A Rare Case of Gastrointestinal Amyloidosis due to Monoclonal Gammopathy of Undetermined Significance

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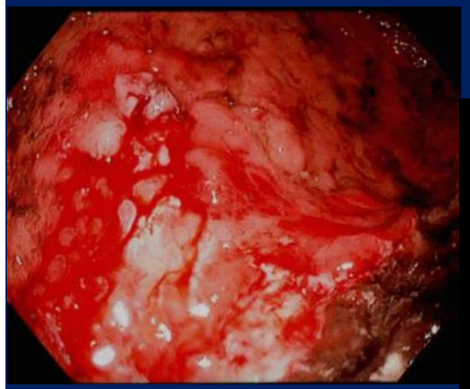
Introduction

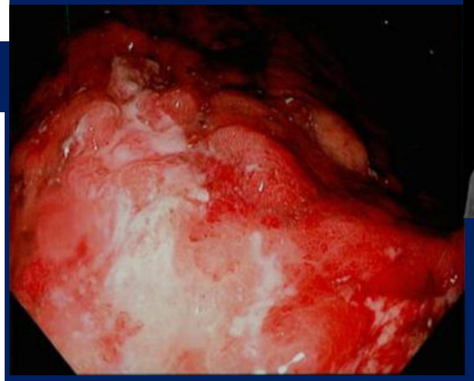
- Amyloidosis of the gastrointestinal (GI) tract is caused by extracellular deposition of fibrils, made up of a variety of serum proteins. It can result from either mucosal or neuromuscular infiltration.
- Symptomatic GI amyloidosis usually presents with one of four syndromes: GI bleeding, malabsorption, protein-losing gastro enteropathy, or GI dysmotility.
- Diagnosis of GI amyloidosis requires a tissue biopsy with positive staining of amyloid by Congo red or the presence of amyloid fibrils on electron microscopy.
- Prognosis of patients with amyloidosis and GI involvement appears to be worse than those without GI involvement.

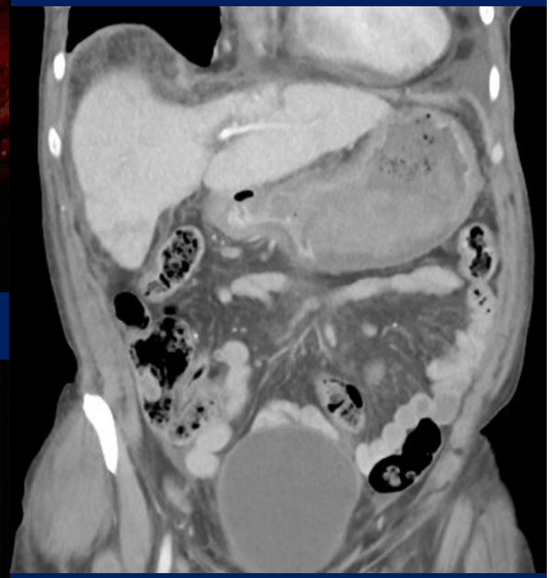
Lab Findings

- IgA 62
- IgG 179
- IgM 13
- Kappa/Lambda Free light chain 66.13
- Kappa Free light chain 39.68
- Lambda Free light chain 0.60
- Gastrin 289
- Electrophoresis: hypogammaglobulinemia, M spike, microalbumin 2.3, protein 193
- CA 125 136.0
- LDH 317



Lining of the duodenum





CT scan of the abdomen

Case Presentation

History of Present Illness:

- A 64-year-old female with a past medical history significant for hypertension, systolic heart failure, depression, hyperlipidemia and hypothyroidism presented with right lower quadrant abdominal pain.

Physical Exam:

- Vitals stable, lungs clear to auscultation, abdominal exam benign

Hospital Course:

- Initial CT abdomen/pelvis, showed evidence of mesenteric, upper abdominal, retroperitoneal adenopathy and omental findings concerning for metastatic disease or carcinomatosis. EGD with biopsy revealed multiple gastric and duodenal ulcers.
- She presented five months later with hematemesis, weight loss, melanotic stool and abdominal pain. EGD revealed findings suggestive of gastric malignancy likely involving both the body and cardia of the stomach. CT abdomen/pelvis demonstrated worsening findings with hyperintensity of gastric lining and increased adenopathy in the chest.
- CA 125 and LDH levels were elevated. Biopsy of gastric mucosa and duodenum showed findings suggestive of amyloidosis, with AL (Kappa) type amyloid deposition and no adenomatous change or malignancy. Analysis with Congo red staining was positive for amyloidosis.
- Bone marrow biopsy revealed findings consistent with MGUS. A PET/CT demonstrated moderate irregular gastric wall thickening that was faintly metabolically active and infiltration of the omental fat, concerning for intraperitoneal spread.
- She was due to start VRd induction therapy and was considered for an autologous transplant. She was started on CyBorD nine months after initial presentation.
- She passed away at home one month later

Case Conclusion

- This case illustrates that, while rare, it is important for clinicians to be aware of amyloidosis of the GI tract. Due to the progressive nature of the disease, early diagnosis is imperative for effective treatment and prognosis.

References

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