AMSER Case of the Month December 2021

59-year-old male with acute onset nausea and vomiting

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Patient Presentation

- HPI: 59-year-old male with 3-day history of intractable nausea and non-bloody, non-bilious vomiting. Pt also with hx of skin thickening, joint pain, and weight loss.
- PMHx: GERD and iron deficiency anemia
- Surg Hx: Prostate cancer s/p prostatectomy (7 years prior), remote hx of appendectomy.
- Medications: Baclofen, Ferrous sulfate, Ondansetron, Pantoprazole, Prednisone, Prochlorperazine, Rifaximin
- Vitals: BP 127/78, HR 129, SpO2 96% on RA, T 36.5 C
- Relevant labs:
  - BMP: wnl
  - CBC: WBC 25.9, Hgb 13.1, Plt 601
  - Lactate 3.8

What Imaging Should We Order?

ACR Appropriateness Criteria

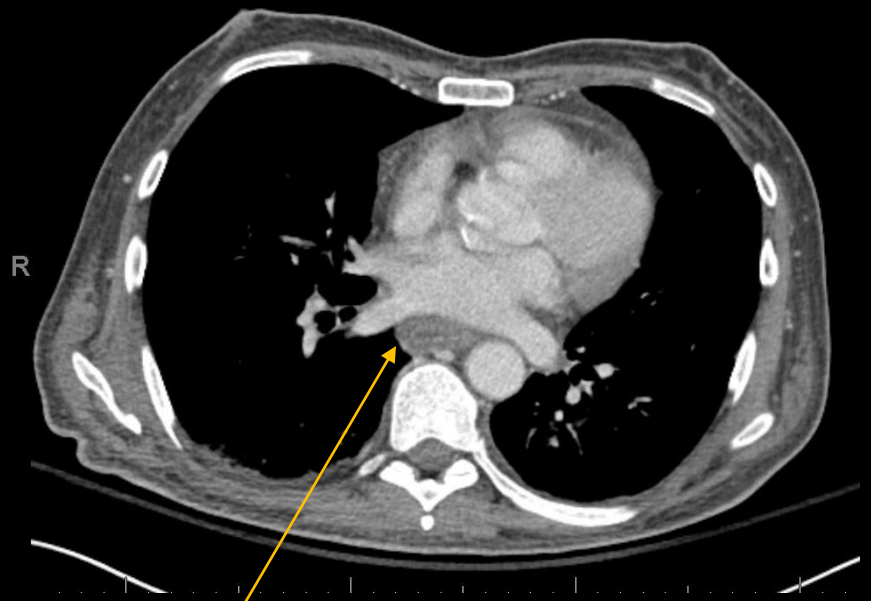
Variant 1: **Suspected small-bowel obstruction. Acute presentation. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast	May Be Appropriate	☢☢☢
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate	O
Radiography abdomen and pelvis	May Be Appropriate (Disagreement)	☢☢☢
Fluoroscopy small bowel follow-through	May Be Appropriate	☢☢☢
MRI abdomen and pelvis without IV contrast	May Be Appropriate	O
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT enteroclysis	Usually Not Appropriate	☢☢☢☢
CT enterography	Usually Not Appropriate	☢☢☢☢
MR enterography	Usually Not Appropriate	O
US abdomen and pelvis	Usually Not Appropriate	O
Fluoroscopy small bowel enteroclysis	Usually Not Appropriate	☢☢☢
MR enteroclysis	Usually Not Appropriate	O

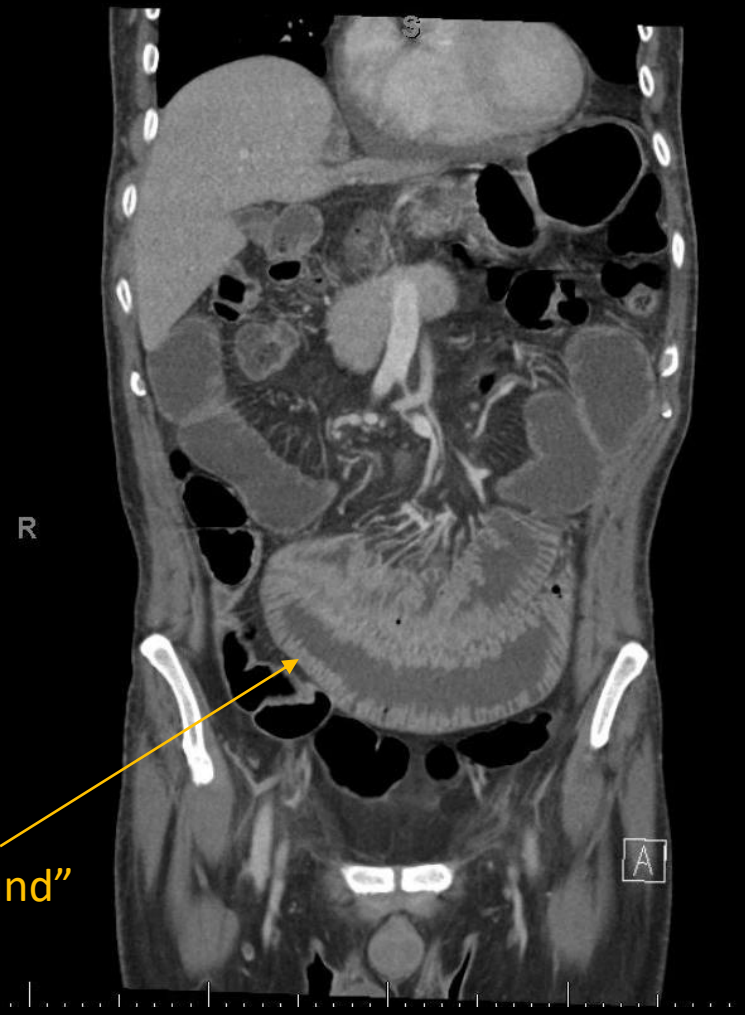
CT A/P w/ contrast completed.

CT Findings: Unlabeled

CT Findings: Labeled

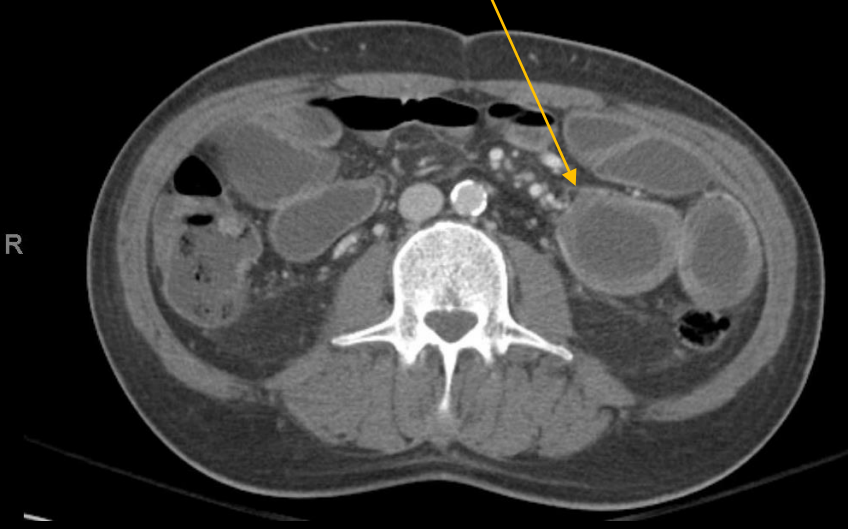


Distal esophageal thickening



"Hide-bound" sign

Apparent thickening of small bowel wall



Small bowel diffusely dilated w/o definite transition point

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Final diagnosis:

Scleroderma

Case Discussion: Systemic Scleroderma

- Major disease subsets:
  - Limited cutaneous systemic sclerosis (CREST syndrome)
    - Calcinosis cutis, Raynaud's phenomenon, Esophageal Dysmotility, Sclerodactyly, Telangiectasia
  - Diffuse cutaneous systemic sclerosis
    - Greater risk of respiratory, cardiac, and renal manifestations, with faster disease progression and increased morbidity and mortality
- Pathophysiology
  - Pathogenesis remains incompletely understood
  - Immune activation, vascular damage, and excessive synthesis of extracellular matrix with deposition of collagen contribute

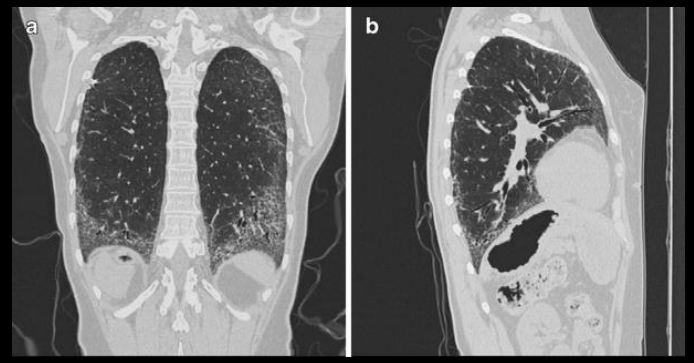
Case Discussion: Systemic Scleroderma

- Examples of Radiologic Manifestations

Esophageal dilatation and dysmotility

Journal of Scleroderma and Related Disorders. 2020;5(1):21-32. doi:10.1177/2397198319848550

Pulmonary fibrosis



Curr Rheumatol Rep. 2010; 12(2): 156–161. doi: 10.1007/s11926-010-0095-0

Hide-bound sign (pathognomonic)

Radiology 1999; 213:No 3 doi: 10.1148/radiology.213.3.r99dc21837

Calcinosis cutis

Current Opinion in Rheumatology30(6):554-561, November 2018. doi: 10.1097/BOR.0000000000000539

Case Discussion: Systemic Scleroderma

Diagnosis:

- Characteristic physical exam findings
- Laboratory testing
  - CBC w/ diff, serum creatinine, CK, urinalysis
  - ANA, Anti-centromere, Anti-topoisomerase I (Anti-Scl-70), Anti-RNA Polymerase III
- Imaging
  - High Resolution Computed Tomography (HRCT)

Prognosis:

- Substantial increase in mortality
- Most deaths related to pulmonary fibrosis, pulmonary arterial hypertension, or cardiac causes

References

1) Denton, C. P., Black, C. M., Korn, J. H., & de Crombrugghe, B. (1996). Systemic sclerosis: Current pathogenetic concepts and future prospects for targeted therapy. *The Lancet*, *347*(9013), 1453–1458. https://doi.org/10.1016/s0140-6736(96)91687-6
2) Pickhardt, P. J. (1999). The “hide-bound” bowel sign. Radiology, 213(3), 837–838. https://doi.org/10.1148/radiology.213.3.r99dc21837
3) Reveille, J. D., & Solomon, D. H. (2003). Evidence-based guidelines for the use of immunologic tests: Anticentromere, SCL-70, And nucleolar antibodies. Arthritis & Rheumatism, 49(3), 399–412. https://doi.org/10.1002/art.11113
4) Tyndall AJ, Bannert B, Vonk M, *et al.* Causes and risk factors for death in systemic sclerosis: a study from the EULAR Scleroderma Trials and Research (EUSTAR) database. *Annals of the Rheumatic Diseases* 2010;**69:**1809-1815.