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Presumptive lupus enteritis

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Title: Presumptive Lupus Enteritis

Presentation:

A 22 year old student of engineering presented with a two week history of facial puffiness and pedal edema. Investigations revealed anemia, lymphopenia, urinary nephritic sediment, low complements, Antinuclear antibodies with a homogenous pattern on immunofluorescence and high titers of Anti-double stranded DNA antibodies. A renal biopsy was performed and it was uneventful. It confirmed Class 4 Lupus nephritis (NIH Activity Index 10/24, Chronicity 1/12). A week later, she went to a surgeon elsewhere with severe abdominal pain, vomiting and obstipation with progressive abdominal distension. She did not have melena. Examination revealed tachycardia with normal blood pressure and diffuse abdominal tenderness. Percussion revealed a periumbilical tympanitic note with dullness in the flanks suggestive of free fluid; bowel sounds were sluggish on auscultation. Suspecting obstruction, a Ryle's tube was inserted and she was referred here. Nutrition was maintained intravenously.

Assessment: An X ray of the abdomen in the supine posture showed centrally located air-filled small bowel loops, suggesting free fluid in the abdomen.[figure 1] Computed tomography of the abdomen demonstrated the 'Target sign' which signifies a diffusely edematous bowel wall with circumferential wall thickening. [figure 2] Ascites, pleural effusion and loss of cortico-medullary differentiation of the kidney was also noticed. CT- angiography did not reveal any evidence of venous or arterial thrombosis. Serum levels of amylase and lipase were not raised. Antiphospholipid antibodies were not present.

Diagnosis: Lupus enteritis, defined as either vasculitis or inflammation of the small bowel, is a rare complication of Systemic Lupus Erythematosus and the pathogenesis is poorly understood. It usually presents with abdominal pain, vomiting or diarrhea in patients already diagnosed with Systemic Lupus Erythematosus - although in an analysis of case reports, only a thirds had other clinical manifestations of active lupus.[1] Important differentials are pancreatitis, intestinal pseudo-obstruction and abdominal infections- such as salmonellosis- especially in the presence of immunosuppressive agents. [2] The target sign on CT, suggesting abnormal bowel wall enhancement, is not specific to a particular diagnosis and has been reported in inflammatory bowel disease, vascular disorders, intestinal infections and ischemic colitis. In lupus, it can be attributed to a nonspecific vasculitis of the mesenteric vessel branches or bowel ischemia secondary to hypercoagulable state. The patient's age and the absence of antiphospholipid antibodies made the latter less likely in her, and therefore a diagnosis of lupus enteritis was made.

Management: She was pulsed with intravenous methylprednisolone for three days followed by oral prednisolone, and cyclophosphamide infusions were initiated. A week later her abdominal pain subsided and oral feeds were tolerated. Lupus enteritis typically responds well to immunosuppression, steroids being the usual choice of first line drugs. [3] This case illustrates the importance of early imaging diagnosis of a potentially life-threatening condition.

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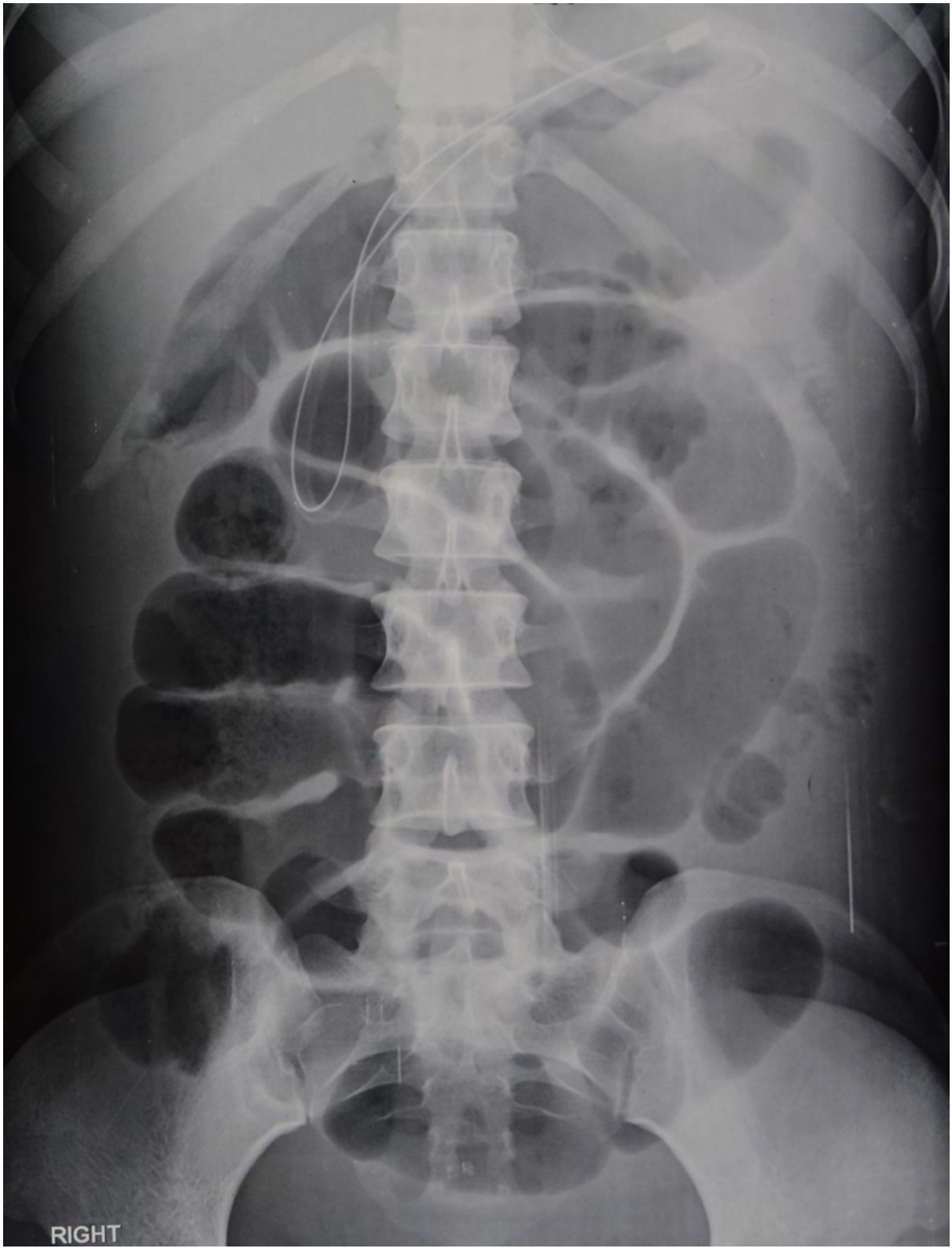
Figure Legends:

1. Abdominal radiograph (supine position) showing multiple air filled, centrally located bowel loops.

2. Computed tomography of the abdomen showing ‘target sign’ of the small intestinal loops signifying edematous bowel wall with circumferential wall thickening (straight arrows). These loops show a double rim of hyperdensity with the central lumen being of fluid density, in contrast to the normal appearing bowel loops seen (open arrowheads). Also noted are mesenteric vasculature leading on the loops (curved arrow) and massive ascites (asterisks).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