Case Report

Jejunojejunal Intussusception Causing Small Intestine Obstruction as the Single Presentation of Amyloidosis: A Uniqueness Report

Amirhossein Hosseini[1], Mehrnoush Hassas Yeganeh[2*], Reza Sinaei[3], Hamid Hosseinzadeh[4], Naghmeh Sharifi[5], Shiva Khoobyari[6]

[1] Pediatric Gastroenterology, Hepatology and Nutrition Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran.

[2] Shahid Beheshti University of Medical Sciences, Tehran, Iran.

[3] Department of Pediatrics, School of Medicine, Kerman University of Medical Sciences, Kerman, Iran; Clinical Research Development Unit, Afzalipour Hospital, Kerman University of Medical Sciences, Kerman, Iran.

[4] Rowan University, Glassboro, NJ, USA.

[5] Mofid Children Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

[6] Department of Pathology, University of Washington, Seattle, WA.

ABSTRACT

A group of diseases resulting from the abnormal aggregation of amyloid fibrils, mainly in the extracellular spaces of tissues, is known collectively as amyloidosis. Clinical presentations of gastrointestinal (GI) amyloidosis are mainly GI bleeding, malabsorption, protein-losing enteropathy, and dysmotility. Here, we describe an 87-year-old woman presenting with acute onset of sharp, constant, and non-radiating epigastric pain that was accompanied by nausea or vomiting. After a thorough investigation, we finally found an isolated jejunojejunal intussusception by laparotomy. The pathological study revealed massive small bowel involvement by amyloidosis in immunostaining. To our knowledge, this is the first report of small intestinal obstruction resulting from intussusception due to intraluminal amyloid polypoid nodules, as the first presentation of light-chain amyloidosis. Amyloidosis should be considered in old patients with intraluminal masses of the proximal small bowel.

Keywords: Jejunum, Intussusception, Obstruction, Amyloidosis

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Corresponding author:
Mehrnoush Hassas Yeganeh, MD
Shahid Beheshti University of Medical Sciences, Tehran, Iran.
Tel: + 98 09133401146
Fax: + 98 3433257470
Email: Me.yeganeh13@gmail.com

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BACKGROUND

Amyloidosis is a collective term that refers to a group of diseases caused by the deposition of misfolded amyloid fibrils, mainly in the extracellular spaces of tissues, leading to organ dysfunction or failure (1). The pathological deposition of amyloid proteins in almost any body organ can interfere with the structure and function of tissues and can lead to permanent organ damage and death (2).

Gastrointestinal (GI) tract amyloidosis is not common, and when present, the disease is usually diffuse, with other organs involved. Very uncommonly but well documented, there is the isolated, localized presentation of amyloidosis in the GI tract, characterized by the absence of systemic involvement. The reported cases most commonly involve the esophagus, the colon, and the rectum (3). However, the least common part of the GI system involved is jejunum/ileum (%5.9 of most case series) (4).

We describe a patient with small bowel obstruction secondary to jejunojejunal intussusception as the only presentation of amyloidosis that was diagnosed on an abdominal computed tomography (CT) scan, which we believe is the first reported case in the world.

CASE REPORT

An 87-year-old woman with a medical history of laparoscopic cholecystectomy was admitted to the emergency department with signs and symptoms of bowel obstruction. She presented with an acute onset of sharp, constant, and non-radiating epigastric pain that was accompanied by nausea or vomiting. She was afebrile with stable vital signs. Findings on physical examination were unremarkable. Laboratory studies were within the normal limits, including complete blood count, serum electrolytes, liver function tests, and coagulation studies. A CT examination of the abdomen and pelvis with intravenous (IV) contrast material (Omnipaque 350 mg/mL 96@. 4 mL) was performed on admission and revealed several areas of dilated, fluid-filled, and enhancing small bowel loops with adjacent fat stranding and free fluid. A segment of the decompressed small bowel, which appears slightly thick-walled, was seen at the zone of transition. With a pre-operative impression of bowel obstruction, she underwent exploratory laparotomy, which showed small bowel obstruction secondary to intussusception from the intra-luminal jejunal mass. Other positive findings during surgery were a moderately dilated small bowel with multiple intra-luminal masses, a short segment of soft but markedly thickened jejunum, and palpably enlarged lymph nodes in the adjacent mesentery. A segment of about 30 cm of jejunum was resected, and primary anastomosis was done. Gross pathological examination revealed a segment of small bowel measuring 33 cm in length × 4 cm (proximal margin) × 5 cm (distal margin). The serosal surface showed patchy areas of hemorrhagic dusky discoloration. The specimen was opened along the antimesenteric side, revealing multiple hemorrhagic, partially necrotic polypoid nodules ranging in size from 0.4 × 0.5 × 0.2 cm up to 5.5 × 4.2 × 1.2 cm. The most distal nodule was located approximately 0.3 cm away from the distal margin. Grossly, the nodules were not penetrating through the serosa. Both ends of the specimen showed a normal gross appearance.

Microscopic examination showed massive small bowel involvement by amyloidosis, involving mostly submucosa but extending to muscularis propria, forming polypoid nodules (the largest was 5.5 cm) (figure 1). The amyloid nature of the deposits was confirmed by crystal violet and Congo red stains with "apple green" birefringence (figure 2). Immunostaining for amyloid-A revealed patchy positivity within small areas of the deposits. The diagnosis of a localized, pseudo-tumoral form of jejunal amyloidosis was proposed. Clinical improvement was noted with subsequent discharge from the hospital. With the final diagnosis of primary amyloidosis, she was referred for follow-up with the diagnosis.

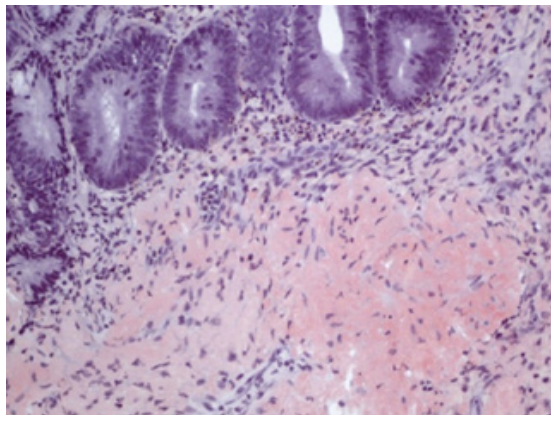

Figure 1. Jejunal specimen revealing extensive amyloid deposition of amorphous, salmon-pink, sparsely cellular, hyaline material within the submucosa and focally within the lamina propria (hematoxylin counterstain, original magnification ×200)

DISCUSSION

Delays in the diagnosis of amyloidosis as a complex disease are associated with significant morbidity and mortality (2). Clinical manifestations of amyloidosis are diverse and non-specific (5,6).

Clinical presentations of GI amyloidosis are mainly GI bleeding, malabsorption, protein-losing gastroenteropathy, and gastrointestinal dysmotility (7). Gastrointestinal

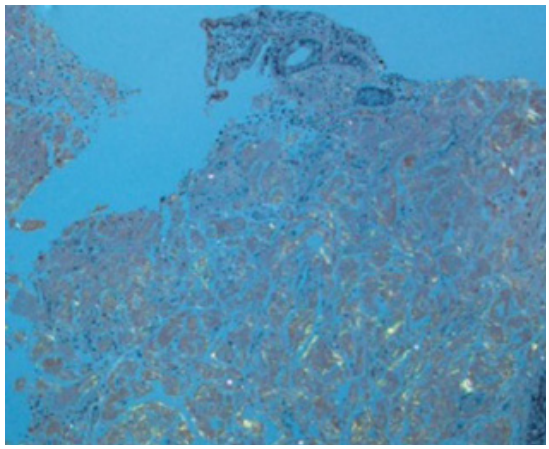

Figure 2. The apple-green color birefringence of the deposits using polarizing microscopy (×100).

dysmotility presents most commonly as nausea, vomiting, dysphagia, gastroparesis, gastroesophageal reflux, constipation, or even pseudo-obstruction (8).

While patients with amyloidosis AA usually present with diarrhea and malabsorption, patients with AL amyloidosis often present with constipation, mechanical obstruction, or chronic intestinal pseudo-obstruction (9).

These manifestations can happen in the course of systemic amyloidosis. However, very rarely, they can be the first presentation of amyloidosis (10).

As the presenting manifestation, there is only one reported case of true obstruction of the small bowel in amyloidosis. Jones and colleagues reported a case of obstruction due to small bowel encasement with a thick membrane in a previously healthy 62-year-old man. At surgery, they discovered a peculiar membrane encasing the entire small bowel. Histopathology revealed thick bands of collagen on the peritoneal surface, which was Congo red positive and showed apple-green birefringence. The findings were matched with encapsulating peritonitis due to amyloidosis. The patient later turned to show the lab findings consistent with amyloidosis (11).

Our patient presents with partial small bowel obstruction. Areas of dilated, fluid-filled, and enhancing small bowel loops were detected in her abdominal CTs. The patient was treated surgically by resection-primary anastomosis of the affected area of the jejunum due to the emergent nature of the obstruction. Following surgery, the patient became symptom-free. With the microscopic and histochemical confirmation of the diagnosis of amyloidosis, she was referred for follow-up with amyloidosis.

Endoscopic findings are variable based on the organ and the depositing amyloid. The duodenal specimens may contain a fine granular appearance or polypoid protrusions. Deposition of AL, β2-microglobulin-related (Aβ2M) amyloid, and Transthyretin amyloid (ATTR) occur submucosally, while AA amyloid is easily detected in the superficial layer of the mucous membrane (12). Histopathological investigation in our patient revealed the involvement of the small bowel by massive amyloidosis, mostly submucosa, but also extending to muscularis propria.

To our knowledge, this is the first report of small intestinal obstruction resulting from intussusception due to intraluminal amyloid polypoid nodules, as the first presentation of light-chain amyloidosis.

In conclusion, we believe that although a rare entity, in any old patient with small intestine obstruction, amyloidosis has to be considered in the differential diagnosis. Besides, amyloidosis should also be considered in old patients with intraluminal masses of the proximal small bowel. Histological evaluation is necessary to confirm the diagnosis of amyloidosis.

Ethics approval and consent to participate

This study protocol was approved by the Ethics Committee of Shahid Beheshti University of Medical Sciences, Tehran, Iran (ID: IR.SBMU.MSP.REC.1397.788).

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