EDITOR'S QUIZ: GI SNAPSHOT

Curious case of gut dysmotility

A previously fit 78-year-old man presented with a 2-month history of dysphagia, epigastric pain, bloating, intermittent vomiting and 1½ stone weight loss. There was no history of new medications or recent travel abroad. Oesophagogastroduodenoscopy (OGD) showed a 'cup and spill' stomach with mild gastritis. Oesophageal biopsies were normal. CT scan of abdomen/pelvis showed extensive small bowel dilatation to the caecum but no transition point (figure 1A). Exploratory laparotomy revealed dilated small bowel loops without mechanical obstruction. Initial duodenal biopsies showed non-specific flattened villi with thickening and eosinophilia of submucosal vessels. Autoimmune screen including tissue transglutaminase and anti-neuronal antibodies were negative. The patient had normal renal and cardiac function.

The patient could not tolerate enteral feeding due to persistent vomiting and started parenteral nutrition with a nasogastric tube for decompression. Barium meal and follow through confirmed delayed gastric emptying and increasing small bowel distension with slow transit (figure 1B). A diagnosis of intestinal failure secondary to gut dysmotility of uncertain aetiology was made and he was transferred to a regional centre for Intestinal Failure. OGD and colonoscopy were performed to obtain more biopsies. Push enteroscopy showed duodenojejunal dilatation but no mechanical strictures. A laparoscopy was arranged to insert a venting gastrostomy because he was intolerant of the nasogastric tube and to obtain a full thickness small bowel biopsy as results from endoscopic biopsies were not yet available.

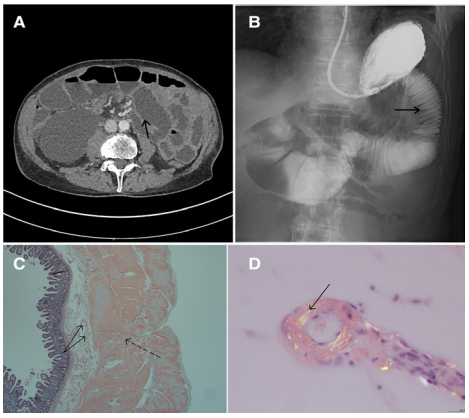

Figure 1 (A) CT showing dilated, fluid-filled loops of small bowel with thickened wall. (B) Barium follow through showing dilated small bowel and delayed transit. (C) Full thickness small bowel stained with Congo red in submucosal blood vessels (solid arrows) and diffuse staining in muscularis propria (dotted arrow) (×2 magnification). (D) Submucosal vessel stained with Congo red and polarised to show apple-green birefringence (×20 magnification).

QUESTION

What is your diagnosis? How would you manage this condition?

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ANSWER

The small bowel biopsy stained with Congo red showed significant amyloid deposition within submucosal blood vessels (figure 1C). When polarised, this showed apple-green birefringence typical of amyloidosis (figure 1D). Colonic biopsies showed similar findings. Immunohistochemical staining of amyloid deposits did not react with serum amyloid A protein, transthyretin or kappa and lambda immunoglobulin light chains. A diagnosis of amyloidosis of uncertain type was made. Following a bone marrow biopsy, he was diagnosed with plasma cell myeloma.

Amyloidosis is rare with an annual incidence of 5.1–12.8 per 1 million cases in the UK, higher in males.[1] It is characterised by insoluble protein deposition, which is resistant to digestion, within tissues (cardiac, renal, GI, hepatic) leading to progressive dysfunction and failure.[2] GI amyloidosis presents with a myriad of symptoms, for example, abdominal pain, refractory dyspepsia, weight loss or haemorrhage. The small bowel is most commonly affected.[3] Our patient displayed features of gastric and small bowel dysmotility and subsequent intestinal failure.

The characteristic histological feature is green birefringence under polarised light on Congo red staining. Treatment includes myeloablative chemotherapy, stem cell transplantation in suitable candidates and managing the underlying condition.[4] AL amyloidosis has a poorer prognosis than AA amyloidosis and GI involvement indicates worse outcomes (median survival 8 months).[5]

Phey Shen Lee ,[1] **Laura Jopson,**[1] **Stephanie J Needham,**[2] **Christopher G Mountford,**[1] **Nick P Thompson**[1]

[1]Gastroenterology, Freeman Hospital, Newcastle upon Tyne, UK
[2]Cellular Pathology, Royal Victoria Infirmary, Newcastle upon Tyne, UK

Correspondence to Dr Phey Shen Lee, Gastroenterology, Freeman Hospital, Newcastle upon Tyne NE7 7DN, UK; pheyshen.lee@nhs.net

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ORCID iD
Phey Shen Lee http://orcid.org/0000-0002-6007-8227

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