Case Report and Review of the Literature

An Obscure Colon Dilatation and its Underlying Cause: Case Report and Literature Review

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ABSTRACT

Introduction: Congenital as well as acquired diseases may be responsible for the development of a megacolon. In adult patients, Clostridium difficile associated infection as well as late-onset of Morbus Hirschsprung disease are known to cause a megacolon. In addition, malignant as well as benign colorectal strictures may lead to intestinal dilatation. In case of an idiopathic megacolon, the underlying cause remains unclear.

Case Presentation: We describe the case of a 44-year-old male patient suffering from a long history of chronic constipation. He presented with an obscurely dilated large intestine with bowel loops up to 17 centimeters in diameter. Radiological as well as endoscopic examination gave evidence of a spastic process in the sigmoid colon. The patient was treated with a subtotal colectomy and the intraoperative findings revealed a stenotic stricture in the sigmoid colon. Since the histological examination did not find a conclusive reason for the functional stenosis, an immunohistochemical staining was advised. This showed a decrease in interstitial cells of Cajal (ICC) in the stenotic part of the sigmoid colon.

Discussion: This case report describes a patient with an idiopathic megacolon, where the underlying cause remained unclear until an immunohistochemical staining of the stenotic colon showed a substantial decrease of ICCs. Various pathologies leading to a megacolon are reviewed and discussed.

Introduction

The megacolon, with its variety of underlying causes, is a rare but potential life-threatening diagnosis in visceral surgery. Acquired as well as congenital forms are described in the literature. Moreover, idiopathic manifestations of the megacolon leave the causal condition unclear.

Acquired forms often originate from mechanical obstructions. In particular, stenotic processes due to malignant as well as benign strictures but also obstructions like sigma volvulus or functional stenosis without any morphologic reason can lead to a dilated colon [1]. Colonic dilatation caused by congenital factors frequently occur in patients suffering from Hirschspring disease (HD) [2–4]. Late-onset forms of HD are described in the literature but are only rarely diagnosed [5]. If concomitant systemic toxicity is present, a toxic megacolon must be taken into consideration. Infectious colitis or an inflammatory bowel disease (IBD) may be part of the patient’s medical history. A dysfunction of ICCs, the intestinal pacemaker cells, seems to be involved in causing slow-transit-constipation and may be further associated with the development of a megacolon [6]. This idiopathic form comes along with a pathologically distended colon without organic reasons that might explain a dilatation and lengthening of the colon [7, 8]. The management of patients with a megacolon can be challenging, especially if patients...
suffer from a slow-transit-constipation or a chronic constipation for a long period of time.

Here, we present a case of a patient suffering from a megacolon who finally underwent subtotal colectomy. However, the underlying cause for the functional stenosis of the colon remained unclear until a re-staining of the specimen was performed. Various pathologic conditions that may be associated with the development of a megacolon are reviewed in the discussion section.

Case Presentation

A 44-year-old male patient presented himself at our outpatient clinic with an obscurely dilated abdomen, increasing dyspnea and tensional pain. He reported sequences of constipation altering with sequences of diarrhea since his childhood. The first abnormal intestinal distensions occurred at an age of twenty. His past medical history describes a Hashimoto-thyroiditis, an evacuation of an epidural hematoma after traumatic head injury 30 years ago, and a laparoscopic removal of his appendix and his gallbladder several years ago.

Chronologically, his first visit at the Department of Internal Medicine was one year before the current visit due to abdominal pain especially in the right upper abdomen as well as nightly back pain. At that time, on physical examination the abdomen was clearly above the thoracic level, borborygmus was audible but the abdominal wall was soft, and the patient did not suffer from severe abdominal pain. He reported a weight loss of eleven kilograms within the last year. He suffered from constipation that aggravated as soon as his laxative medication was paused.

The attending gastroenterologist intensified the conservative treatment and a colonoscopy was performed. Due to an impassable obstruction in the proximal sigmoid colon, the examination had to be interrupted. Subsequently, an abdominal computer tomography was performed, which confirmed the colonic distension with segments dilated up to twelve centimeter in diameter. Additionally, a five-centimeter-long stenotic process in the sigmoid colon was described, which was considered responsible for the colonic distension (Figure 1). For further clarification, a second colonoscopy was performed in which a spastic but passable bowel segment in the sigmoid colon was described.

In a further diagnostic step, the colonic transit time using radiopaque pellets was measured. The abdominal X-ray four days after application showed about forty pellets in the right colon (Figure 2). The patient was transferred to our abdominal surgery outpatient clinic. The results of all recently performed radiological examinations were discussed interdisciplinary, concluding that a tumor or an aganglionic segment in the sigmoid colon were the most probable cause for the megacolon.

After nine months without any further recorded outpatient clinic visits, the patient presented himself again at our clinic with an obscurely dilated abdomen, increasing dyspnea and tensional pain. He reported on constipation altering with discharge of very small amount of liquid stool. In our outpatient clinic, a soft rectal tube was inserted, but neither rectal stool nor gas discharge was observed. In the abdominal X-ray (Figure 2), a colonic distension with bowel segments up to 17 centimeters in diameter was found. Additionally, nearly all of the nine-month-ago orally applied radiopaque pellets were still projected onto the right colon.

Figure 1: Abdominal computer tomography revealed a colonic distension up to twelve centimeters. There was evidence of a five-centimeter-long stenotic process in the sigmoid colon (yellow arrows).

Figure 2: A) After orally applied radiopaque pellets, an abdominal X-ray was performed. It revealed a pathological colonic distension. About forty of the radiopaque pellets were projected onto the right colon. The small intestine appeared not distended. B) Nine months later, the initially applied radiopaque pellets were still detected in projection onto the right colon. The colon was distended up to 17 centimeters. This time, the small intestinal loops appeared dilated up to five centimeters as well.
Moreover, distended small intestine loops up to five centimeters in diameter were described as well. Blood tests revealed a leukocytosis of 12.95 G/L, a slight thrombocytosis (425 G/L), an increased fibrinogen (486 mg/dl) and a mild hyperkalemia (5.12 mmol/l). Neither viral antigens nor virus-specific antibodies for hepatitis-A, -B, -C or HIV were found. Furthermore, endomysial as well as anti-tissue transglutaminase antibodies were below detection level. The tumor marker alpha-fetoprotein was negative as well.

Now, given the substantial symptomatic burden, the patient gave his consent to the recommended operation. Subsequently, an explorative laparotomy was performed. As already radiographically assumed, intraoperatively, an obscurely dilated colon was found (Figure 3). A filiform stenotic process in the rectosigmoid junction, most likely triggered by inflammation, seemed to be causal. A subtotal colectomy with a side-to-side ileorectal anastomosis was performed.

Figure 3: The pictures show the patient before (A) and after (C) subtotal colectomy in patient’s profile. Picture A shows the extent of colonic distension as well as a slightly livid color of the abdominal skin. In picture (B), the yellow arrow points out the stenotic process in the sigmoid colon.

Figure 4: The figure shows A) immunohistochemical stainings of the stenosis in the sigmoid colon and B) an unaffected section of the sigmoid colon. i) c-Kit staining was used for interstitial cells of Cajal (ICC) and ii) S100 staining for glia cells of the nerval plexus. (Ai) and (Aii) show a decreased number of ICCs and glia cells in comparison to (Bi) and (Bii).

The histological work-up of the colon showed a non-inflamed diverticulosis in the sigmoid colon. Of particular interest was a section of the sigmoid colon, which showed a stenosis over a length of 13 centimeters. Neither a tumorous process nor any other underlying cause for the stenotic process was found. Therefore, an immunohistochemical staining of the stenotic part of the specimen was conducted with the main focus on the investigation of ICCs and structures of the nerval plexus. The staining of the ICCs was performed by using a c-Kit antibody whereas S100 was used as a marker for glia cells of the nerval plexus. A decrease of both, ICCs as well as glia cells of the nerval plexus, was observed in the stenotic part when compared to an unaffected part in the sigmoid colon (Figure 4).

During the postoperative course a stepwise progression to full diet was achieved and the patient was discharged 10 days after an uneventful postoperative period. During the following months, no further visits were documented. In a telephone contact, the patient reported a slightly increased defecation frequency up to 3 times a day but no other complaints.

Discussion

A variety of underlying diseases such as mechanical obstructions, infectious disease, inflammation as well as congenital disorders may lead to a megacolon. [4, 5, 9-15]. Besides benign and malign tumorous processes causing obstruction, a common cause of a mechanical bowel obstructions leading to a megacolon is the volvulus [9, 10]. It occurs when the intestine twists around a fixed point and may involve either the small or the large bowel [1]. The colonic volvulus mostly affects the sigmoid colon and the cecal region [10]. In the US, the sigmoid volvulus is the third most common cause for colonic obstruction after cancer and diverticulitis [11]. In this actual case, neither the preoperatively diagnostic workup nor the intraoperative findings gave evidence for colonic volvulus.

Non-mechanical causes may be an infectious colitis as well as an inflammatory bowel disease (IBD), potentially leading to a toxic megacolon [12, 13]. Severe colitis can be caused by *Clostridium difficile* infection [12]. Typically, diffuse ulcerations, raised mucosal nodules and pseudomembranes are present at endoscopic examination [14]. Rarely, amoebic colitis, cytomegalovirus colitis as well as infections with *Salmonella*, *Shigella* and *Campylobacter* can also be associated with a toxic megacolon [14, 15]. A toxic megacolon is per definition accompanied by systemic toxicity [16]. Patients with IBD in their
system is assumed to be causal [27, 28, 29]. In cases of Ogilvie’s syndrome, no mechanical reasons for the intestinal obstruction can be found. The here presented patient unlikely suffered from an Ogilvie’s syndrome since no recent surgery took place and a long history of constipation was reported.

In the presented case the AMC may be the best fitting diagnosis. The term describes a condition of a pathologically distented colon without organic reasons, explaining the dilatation and lengthening of the colon [7, 8]. A review of Cuda et al. proposed following criteria for the diagnosis of AMC: (1) the exclusion of organic disease; (2) a sigmoid diameter of ~10 cm on abdominal X-ray or barium enema; (3) and symptoms including constipation, distension, abdominal pain and gas distress [8]. AMC still remains a diagnosis by exclusion but a dysfunction of ICCs was suggested to be the most profound histopathological finding [8]. These cells can undergo cyclic, spontaneous depolarisation leading to slow electric waves, which migrate towards the muscle cells [30]. Therefore, they play an important role as pacemaker cells, driving the activity of smooth muscle cells in the gastrointestinal tract [6, 31]. Lesions of these interstitial gut-pacemaker cells may cause a slow-transit constipation that further contributes to the development of a megacolon.

An immunohistochemical staining of the surgical specimen provided important insights: ICCs express c-kit, a receptor tyrosine kinase that is specific for ICCs. Hence, antibodies to the c-Kit protein label ICCs selectively. However, nerve or glia cells are not contributing to c-Kit positive cells [32, 33]. An antibody to S100 was used to stain glia cells. The immunohistochemical staining of the stenotic part of the colon revealed a lower number of ICCs in a reduced number of nerve plexus cells compared to an unaffected part of the sigmoid colon.

Taken together, the substantial decrease in ICCs seems to be the most reasonable pathophysiological mechanism, which led to the described megacolon.

**Conflicts of Interest**

None

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