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AA amyloidosis presented with ileus by forming a mass in the small intestine: A case report

İnce bağırsakta kitle oluşturarak ileus ile kendini gösteren AA amiloidoz: Bir olgu sunumu

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Abstract

Intestinal amyloidosis frequently encountered as a part of systemic amyloidosis, but rarely can be confined in the gastrointestinal tract. A 54-year-old male presented with the complaint of gas and stool discharge. Urgently segmental bowel resection was performed for ileus. Macroscopically nodular lesions, the largest at 7x3x0.7 cm in size were observed in the intestinal lumen. Microscopically; the accumulation of dense eosinophilic material that formed a mass in the submucosal area was noted. This material was positive with Crystal Violet, Congo Red and Amyloid A. Kappa and Lambda were negative. No monoclonal gammopathy, increase in serum amyloid A levels, chronic inflammatory disease, infectious disease or malignancy was determined. The case was evaluated as "intestinal AA amyloidosis". While AA amyloidosis was existent in our case, it comprised a mass lesion and caused intestinal obstruction. It is also exraordinary for AA amyloidosis to be confined in gastrointestinal tract.

Keywords: amyloidosis; intestinal amyloidosis; ileus; small intestine; intestinal mass

Öz

İntestinal amiloidoza sistemik amiloidozun bir parçası olarak sıklıkla rastlanır, ancak nadiren gastrointestinal kanalda sınırlı olabilir. 54 yaşında erkek hasta gaz ve gaita çıkaramama şikayeti ile başvurdu. İleus nedeniyle acil olarak segmental bağırsak rezeksiyonu yapıldı. Makroskopik olarak bağırsak lümeninde en büyüğü 7x3x0.7 cm boyutlarında nodüler lezyonlar gözlendi. Mikroskobik olarak; submukozal alanda kitle oluşturan yoğun eozinofilik materyal birikimi kaydedildi. Bu materyal Crystal Violet, Congo Red ve Amiloid A ile pozitifti. Kappa ve Lambda negatifti. Monoklonal gamopati, serum amiloid A düzeylerinde artış, kronik inflamatuar hastalık, enfeksiyöz hastalık veya malignite saptanmadı. Olgu "intestinal AA amiloidoz" olarak değerlendirildi. Olgumuzda AA amiloidoz mevcut iken, kitle lezyonu oluşturmuş ve intestinal obstrüksiyona neden olmuştur. AA amiloidozun gastrointestinal kanalda sınırlı olması da olağan dışıdır.

Anahtar kelimeler: amiloidoz; intestinal amiloidoz; ileus; ince bağırsak; bağırsak kitlesi

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Introduction

Amyloidosis is a rare disease group that is characteristically express extracellular fibrillar protein deposition in various tissue. Although many amyloidogenic proteins have been identified to date, amyloidosis is classified into four groups. Immunoglobulin light-chain (AL) amyloidosis (primary amyloidosis) is a plasma cell disorder and the most common type of amyloidosis. Serum amyloid A (AA) amyloidosis (secondary amyloidosis) is associated with chronic inflammatory disease, infections and malignancies. ß2-microglobulin amyloidosis is associated with dialysis and is the type of amyloidosis that seen in end-stage renal disease patients. TTR (ATTR) accumulation occurs in hereditary amyloidosis and in senile systemic amyloidosis [1, 2].

Amyloidosis can be localized or systemic. Involvement of various organs may be seen in systemic amyloidosis. Cardiac, dermal, renal and peripheral neurological involvement is common [3]. Gastrointestinal tract is also frequently involved in amyloidosis and the most commonly affected gastrointestinal site is the small intestine. Although amyloidosis is rarely seen locally in the gastrointestinal tract, it is frequently encountered as a part of systemic amyloidosis [4, 5].

The presentation in amyloidosis is variable and can vary depending on the organ involved and the degree of involvement. Amyloid can accumulate in many tissues such as muscles, vessels, nerves in the intestines. AL amyloidosis is usually accumulate in the gastrointestinal tract in submucosal area, muscularis mucosa and muscularis propria. This accumulation usually cause polypoid masses and myopathy and these patients often presented with constipation, obstruction or pseudo-obstruction. On the contrary, AA amyloidosis tends to involve the lamina propria and present with ulceration, bleeding or malabsorption [6].

Here in, we present a case of AA amyloidosis presented with ileus by forming a mass in the small intestine.

Case report

A 54-year-old male presented with the complaint of gas and stool discharge. On physical examination, the abdomen was distended, bowel sounds could not be detected, and widespread abdominal tenderness was appointed. Rectal examination revealed empty rectum and no mass was detected. Computed tomography images revealed dilatation in the stomach, duodenum and jejunum, as well as air-fluid levels in the jejunum. Dilatation was not observed in the ileum and colon. The patient was urgently operated for ileus; a mass lesion that causing obstruction in the jejunum was observed. When segmental bowel resection was performed to excise the mass, it was noteworthy that there were >100 nodular lesions in the lumen of the remaining intestinal tissue. An isoperistaltic ileoileostomy anastomosis was performed side by side from the relatively solid area. In the macroscopic examination of the 13 cm long small bowel resection material; a nodular lesion, 7x3x0.7 cm in size, covered with ulcerated mucosa was observed in the intestinal lumen. In addition, a 2.5x2x1.5 cm polypoid lesion and a nodular lesion of 3x1.5x0.5 cm in size were observed (Figure 1). Microscopically; the mucosa was ulcerated, and the accumulation of dense eosinophilic material that formed a mass in the submucosal area was noted. It was observed that the same material was deposited on the vessel walls and muscularis propria. Positive staining was determined with Crystal Violet and Congo Red stains in this material (Figure 2). While Amyloid A stain was positive immunohistochemically, Kappa and Lambda light chains were negative. The case was pathologically reported as AA amyloidosis.

Subsequent esophagogastroduodenoscopic examination revealed mild edema in the antrum, nodularity and polypoid lesions in the duodenum. No pathological findings were found in colonoscopic examination. Amyloidosis was detected in the antrum and duodenum biopsies on histopathological examination, and no amyloidosis was detected in the colon. Plasma cell increase and amyloid accumulation was not detected in the bone marrow biopsy, plasma cells were observed at a rate of 5% and these cells were found to be polyclonal. No monoclonal gammopathy was detected in the serum immunelectrophoresis study. There was no increase in serum amyloid A levels. No chronic inflammatory disease, infectious disease, Crohn's disease, familial mediterranean fever or malignancy was determined. With these findings, the case was evaluated as "intestinal AA amyloidosis".

A written consent was obtained from the patient for the publication of this case report and accompanying images.

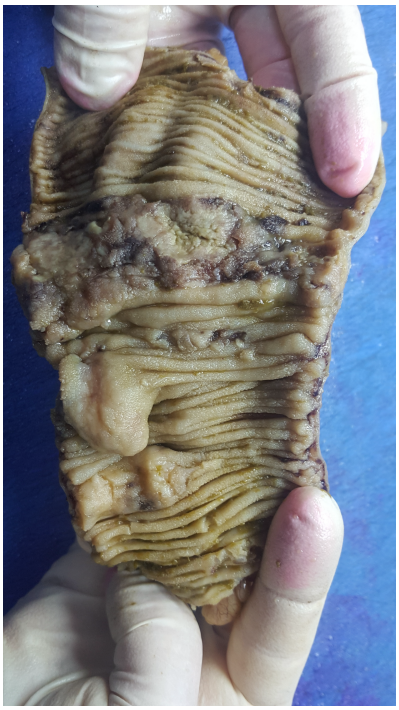

Figure 1: A nodular lesion, 7x3x0.7 cm in size, covered with ulcerated mucosa, a 2.5x2x1.5 cm polypoid lesion and a nodular lesion of 3x1.5x0.5 cm in size were observed in the small bowel lumen.

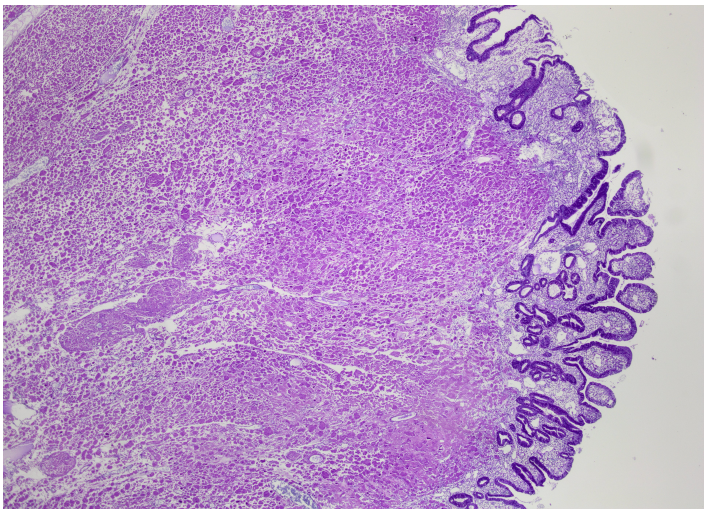

Figure 2: Accumulation of dense material that formed a mass in the submucosal area in the small bowel. Positive staining with Crystal Violet in this material. Crystal Violet x 100.

Discussion

The clinical manifestation of amyloidosis can range from asymptomatic to fatal disease. Intestinal amyloidosis can cause a wide variety of symptoms by causing both structural and functional impairment in the organ where it deposits. Symptoms are usually nonspecific and weight loss, abdominal pain, diarrhea, malabsorption, lower and upper gastrointestinal tract bleeding are common symptoms. The severity of these symptoms is related to the amount and extent of amyloid deposition [3].

Endoscopic findings are also nonspecific as well as clinical findings of intestinal amyloidosis. Erythema, granular appearance, erosion, ulceration, friability, polypoid protrusion, submucosal masses can be observed. Endoscopic findings may be normal in intestinal amyloidosis. Localized amyloidosis in the small intestine is characterized by focal amyloid deposits that form a mass and cover the entire intestinal wall. These are called amyloidomas and are seen as a polioid mass lesion or an ulcerated lesion, as in our case [7].

The diagnosis of amyloidosis is difficult and can be missed because of its rarity, nonspecific symptoms and nonspecific endoscopic findings. Although gastroenterologists and endoscopists play critical role in diagnosis of amyloidosis, histopathological demonstration is necessary for definitive diagnosis. In differentiating amyloid from other protein deposits, Congo red is the most specific stain. Crystal Violet is another histochemical stain demonstrating amyloid. Immunohistochemical Amyloid A, Lambda and Kappa stains allow us to differentiate the type of amyloidosis besides showing that the accumulation is amyloid. While amyloid A positivity indicates AA amyloidosis; Lambda or Kappa positivity supports AL amyloidosis [8].

Amyloid can accumulate in all layers and structures of the intestine and may lead to different conditions depending on the structure in which it accumulates. Deposition in the muscularis propria and myenteric plexus can lead to myopathy and neuropathy and causes atrophy, diarrhea and dysmotility. Deposition in vascular structures can lead to ischemia, infarction, ulceration and bleeding [7]. The localization and presentation of amyloid deposition in gastrointestinal tract depends on the type of amyloid. Generally in AL amyloidosis, amyloid deposition occurs in the submucosal area and in the muscularis propria; it may cause a mass lesion, present with constipation, mechanical obstruction or pseudo-obstruction. In AA amyloidosis, the accumulation is mostly seen in the lamina propria, mucosa and submucosal area in macular form and manifests with granular appearance and friability of the mucosa. AA amyloidosis often causes clinical signs of diarrhea and malabsorption [9, 10]. Although our case has AA amyloidosis; it was noteworthy that amyloid was deposited in the submucosal area, muscularis proria and vessel walls, formed a polypoid submucosal mass and presented with obstruction findings.

Treatment of AA amyloidosis is carried out by treating the underlying disease, such as autoimmune disease, infections or malignancy that causes an increase in amyloid precursors. In addition, symptomatic treatment based on alleviation of complaints may be useful in amyloidosis. Cases with malabsorption can be suplemented by nutritonsand vitamins. Surgical treatment can be applied in cases with bleeding or obstruction. Since we did not determine an underlying disease and didn't detect an increase in serum amyloid A level in our case, our patient did not receive additional treatment after surgery.

Although amyloid accumulation is common in the gastrointestinal tract as a part of systemic amyloidosis, it is rare for amyloidosis to be confined in the gastrointestinal tract [11]. Amyloidosis is a chronic disease and rarely requires immediate surgical treatment.

It is interesting that while AA amyloidosis was existent in our case, it comprised a mass lesion, caused intestinal obstruction, and required urgent surgical treatment. It is also exraordinary for AA amyloidosis to be confined in the gastrointestinal tract.

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