CT and MRI Findings of Small Bowel Involvement of Amyloidosis Mimicking Small Bowel Polyposis Syndrome: a Case Report

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INTRODUCTION

Amyloidosis is an all-inclusive disease of deposition of amyloid proteins in the extracellular spaces, which in localized or systemic form cause tissue damage and dysfunction. Herein, we report a case of small bowel involvement of systemic amyloidosis presenting with multiple polypoid wall thickening mimicking small bowel polyposis syndrome in an age 75 male. Interestingly, polypoid wall thickening and amyloidoma showed hypointensity on T2-weighted images. To our knowledge, there has been no literature describing MRI findings of poylpoid wall thickening and amyloidoma. Although the underlying mechanisms are unclear and need validation, hypointensity on T2-weighted images could be valuable in diagnosing small bowel involvement of amyloidosis in patients presenting with poylpoid wall thickening and amyloidoma.

Keywords: Small bowel; Intestinal; Amyloidosis; Amyloidoma; Computed tomography; Magnetic resonance imaging

CASE REPORT

An age 75 male with anemia presented to our hospital. He had no previous significant
medical or family history, except for hypertension. He underwent gastrointestinal endoscopy, which revealed multiple polypoid lesions in the duodenum (Fig. 1a). Endoscopic biopsy results demonstrated non-specific severe duodenitis and chronic gastritis. Subsequently, he underwent contrast-enhanced abdominal computed tomography (CT), revealing widespread polypoid wall thickening in the duodenum and entire small intestine. And in the jejunum, an annular mass was also evident (Fig. 1b), mimicking small bowel polyposis syndrome and possible development of small bowel cancer. Additionally, small bowel follow through was performed, revealing impaired motor activity, fold thickening, and polypoid protrusions (Fig. 1c). The patient refused further evaluation and management thereafter.

Eight years later, however, he visited the emergency department from suspected acute cholecystitis. He underwent contrast-enhanced abdominal CT and magnetic resonance cholangiopancreatography, and was treated with percutaneous cholecystostomy. Duodenoscopy and CT revealed increased size and number of polypoid lesions with progressed calcifications (Fig. 2a, b). Magnetic resonance imaging (MRI) was performed with a 3.0-T MRI system. T1- and T2-weighted images obtained by fast field echo in-phase and out-of-phase sequence and single shot turbo spin echo sequence. On MRI, the polypoid lesions
exhibited hypointensity on T2-weighted images relative to the unaffected bowel wall and isointensity on T1-weighted images (Fig. 2c-f). Duodenoscopic biopsy results demonstrated eosinophilic amorphous material deposition in the duodenum with positive Congo red stain, consistent with amyloidosis (Fig. 2g, h). Protein electrophoresis, immunofixation electrophoresis, and bone marrow biopsy revealed immunoglobulin λ light-chain-positive monoclonal gammopathy, and a final diagnosis of systemic amyloid light-chain (AL) amyloidosis was confirmed. Because of the patient’s advanced age, chemotherapy was not performed and conservative management was initiated.

**DISCUSSION**

In the literature, presently 36 amyloid proteins are known of which 14 proteins only related with systemic amyloidosis and 19 proteins only related with localized forms, and three proteins related with localized and systemic amyloidosis (1). Among these, AL is one of three proteins that can development systemic and localized form,

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**Fig. 2.** Eight years later. (a) The duodenoscopy image showed increased size and number of multiple polypoid lesions. (b) The axial CT scan showed dense progressed calcification (arrow) in the focal annular mass. (c) The coronal T2-weighted image showed multiple polypoid wall thickening showing hypointensity (arrows). (d-f) Axial T2- (d), in-phase T1- (e), and out-of-phase T1-weighted (f) images showed the amyloidoma as T2 hypointensity and T1 isointensity (arrows). The focal T1 and T2 hyperintensity (arrowheads) were caused by dense calcification with microscopic fat showing signal drop on out-of-phase T1-weighted image.
and AL amyloidosis is the secondary disease to plasma cell dyscrasias and approximately 10% of patients are related with multiple myeloma (6). In systemic amyloidosis, the most commonly affected system is the gastrointestinal tract, especially the small intestine (2, 7).

In the barium study, the most common finding is symmetrical fold thickening from edema caused by vascular deposition of the amyloids and ischemia. Impaired motor activity, fine granular densities, jejunalization of the ileum, polypoid protrusions, and amyloid tumor are also known (2). Although pathological correlation was not investigated, the polypoid protrusions and fold thickening in our case of AL amyloidosis corresponded with the previous study (8), reporting that amyloid deposition at muscularis mucosa and submucosa in AL amyloidosis.

The CT findings of small intestinal amyloidosis are non-specific and diverse. It includes bowel wall thickening, dilatation, focal amyloidoma, and mesenteric infiltration (2-5). Although there was no mention of polypoid wall thickening in the previous studies, it seems to be a manifestation of the polypoid protrusion on barium study.

Interestingly, polypoid wall thickening and amyloidoma demonstrate hypointensity on T2-weighted images relative to the unaffected bowel wall and isointensity on T1-weighted images. To our knowledge, there has been no literature describing MRI findings of polypoid wall

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Fig. 2. (g) The photomicrograph (x 50, Hematoxylin & Eosin stain) showed abundant amorphous eosinophilic material deposits. (h) The photomicrograph (x 100, Congo red stain) showed amyloid deposit as red to pink (arrows).
thickening, and amyloidoma. Based on the kinetics of fibril formation, known as nucleated growth like of crystallization in in vitro studies (9), polypoid wall thickening and amyloidoma are expected to be exhibit densely aggregated protein deposition. So, we hypothesize that hypointensity on T2-weighted imaging may be attributable to high protein concentrations. However, there are many factors that should also be considered, including edema, macrophage, fibrosis, calcification, iron, and other unknown mechanisms. Thus, the mechanism of hypointensity on T2-weighted images is unclear and further studies are needed.

Conversely, previous studies (3, 4) reported MRI findings of small intestinal amyloidosis that demonstrated a diffuse wall thickening pattern, not polypoid wall thickening or amyloidoma. They reported diffuse wall thickening of the small intestine without mention of T2 signal intensity that resembles isointensity. Although morphological features on MRI are beneficial in evaluating small intestinal amyloidosis, T2 signal intensity is inconspicuous in the case of diffuse wall thickening patterns.

Generally, the differential diagnosis of small intestinal amyloidosis is diverse according to radiological findings, including infectious enterocolitis, bowel ischemia, other infiltrating diseases, such as small bowel lymphoma, and even adenocarcinoma, when focal amyloidoma was observed. In our case, multiple polypoid wall thickening mimics small bowel polyposis syndrome, such as Peutz-Jeghers syndrome. Clinical factors, such as family history, age, colonic polyposis, and mucocutaneous pigmentation, are fundamental in the differential diagnosis of small bowel polyposis syndrome. Also, adenomatous or hamartomatous polyps are likely to exhibit isointensity or hyperintensity on T2-weighted images relative to the bowel wall, hypointensity on T2-weighted images could be valuable in the differential diagnosis of polyposis syndrome.

In conclusion, we report a case of small bowel involvement of systemic AL amyloidosis presenting with multiple polypoid wall thickening mimicking small bowel polyposis syndrome. Although the underlying mechanisms are unclear and need validation, hypointensity on T2-weighted images could be valuable in diagnosing small bowel involvement of amyloidosis in patients presenting with polypoid wall thickening and amyloidoma.

REFERENCES