Mechanical Bowel Obstruction Due to Localized Extensive Amyloidotic Involvement in the Left Colon

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Abstract Amyloidosis is a disorder caused by extracellular deposition of insoluble protein fibrils in various tissues. Colonic amyloidosis is a rare clinical manifestation which can mimic tumor, inflammatory bowel disease, or ischemic colitis. The most common symptoms of colonic amyloidosis include bleeding, ulceration, diarrhea, and abdominal pain. However, colonic obstruction due to extensive submucosal amyloidosis is extremely rare. Considering the wide variety of symptoms and potentially effected organs, the diagnosis can be challenging particularly in emergent conditions. In this paper, a case with mechanic bowel obstruction due to extensive amyloidotic involvement of the left colon is presented.

Keywords Gastrointestinal tract · Amyloidosis · Ileus · Colectomy

Introduction

Amyloidosis is a heterogeneous group of disorders characterized by systemic or localized extracellular deposition of an

abnormal protein material in the interstitium of organs and tissues, often resulting in functional impairment [1]. Although amyloidosis can occur in virtually any organ system in the body, including the gastrointestinal tract, localized amyloidosis in the gut is rare [2, 3]. Here, we present a case that required emergent surgery due to extensive localized amyloidotic involvement of the descending colon.

Case Report

A 76-year-old male patient was admitted to the emergency unit with a 10-day history of constipation, progressive abdominal distention, and discomfort. For several years, he had suffered from difficulties of defecation and intermittent rectal bleeding. Very recently, he had undergone full colonoscopy a month prior to presentation. Severe luminal narrowing and wall thickening were detected in the left colon and also the colonic mucosa was too fragile and friable due to excessive edema. Multiple biopsy was taken during colonoscopy, and histopathological analyses unveiled fibrosis, congestion, and nonspecific findings of chronic inflammation.

His blood pressure was 140/90 mmHg, pulse was 88 beats/min, and the temperature was 37.8 °C. Abdominal examination revealed diffuse abdominal distention and rebound tenderness with marked guarding. Bowel sounds were diminished. Digital rectal examination was normal. Full blood count, biochemical profile, and chest X-ray showed no abnormalities. All colonic segments, proximal to the left colon, were dilated in upright direct abdominal X-ray.

A subsequent computed tomography (CT) of the abdomen, and pelvis with oral and intravenous contrast demonstrated diffuse mural thickening, and narrowing extending from the descending colon to the rectosigmoid, and noticeably dilatation in all proximal colonic segments (Fig. 1a). A provisional diagnosis of large bowel obstruction secondary to a tumor in

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Fig. 1 **a** Diffuse mural thickening and narrowing extending from the descending colon to the rectosigmoid in abdomen CT scan (*white arrows*), **b** serosal surface discoloration extending from the descending colon to the sigmoid colon foci of red/purple (*white arrow*), **c** amyloid deposition in the vessel walls was demonstrated by apple-green birefringence under polarized light on Congo Red stain (*white arrows*), and **d** AA amyloid immunohistochemical stain showed the amyloid deposition in the vessel walls (*white arrows*)

the descending colon was made, and emergency surgery was planned.

Exploratory laparotomy revealed a huge bizarre mass lesion consistent with spreading-type of colonic tumor extending from the dilated descending to the sigmoid colon with serosal surface discoloration (foci of red/purple) possibly due to bouts of acute inflammation (Fig. 1b). Diffuse wall thickening with intense edema, and induration like phlegmonous infiltration was noticed at the involved colonic segment. The remaining parts of the colon were moderately dilated, and handled hardly. A Hartmann procedure was performed.

Histologic examination showed positive staining with Congo red dye, and the apple-green birefringence under polarized light and immunohistochemical findings were positive for amyloid A (AA) (Fig. 1c, d). Extensive laboratory investigations, physical examinations, and history taking did not provide any of the systemic disease. In order to rule out systemic disease, such as multiple myeloma or collagen disease, we performed some specific investigations. There were no plasma cell dyscrasias, serum and urine electrophoresis did not show monoclonal component, and serum autoimmune markers were negative.

Amyloid A finding in immunohistochemical analysis made us think that the case was a localized extensive colonic involvement of secondary amyloidosis.

After 7 days of recuperation, we discharged the patient without any further complication. We refrained from

administering any further drugs but decided to observe the patient intensely over the next 2 years. After a 2-year follow-up, there were neither signs of plasma dyscrasia nor of a systemic process.

Discussion

Amyloidosis can be acquired or hereditary, and systemic or localized. The common forms of systemic amyloidosis are primary (AL), secondary (reactive) (AA), dialysis-related, senile, and familial amyloidotic polyneuropathy.

The clinical diagnosis of gastrointestinal amyloidosis may be difficult in patients without previously known history of amyloidosis. Affected patients may present with nausea, vomiting, gastrointestinal bleeding, malabsorption, or rarely obstruction. In patients older than 30 years with unexplained diarrhea, weight loss, autonomic dysfunction, malabsorption, or proteinuria, amyloidosis has to be considered [4].

The combination of serum and urine immunoelectrophoresis increases the early yield of correct diagnosis [5]. Imaging reveals a normal bowel in most cases, but nonspecific changes such as bowel wall thickening or dilatation, or both may be seen [4]. Endoscopically, the findings of the gastrointestinal tract are nonspecific and include erythema, erosions/ulcerations, granular or plaque-like mucosa of the stomach or small

intestine, large duodenal folds, friability, prepyloric ulcers, and polypoid protrusions [6].

Histological evaluation is required for definitive diagnosis. Apple-green birefringence (when viewed with crossed-polarized light) and red staining (under white light microscopy) with Congo red are pathognomonic [7, 8].

Complications due to amyloidotic involvement at the gastrointestinal tract are uncommon and colonic obstruction is very rare. Here, we described a patient wherein we performed emergent laparotomy with a provisional diagnosis of large bowel obstruction secondary to a tumor in the descending colon. After the operation, histologic findings were positive for AA or secondary amyloidosis that was limited in the left colon. We performed all detailed investigations including plasma and urine electrophoresis, autoimmune markers, electrocardiography, and echocardiography to rule out a systemic process. We did not detect any evidence regarding systemic disease. Also, additional fat-pad biopsies after the operation were normal. Secondary (AA) amyloidosis may complicate a number of chronic inflammatory conditions, such as chronic degenerative diseases, chronic infections, or certain neoplasms. In this case, despite the fact that it could not be proved, we thought that a kind of chronic inflammatory condition—maybe idiopathic—caused secondary amyloidosis.

In conclusion, even if they are rare, it should be kept in mind that some unexpected pathologies such as amyloidotic involvement may lead to colonic obstruction, and endoscopic

biopsy can mislead the clinician for conservative approach. However, progressive disease with severe complications necessitates colonic resection.

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