CASE REPORT

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Chronic Idiopathic Intestinal Pseudo-obstruction

Kronik İdiopatik İntestinal Psödo-obstrüksiyon

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ÖZET

Kronik idiopatik intestinal psödo-obstrüksiyon, abdominal distansiyon, ağrı, bulantı, kusma semptomları ve rekürren intestinal obstrüksiyon atakları ile karakterize nadir görülen heterojen bir sendromdur. Her ne kadar radyolojik çalışmalar barsakta hava sıvı seviyeleri içeren dilate looplar ortaya koysa da, mekanik obstrüktif lezyon görülmemektedir. Bu motilite bozukluğunun iki patofizyolojik tipi miyopatik ve nöropatiktir.Burada, kronik idiopatik intestinal obstrüksiyon tanısı almış bir hasta sunulmakta ve ilgili literatür gözden geçirilmektedir. Intestinal psödo-obstrüksiyon tekrarlayan ataklar ya da barsak obstrüksiyonunun devamlı bulguları ile karakterize, radyolojik obstrüksiyon belirtileri de dahil olmak üzere mekanik obstrüktif lezyon olmaksızın seyreden klinik bir entitedir. Psödo-obstrüksiyon hem ince barsak hem de kolonda ortaya çıkabilir ve kronik ya da akut olabilir. Ogilvie sendromu olarak da bilinen akut kolonik psödo-obstrüksiyon, intestinal obstrüksiyonun en yaygın formu olsa da kronik idiopatik

ABSTRACT

Chronic idiopathic intestinal pseudo-obstruction (CIIP) is a rare heterogeneous clinical syndrome characterized by recurring episodes of symptoms and signs of intestinal obstruction which include abdominal distention and pain, nausea, and vomiting. Although radiologic studies reveal dilated loops of bowel with air fluid levels, but there is no any mechanically obstructing lesion. The two pathophysiologic types of this motility disorder are myopathic and neuropathic. Herein, we reported a patient with CIIP and attempted to review the relevant literature. Intestinal pseudo-obstruction is a clinical entity characterized by recurring episodes or continuous symptoms and signs of bowel obstruction in the absence of a mechanically obstructing lesion, including radiological features of obstruction. Pseudo-obstruction occurs in both the small bowel and colon and it can be either acute or chronic. Although acute colonic pseudo-obstruction, also known as Ogilvie syndrome, is the most common form of intestinal obstruction, CIIP

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intestinal psödo-obtrüksiyon (KİP) ender rastlanan ve ağır, fonksiyon bozukluğuna neden olan, heterojen bir hastalıktır. Kajal'ın interstisyel hücrelerinin KİP patogenezinden sorumlu olduğu düşünülmektedir. Bazen akut karın bulguları görülebilir.

Anahtar Kelimeler: Kronik obstruksiyon, İnce barsak, Kolon, Psödo-obstrüksiyon

has been defined as a rare and severe, disabling and heterogeneous clinical syndrome. Interstitial cells of Cajal (ICC) have been simplied in the pathogenesis of CIIP. Sometimes acute abdominal findings can be seen.

Key words: Chronic obstruction, Small bowel, Colon, Pseudo-obstruction

Introduction

Intestinal pseudoobstruction is a clinical entity characterized by recurring episodes or continuous symptoms and signs of bowel obstruction in the absence of a mechanically obstructing lesion, including radiological features of obstruction.[1] The two pathophysiologic types of this motility disorder are myopathic and neuropathic.[1] Pseudoobstruction occurs in both the small bowel and colon and it can be either acute or chronic. Acute colonic pseudo-obstruction, also known as Ogilvie syndrome, is the most common form of intestinal obstruction, CIIP has been defined as a rare and severe, disabling and heterogeneous clinical syndrome.[2] Interstitial cells of Cajal (ICC) have been implied in the pathogenesis of CIIP.[2] Sometimes acute abdominal findings can be seen.[2] Herein, we reported a patient with CIIP and attempted to review the relevant literature.

Case Report

Herein, we report a 60-year-old woman with mega colon and pseudo-obstruction. She had a history lasting for 3 years, with episodes of progressive abdominal distension and constipation which were interpreted as chronic constipation. Albeit her complaints were progressively severe, she was not admitted for further investigation. Although spontaneous defecation and flatus occurred, she had progressive and severe abdominal distention, edema of anterior abdominal wall (Fig.1), pain, nausea, and vomiting for three days.

Physical examination revealed metallic sounds.

Radiological studies revealed massive dilated loops of bowel with air-fluid levels (Fig. 2). The patient admitted to emergency room at midnight. Also the patient had acute abdomen signs. Colonoscopy could not be done and the patient underwent operation.

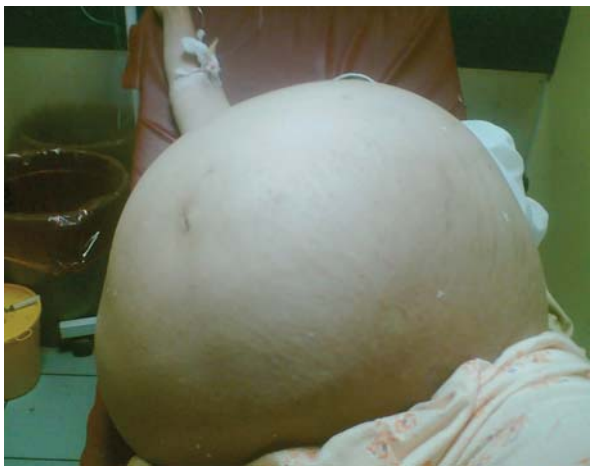

Figure 1. The preoperative presentation of the patient (front view).

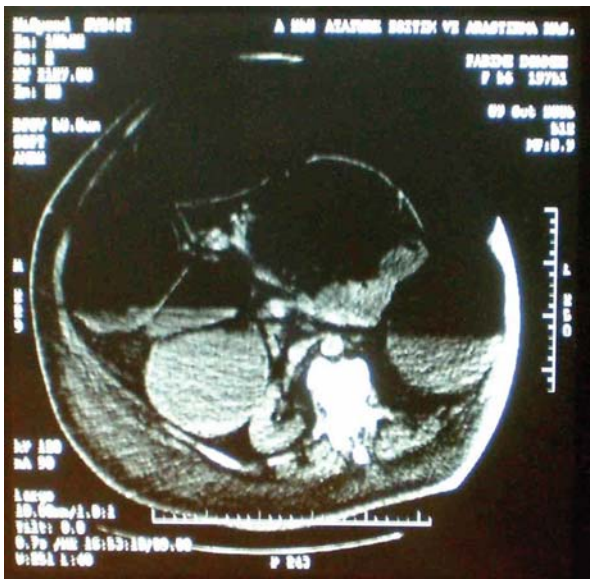

Figure 2. Computed tomography examination showed marked dilated loops of bowel with air fluid levels.

At laparotomy, we observed that the colon was severely dilated, thin-walled, and some loops included ischemic areas (Fig. 3, 4). Because of the ischemic areas, extended left hemicolectomy and Hartmann procedure were performed instead of colostomy. The patient was reoperated because she developed acute abdominal findings at postoperative day 8. During second laparotomy, colonic stump necrosis and intra abdominal abscess were observed. The loss of the patient was sepsis and sepsis-related lung problems at postoperative day 10. Macroscopic examination showed that the colectomy material was 200 cm in length and 20 cm in diameter. Neither a neoplastic process nor obstruction were seen. The number of ICC was very low in the Aurbach plexus, immunohistochemically. These findings were consistent with the diagnosis of CIIP with loss of c- kit positive cells of Cajal. These findings were seen at the colon diffusely. However; histopathological examination of material showed vacuolated myocytes and fibrosis in the muscularis propria (Fig. 5) which is seen in slow transit constipation. So maybe the two entities were together in this patient as a subpopulation of CIIP.

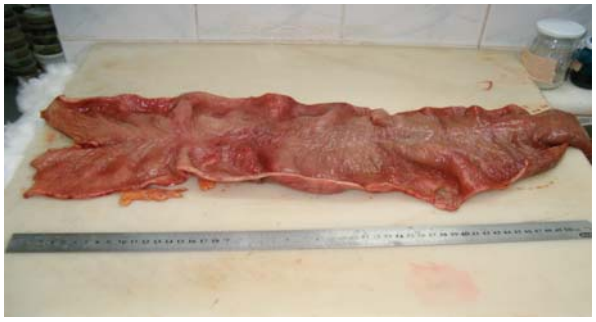

Figure 3. Subtotal colectomy material (open view). Thin wall of the colon is remarkable.

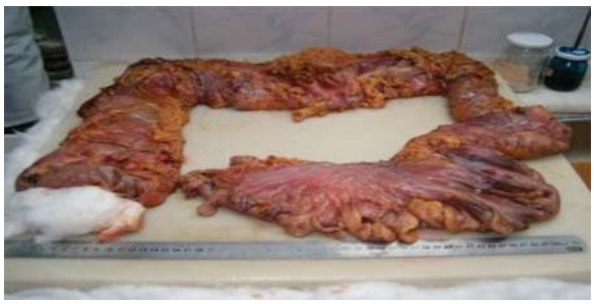

Figure 4. Subtotal colectomy material.

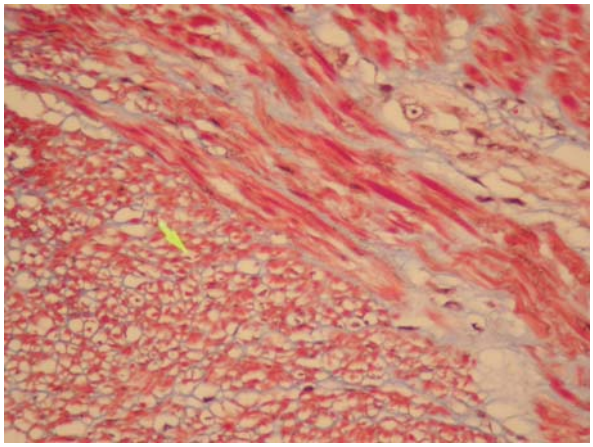

Figure 5. Microscopic appearance of colonic wall. Vacuolated myocytes and fibrosis in the muscularis propria (stained Mason Trichrom).

Discussion

Idiopathic intestinal pseudo-obstruction is an uncommon disorder of gut motility which is characterized by the failure of the intestinal tract to propel its contents appropriately. This leads to signs and symptoms of bowel obstruction and in the absence of an associated systemic disorder or the administration of drugs known to result in bowel dismotility, is termed CIIP. The primary feature is clinical and radiological evidence of intestinal obstruction in the absence of a mechanical lesion. The condition was first recognized by Dudley *et al*.[3] in 1958 who described 13 patients with clinical features of bowel obstruction in the absence of a mechanical cause. The clinical syndrome of CIIP most commonly occur secondary to diseases such as progressive systemic sclerosis, amyloidosis, and small cell carcinoma of the lung. The term CIIP was first coined by Maldonado, *et al*.[4] in 1970, who described five patients with recurrent unexplained episodes of intestinal obstruction, diarrhea, and weight loss, which sometimes progressed to death from starvation. Characteristic symptoms, shared by mechanical obstruction, include cramping abdominal pain, vomiting, constipation often followed by diarrhea, tender distended abdomen, high pitched bowel sounds and abdominal x-rays revealed dilatation of small and large intestine and delayed gastric emptying on various admissions. Dilatation of other viscera, such as the renal pelvis, ureter or urinary bladder, is identified in a minority of patients. In most patients a major differentiating feature from obstruction may be the presence of diarrhea

rather than constipation. Steatorrhea is secondary to an overgrowth of anaerobic bacteria in the motionless dilated loops of bowel.

Interstitial cells of Cajal have been shown to be the pacemaker cells of the intestine and have been implied in the pathogenesis of a number of gastrointestinal motility dysfunctions. Most patients with visceral myopathy have light microscopic changes of smooth muscle fibrosis and vacuolar and other degenerative changes in the circular and longitudinal layers of the intestinal wall, although the changes often involve only one muscle layer. Future routine use of special techniques such as electron microscopy, immunohistochemical staining for specific contractile elements, and more refined staining for neurological elements will decrease the number of patients in whom histological examination is apparently "normal".

Currently, treatment of CIIP is not standardized. Although laxatives, anticholinesterase agents, prokinetics and other drugs have been used in the past its usefulness was demonstrated of in controlled trials. It has been showed that neostigmine, via IV, 2.5 mg over 2 to 3 min; ameliorate symptoms of acute colonic pseudo-obstruction. Some authors provided that erythromycin could be benefit for preventing and/or diminishing the attacks of pseudo-obstruction.[1]

Colonoscopic decompression is a standard method to assure to remove air from the colon in these patients. The risk of perforation of the colon is very high, because massive dilated segments of the colon. This procedure can be achieved to reduce dimension of dilated colon, but dilatation usually recurs. The role of surgical treatment in these disorders remains poorly defined. Patients vary in their clinical expression and problems, despite even identical histology. Surgical treatment therefore needs to be tailored to the individual symptoms and objective evidence of regional disturbances in transit. Surgery in the form of bypass, limited resection, or decompressing venting stomas has had varying success in our patient and in the literature.[5,7] Visceral neuropathies are more commonly sporadic, although there have been reports of familial cases.[8,9] Patients having laparotomy for apparent obstruction who are found to lack a mechanical cause should have tissue processed to allow special examinations. The prognosis in these patients often depends on that of the underlying disorder. In contrast there is a group of patients with a similar pseudo-obstructive syndrome in whom there is no recognized underlying non-gastrointestinal disease. These patients with CIIP are all thought to have an abnormality of either enteric smooth muscle (visceral myopathy) and interstitial cells of Cajal.[10,11]

In summary, primary CIIP is a heterogeneous uncommon disorder of gut motility which must be differentiated from mechanical intestinal obstruction.

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