Chronic idiopathic intestinal pseudo-obstruction: Report of a case

Kronik idiopatik intestinal psödo-obstrüksiyon

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Kronik idiopatik intestinal psödo-obstrüksiyona genellikle enterik sinir yada düz kasın hastalığı sebep olur ve rekürren intestinal obstrüksiyon ataklarına yol açar. Hastalığın klinik belirti ve bulguları ile ya da rutin labarotuar incelemeleri ile ayırıcı tanısının yapılması imkansızdır ve çoğunlukla hastaya kesin tanı konuluncaya kadar birkaç kere gereksiz laparatomi yapılmış olur. Kesin tanı için tam kat barsak biyopsisi gereklidir. Burada; PAS boyası ile dejenere olmuş myofibrillerde, sitoplazmada inklüzyon cisimlerinin mikroskopik olarak tanımlanması ile tanı konmuş ve kronik idiopatik intestinal obstrüksiyon tanısı almış bir hasta, literatür ışığında sunulmaktadır. Bu hasta altıncı dekatta tanı alan birkaç hastadan biridir.

Anahtar kelimeler: Kronik intestinal psödo-obstrüksiyon, visseral miyopati, inklüzyon cisimciği

Chronic idiopathic intestinal pseudo-obstruction is usually caused by disease of the enteric nerves or smooth muscle and results in recurrent episodes of intestinal obstruction. It is impossible to identify by clinical appearance and routine laboratory study, and there are a few reports of mostly unnecessary laparotomy. A full-thickness intestinal biopsy is necessary for a certain diagnosis. Here, a patient diagnosed microscopically (In periodic acid-Schiff [PAS] staining, PAS is positive and inclusions (PASx1000) consist of degenerated myofibrils) with chronic idiopathic intestinal pseudo-obstruction is presented in light of the available literature. Diagnosis of chronic idiopathic intestinal pseudo-obstruction in this patient was delayed until the sixth decade.

Key words: Chronic idiopathic intestinal pseudo-obstruction, visceral myopathy, inclusion bodies

INTRODUCTION

Chronic idiopathic intestinal pseudo-obstruction (CIIP), a syndrome of ineffectual motility due to a primary disorder of the enteric nerve or muscle, is rare and is a disorder of intestinal motility, characterized by recurrence of continuous symptoms and signs of intestinal obstruction in the absence of true mechanical obstruction. It is impossible to identify by clinical appearance and routine laboratory research, and there are a few reports of mostly unnecessary laparotomy. Early diagnosis of intestinal pseudo-obstruction is important to avoid repeated laparotomies. Generally, the age at first operation is in the fifth or sixth decade of life. A full-thickness intestinal biopsy is necessary for a certain diagnosis.

Herein, we present a patient who was diagnosed as having intestinal obstruction (etiology: dolichocolon and/or megacolon or brid ileus) and operated, and who was later diagnosed as CIIP. We discuss the case in light of the available literature. Diagnosis of CIIP in this patient was delayed until the sixth decade of life.

CASE REPORT

A 65–year-old male patient was repeatedly examined in the emergency unit for abdominal distension, vomiting and absence of bowel movements for more than 4-5 days. His history began two years before, with episodes of progressive abdominal distension and constipation. In the last year, these episodes increased in severity and frequency, leading to food abstention and secondary denutrition. There was no history of metabolic, neurological, cardiovascular or pulmonary disease, and no history of abdominal cancer, inflammatory processes or trauma. No concomitant medication was taken. There was only a history of gastric operation due to peptic ulcer at the age of 30 years.

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Physical examination during the obstructive episodes revealed marked abdominal distension with hypoactive bowel, in the absence of abdominal mass or peritoneal signs. Laboratory screening showed normal values of serum electrolytes, hemoglobin, hematocrit, white blood cells, blood glucose, creatinine, and coagulation profile.

Abdominal ultrasound did not reveal any sign of small bowel or colonic mechanical obstruction; distended bowel segments were observed, and there was a little free peritoneal fluid in the peritoneal cavity.

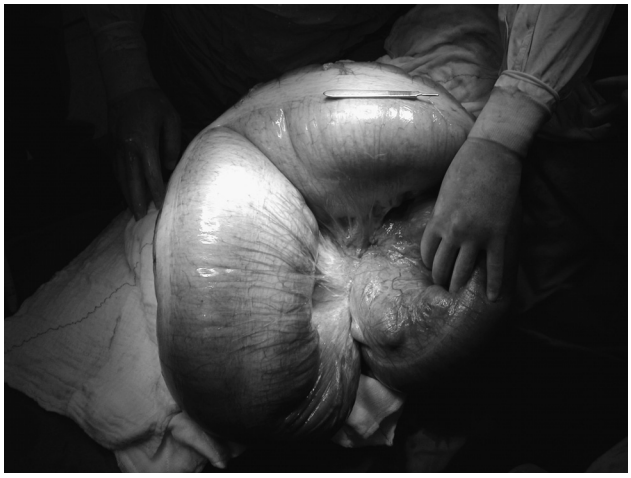
Laparotomy was done with general anesthesia under emergency conditions. The colon and sigmoid colon reaching 20 cm in diameter was found torsioned during abdominal exploration, and intraoperative diagnosis was dolichocolon (Figure 1). Due to the procedure, total colectomy plus end ileostomy were performed.

No postoperative complication was observed after colectomy and no obstructive episode recurred.

Cross-sections 4-5 microns in thickness from tissue-immersed paraffin were taken and stained with hematoxylin-eosin (HE). Nonspecific inflammation findings were monitored. It was observed that the longitudinal (outer) muscle layer of the musculus propria was atrophic in some areas, while in others it was thicker. In addition, severe vacuolar degeneration was observed; there was a gray-light pink color between cytoplasmic inclusions. There was no special observation in the circular muscle layer. Severe vacuolar degeneration (HEx200) and cytoplasmic inclusions (HE, x400) were seen in the longitudinal muscle layer. Periodic acid-Schiff (PAS) staining was positive and inclusions (PASx1000) consisted of degenerated myofibrils (Figure 2).

DISCUSSION

The etiology of CIIP is unknown. This syndrome is classified as primary or secondary. Primary CIIP is also classified as either myopathic or neuropathic depending on the histopathology. This form is seen commonly in children and is usually symptomatic during or immediately after birth. The secondary or acquired form is more common in adults than children. The causes of secondary CIIP are gastrointestinal smooth muscle and nervous system disease, and endocrinologic, metabolic, autoimmune disorders, and infections (i.e., cytomegalovirus [CMV], Epstein-Barr virus [EBV]) (1).



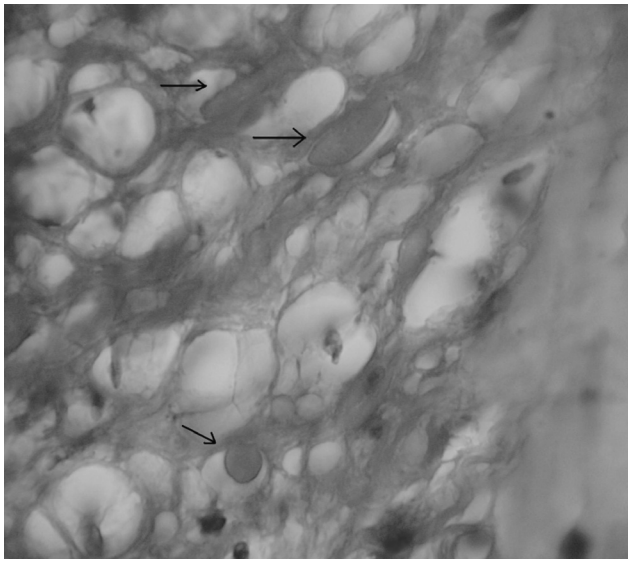
Resim 1. Operative view of the colonic segments

The most important factor for diagnosis of pseudo-obstruction is to have a high index of suspicion. The patient's history may provide a clue to the absence of any mechanical obstruction.

The condition of CIIP is rare. Literature related to primary CIIP consists of patients with familial visceral myopathy showing different clinical syndromes and underlying pathologies (2).

Lack of the normal pacemaker activity, usually generated by the interstitial cells of Cajal, could account for the obstruction (3). Neurotransmitters such as nitric oxide (NO) can also play a role in generating pseudo-obstruction.

Mann and coworkers (2) believe that a dilated gut



Resim 2. PAS is positive and inclusions (PASx1000) consist of degenerated myofibrils

is an indisputable abnormality. They also believe that histological assessment remains the gold standard for diagnosis, although its interpretation is sometimes subjective or indeterminate, and occasionally the appearance is normal. Advances in the use of special techniques such as contractile protein immunohistochemistry are likely to decrease the number of patients with "normal" histology in the future (2). In our case, it has not been decided whether or not the staining with HE plus dilated colon reaching 20 cm in diameter and staining with PAS (+) indicate a typical histology.

Most patients have obstructive symptoms for many years prior to laparotomy. Recognition of the true underlying abnormality was often delayed in the published reports (4). In our case, the diagnosis was delayed to the age of 65 years.

The most common misdiagnosis is obstruction of the duodenum by the superior mesenteric artery. Other diagnoses included achalasia, diverticulosis, psychogenic vomiting, functional bowel disease, megacolon, colitis, megaduodenum, malabsorption syndrome, and adhesions (4). In our case, the preoperative diagnosis was adhesion due to operation or megacolon.

The role of surgical treatment in these disorders remains poorly defined. Patients vary in their clinical expressions 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, such as might be observed with radio isotope studies. Varying success has been achieved in our patients and in the literature with surgery in the form of bypass, limited resection, or decompressing venting stomas (5, 6). Our patient with total colectomy + terminal ileostomy has no problem during the three-year follow-up.

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. Patients undergoing laparotomy for apparent obstruction who are found to lack a mechanical cause should have tissue processed to allow special examination. Such processing includes the storage of tissue in liquid nitrogen for specific immunohistochemical staining and glutaraldehyde for electron microscopy (4). As a result, most cases could not be diagnosed by PAS (+) staining, and diagnosis based on HE with inclusion corpuscles was doubted.

In summary, primary CIIP is a heterogeneous disorder. The major aims in management should be early diagnosis, avoidance of unnecessary repeated laparotomies, maintaining nutrition, symptom control, attempts at restoring motility, and the commitment to long-term care.

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