisciplinary approach includes the follow-up of respiratory functions, physiotherapy, the management of comorbid diseases and the regulation of nutrition with oral therapy in the preoperative period, in the postoperative period, it is necessary to focus on preventing pneumonia with early extubation, chest physiotherapy and early mobilisation.[5] Both of our patients were followed up by relevant departments in the intensive care unit with a multidisciplinary approach.

While 65% of the patients with cystic fibrosis are referred with gastrointestinal symptoms, Smith et al.[4] reported that intestinal obstruction and ileus were observed in 24% of critically ill children with cystic fibrosis in their study who were hospitalised in the intensive care unit. Although DIOS may affect patients of all age groups, it is frequently seen in adolescents and young adults.[6] Our patients were in the adolescent age group. The frequency of DIOS in young adults is 18.1%.[7]

The passage of faeces in the intestines slows down due to insufficient fluid secretion as a result of the defect in chloride channels, dehydration, and decreased fat reabsorption caused by pancreatic insufficiency; this plays a role in the pathogenesis of distal intestinal obstruction syndrome. In the differential diagnosis of DIOS, diseases such as chronic constipation, invagination, appendicitis, inflammatory bowel diseases and fibrosing colonopathy are included.[6] The diagnosis of complete or partial DIOS is made using the diagnostic criteria established by the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN) Cystic Fibrosis Study Group. Intestinal obstruction with air-fluid levels in the small intestines on direct abdominal X-ray and/or biliary vomiting, fecaloid in the ileocecal region and abdominal pain and/or distension constitute the diagnostic criteria.[8] Typical symptoms of the syndrome include abdominal distension, vomiting, weight loss and constipation. The most common obstruction is in the ileocecal junction. Direct abdominal radiographs are used in the diagnosis, with abdominal US and CT shown to help the diagnosis.[9] CT is the gold standard radiological examination with the advantage of precisely visualising the point of obstruction in defining intestinal obstruction and an obstructive mass.[10] Both of our patients were diagnosed with DIOS with ESPGHAN diagnostic criteria and radiological imaging methods, with air-fluid levels on standing direct abdominal radiography, intestinal obstruction and fecaloids seen in advanced imaging methods, constipation, complaints of abdominal pain, abdominal distension and vomiting, and they were all followed up.

The primary treatment of distal intestinal obstruction syndrome is non-surgical, and the conservative approach is successful in most cases. Most of the patients respond well to pancreatic enzymes, hydration, mucolytic agents, intestinal lavage solutions, stool softeners or laxatives, oral polyethylene glycol solution, enema and nasogastric drainage; the colonoscopic approach may eliminate the need for surgical treatment. The regulation of nutrition, oral osmotic laxatives, polyethylene glycol or n-acetylcysteine, is used to avoid DIOS attacks.[11] Surgical intervention is applied when there is no response to medical treatment, and the obstruction cannot be resolved, and when intussusceptions or volvulus develops. In a case series of 80 patients with DIOS, surgical treatment was required in 12.5%.[12] It has been reported in recent publications that the need for surgery has decreased to 3.9%.[5] In another study, it has been reported that only one (4.7%) of 21 DIOS attacks have required surgical intervention, while a pediatric patient who required surgery has been successfully treated with enterotomy and washing procedures.[13] Farrelly et al.[12] examined different surgical procedures over 20 years; they reported that most of the surgically treated patients were successfully treated with enterotomy and washing or small bowel resection with primary anastomosis. Conservative treatments were started after both patients were diagnosed. However, a complete response was obtained with conservative treatment in our first case at the age of 12, and the clinical picture of obstruction regressed. As there was no response to conservative treatments in our 17-year-old patient in his second DIOS attack, laparotomy was performed, enterotomy and washing procedures were performed, and his obstruction was treated.

In conclusion, DIOS is a common gastrointestinal complication that should be considered in patients with cystic fibrosis. Other colon and intestinal pathologies may also occur in these patients. Thus, it is critical to make a fast and accurate diagnosis and provide treatment. In eligible and stable patients, conservative and less invasive approaches should be first attempted to resolve the attack. Surgical treatment methods can also be used when patients with appropriate clinical symptoms do not respond to conservative treatment.

Ethics

Informed Consent: Patient consent was obtained for this report.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.M., A.S., D.Y., Ö.Ö.H., F.E., S.T.Ç., D.Ö., Concept: M.M., A.S., D.Y., Ö.Ö.H., F.E., S.T.Ç., D.Ö., Design: M.M., A.S., D.Y., Ö.Ö.H., F.E., S.T.Ç., D.Ö., Data Collection or Processing: M.M., A.S., F.E., Analysis or Interpretation: M.M., A.S., D.Y., Ö.Ö.H., F.E., S.T.Ç., Literature Search: M.M., A.S., D.Y., Ö.Ö.H., S.T.Ç., Writing: M.M., A.S., F.E.

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