

Localized Amyloidosis of Colon- A Rare Case Report

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Abstract: Amyloidosis of the Gastrointestinal tract with biopsy proven disease is rare. We present the clinical and histopathological features of localised intestinal amyloidosis with a rare case report.

The patient had nonspecific gastrointestinal symptoms including haematochezia. Prior to treatment, the patient was suspected of having colo-colic intussusception on the basis of CT scan findings. The patient was treated with right hemicolectomy for caecal mass. The postoperative pathological diagnosis determined the lesion to be deposition of amyloid material with no evidence of malignancy. It was confirmed on congo red staining.

Localised gastrointestinal amyloidosis is rare in incidence, but it should be considered in differential diagnosis of gastrointestinal tumours and confirmation can be done on biopsy. Although the condition is benign, there is a tendency of recurrence as suggested by literature.

Keywords: Amyloidosis, colon, intestine, mass

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I. Introduction

Amyloidosis is a group of disorders in which amyloid protein, known as amyloid fibrils build up in tissue. It commonly shows a systemic involvement. Localised deposition of amyloid is a rather uncommon form, and amyloid deposit confined to the intestine is extremely rare.

II. Case Report

A 45 year old female was admitted to general surgery department of our hospital with one month history of pain in lower abdomen, diarrhoea and haematochezia. She had undergone hysterectomy 10 years back, and recovery was uneventful. She is G2P2A0. Family history was not significant. On general examination, there was no abnormality found. Per rectal examination was normal. Chest and abdominal x ray were normal. On ultrasonography and Computed tomography scan, colo-colic intussusception was suspected involving 10 cm of ascending colon. Patient underwent exploratory laparotomy and caecal mass was found, and right hemicolectomy was done. On histopathological examination, deposition of amorphous amphophilic eosinophilic material ? amyloid, suggestive of intestinal amyloidosis. It was confirmed with congo red staining.

III. Discussion

Amyloidosis is a disorder characterized by extracellular deposition of amyloid in various tissues and organs. As the unique feature of amyloid substance was, the component of the precursor protein that forms the fibrillar deposit has been now accepted as the basis for the classification of amyloidosis^[1]. Up to the present, several types of the precursor proteins such as primary or light chain (L)-associated AL amyloidosis, secondary amyloidosis with acute-phase reactant serum amyloid A protein (AA amyloidosis), familial amyloidosis (ATTR amyloid), hemodialysis-associated amyloidosis (A β 2 amyloid), senile and localized amyloidosis. Secondary amyloidosis is associated with infectious, inflammatory, or less commonly, neoplastic disorders and renal dysfunction is the most common symptom of AA amyloidosis at diagnosis.

Amyloidosis of the gastrointestinal tract with biopsy proven disease is rare. A retrospective study has been conducted in past which reviewed 2334 patients with all forms of amyloidosis evaluated over 13 years and showed gastrointestinal involvement in 3.2% cases (76 patients). Biopsy proven patients of gastrointestinal amyloidosis present with diarrhoea, weight loss, gastro-paresis, malabsorption, intestinal pseudo-obstruction, and bleeding^[2,3]. The walls of the submucosal blood vessels are the most frequent sites of amyloid deposits, and diffuse deposits may result in impaired motility, malabsorption, and ulceration because of the related ischemia^[4]. The clinical manifestation of colonic amyloidosis may be similar with other colonic diseases, such as inflammatory bowel disease, ischemic colitis, collagenous colitis, and malignancy^[5,6]. The absence of systemic symptoms may make diagnosis difficult.

. It is important in this situation to determine whether amyloidosis is local versus systemic as the management and the prognosis are quite different^[7]

Endoscopic findings such as fine granular appearance and polypoid protrusions are common and may reflect amyloid deposition in the mucosa or submucosa of the alimentary tract^[8].

Radiologic findings of abdominal involvement in systemic amyloidosis are nonspecific and can present in a variety of ways. Heterogeneous appearance of liver, periportal involvement, diffuse low signal intensity of spleen on T2-weighted images and thickened bowel wall can be helpful if accompanied by history of chronic inflammatory disease and clinical suspicion for amyloidosis^[9]

Patients with amyloidosis have been treated by IV hyperalimentation; by administration of corticosteroids, immunosuppressive agents, dimethyl sulfoxide, or cochlincine; or by surgery. Because no specific therapy is available, it is difficult to select the most effective therapeutic regimen. Effective preventive treatment against localized amyloidosis deposits in the gastrointestinal tract is also uncertain. The affected mucosa is fragile and tends to bleed, which can cause suture failure; therefore, it has been suggested that surgery should be limited to extreme emergencies, such as in patients with massive bleeding, a perforation, or an obstruction^[10].

IV. Conclusion

Intestinal amyloidosis should be considered in patients with unresponsive to conventional treatment because its clinical manifestation is variable and non-specific.

Acknowledgements

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FIGURES

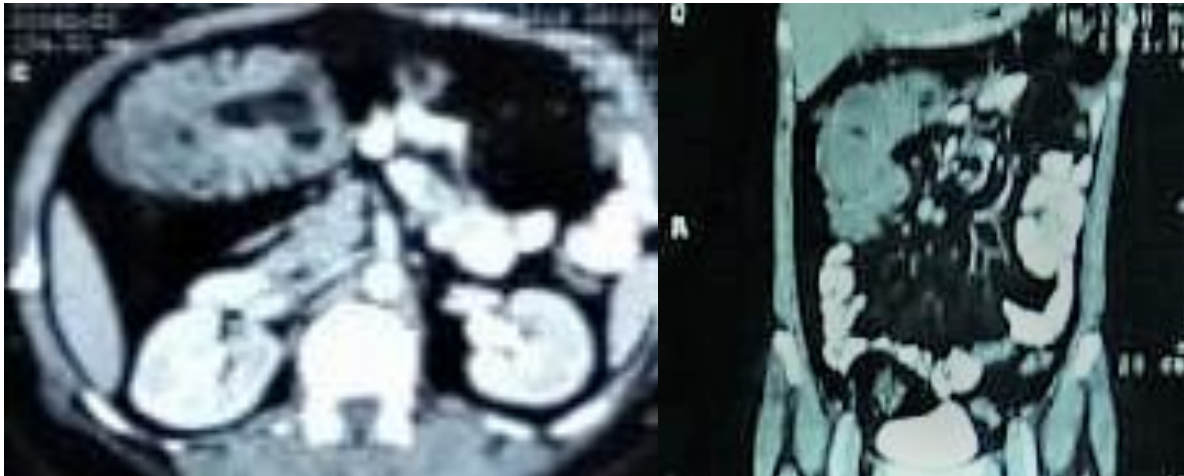


FIGURE 1: Axial and Coronal CECT: showing diffuse circumferential thickening of ascending colon and hepatic flexure appearing as colonic mass with features of intussusception.

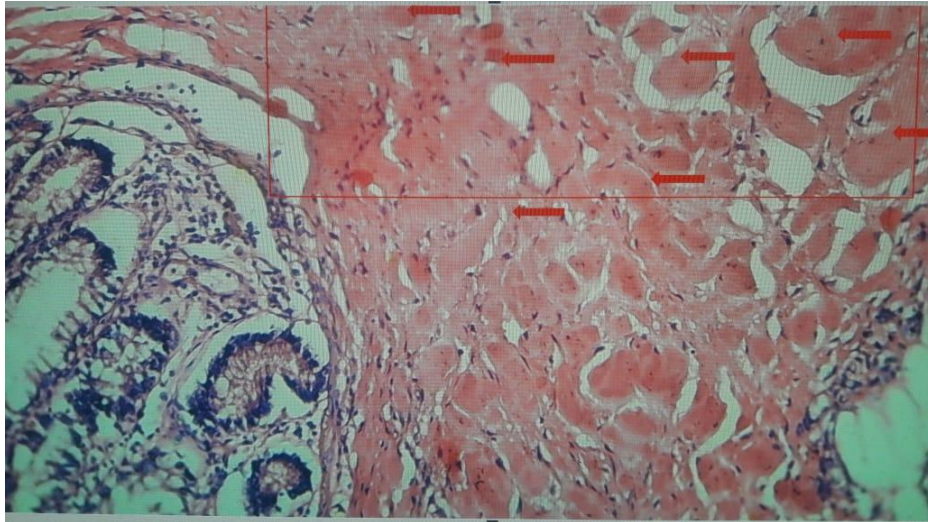


FIGURE 2: Congo red stain of colonic mucosa showing deposition of amorphous eosinophilic material in lamina propria and positive staining with congo red.

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