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None of the authors are recipients of a research scholarship. This paper is not based on previous communication to a society or meeting. It is a case report of a patient clinical presentation in which the authors were involved in the surgical care and management of the patient discussed. The patient discussed has provided consent for the publication of their case.

Word Count = 750 words + references

This case report includes: 3 images attached

Amyloidosis is a rare disorder characterised by the extracellular deposition of abnormal, insoluble peptides and fibrillar proteins<sup>(1),(2)</sup>. Nearly every organ can be affected, including the gastrointestinal tract and the clinical

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symptoms of gastrointestinal amyloidosis are varied making the clinical diagnosis challenging<sup>(1),(3),(4),(5)</sup>. We report a case of small bowel amyloidosis causing partial small bowel obstruction which was initially misdiagnosed as stricturing crohn's disease.

A 60 year old woman with a past history of rheumatoid arthritis, osteoarthritis and coeliac disease presented with 24hr history of loose bowel motions and abdominal pain. They had been diagnosed with crohn's disease 3 months prior, following a similar presentation which was treated and responded clinically to high dose steroids. This diagnosis was made on the basis of CT enterography which showed a short segment of concentric bowel wall thickening in the distal ileum, suggestive of crohn's stricture (Figure 1), and pillcam imaging suggestive of the same pathology. Colonoscopy up to the terminal ileum was normal. Following a discussion at the gastroenterology MDT, a diagnosis of crohn's disease was made.

The patient was initially treated as an acute crohn's flare with hydrocortisone and whilst improving initially, after 48hrs had increasing abdominal pain, distention and bilious vomiting. CT scan undertaken at this time (Figure 2) demonstrated an incomplete small bowel obstruction with a transition point at the distal ileal stricture which had progressed from the CT enterography performed 3 months prior.

The patient underwent a laparotomy and small bowel resection with formation of a double barrel end ileostomy. Intraoperative findings revealed a short segment of ileum which was resected, approximately 60cm from the terminal ileum. There was evidence of small bowel distension upstream to this but there were no further lesions seen. In view of the concern of malnutrition and prolonged symptoms prior to presentation, a stoma was formed. The patient had an unremarkable postoperative course and was discharged day 6 post operatively. She continues to have follow up treatment with Rheumatology.

Histopathology from the resected ileum showed evidence of ulceration, associated local chronic active mucosal inflammation and transmural thickening by semiconfluent fissured, nodular deposits of hyaline material within the submucosa and muscularis propria. Congo red stained sections showed the dense hyaline material stained bright orange, with green birefringence under cross polarisation, confirming the classical appearance of amyloid (Figure 3). Amongst the deposits there was also a nodular transmural inflammatory infiltrate which focally

extended out into the mesentery. The infiltrate consisted principally of mature lymphocytes with lesser numbers of plasma cells and small numbers of larger lymphoid cells. There was only focal minor associated mural fibrosis and the macroscopic stricture appeared to be entirely consequent to the massive amyloid deposits, which had completely obliterated the muscularis propria in the centre of the stricture.

The amyloid showed selective staining for kappa light chains by immunohistochemistry. In addition, the lymphocytic infiltrate showed monoclonal expression of kappa light chain mRNA by ISH and had an immunophenotype consistent with an extranodal marginal zone lymphoma.

Amyloidosis is a rare disorder with an incidence between 5 and 13 per million persons per year<sup>(2)</sup>. There are a number of different subtypes of amyloidosis, classified by the precursor proteins involved in protein deposition.<sup>(6)</sup>

Primary amyloidosis known as AL amyloidosis caused by light chain immunoglobulins, is the most common form of amyloidosis and is associated with haematological malignancies<sup>(7)</sup>. In secondary amyloidosis, known as AA amyloidosis, there is evidence of coexisting disease leading to the production of serum amyloid A protein, an acute phase reactant found in infectious and chronic inflammatory diseases such as rheumatoid arthritis<sup>(7),(5),(8)</sup>.

The gastrointestinal tract can be affected in 60% of patients with AA amyloidosis, while only 1–8% of patients with amyloidosis caused by light chain immunoglobulins (AL amyloidosis) have gastrointestinal involvement<sup>(8)</sup>. The small bowel is the most commonly affected site of amyloid deposition in the gastrointestinal tract<sup>(5)</sup>.

Clinical symptoms vary depending on the layer of bowel wall involved. Mucosa predominant amyloid deposition can present with diarrhoea and malabsorption, whereas muscularis predominant disease can present with obstruction<sup>(1),(9)</sup>. Radiological findings in gastrointestinal amyloidosis are non-specific and may include small-bowel dilatation, symmetric bowel wall thickening, mesenteric infiltration, and mesenteric adenopathy<sup>(10)</sup>.

The diagnosis of amyloidosis is based on congo red staining of the histological specimen which produces the characteristic orange-red appearance in normal light and classic green birefringence under polarised light<sup>(3)</sup>.

Our case illustrates that amyloidosis of the small bowel can mimic Crohn's disease in its presentation. It highlights the need to consider other causes of small bowel pathology in the context of patients who present with non-typical location within the small bowel and in the presence of other autoimmune conditions. Patients with small bowel lesions that progress over a short period of time and fail to respond to non-surgical management, should be considered for surgical resection. Multidisciplinary management is the key to the successful management of this uncommon condition.

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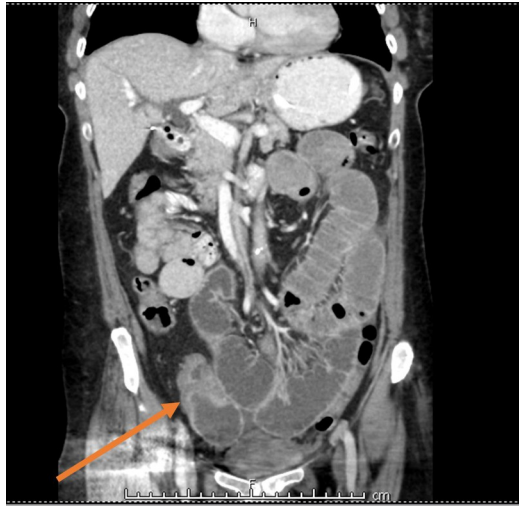


Figure 2. CTAP during acute admission with partial small bowel obstruction showed progression of the thickening in distal ileum seen on imaging 3months prior (see figure 1)

ANS\_17084\_ANS\_17084\_amyloidosis figure 2.jpg

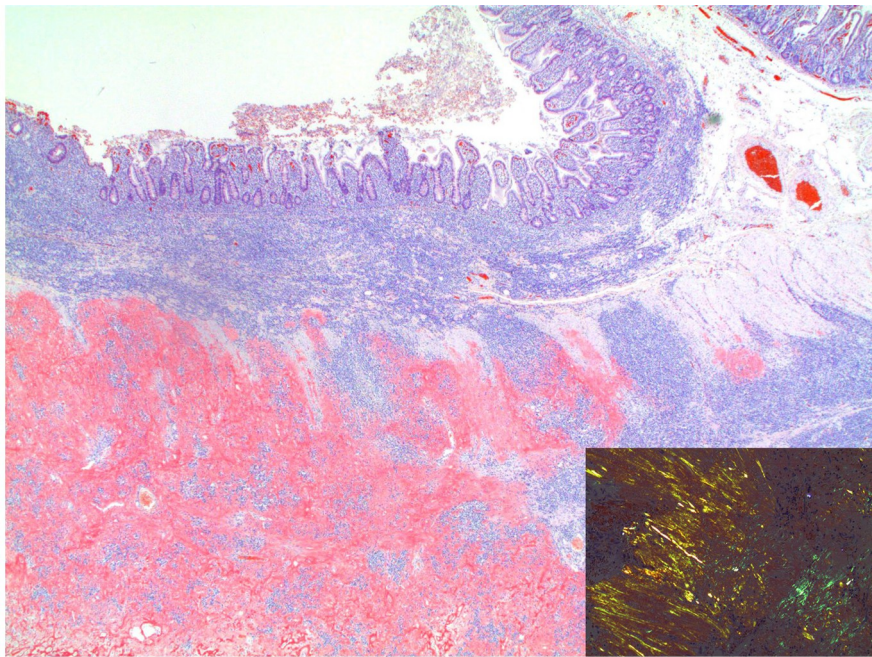


Figure 3. Congo red section x2 objective: massive mural amyloid deposit destroying the muscularis propria. Inset: x10 objective, polarized, demonstrating green birefringent amyloid material.

ANS\_17084\_ANS\_17084\_amyloidosis figure 3.jpg



Figure 1. CT enterography showed concentric bowel wall thickening in the distal ileum suggestive of crohn's stricture

ANS\_17084\_ANS\_17084\_Amyloidosis figure 1.jpg

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