CASE SERIES

AUTOIMMUNE DISEASE PRESENTING AS ACUTE ABDOMEN

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Mesenteric vasculitis and resultant bowel ischemia is rare but serious complication of autoimmune disorders. Early detection and treatment is the key to avoid potentially fatal outcomes of bowel perforation and peritonitis. In this series, we present patients presenting with acute abdominal pain and having CT imaging features of bowel ischemia who responded well to immunosuppressive therapy. The aim of this work is to familiarize health professionals with possibility and imaging features of mesenteric vasculitis.

Keywords: Acute abdomen; Autoimmune; Laparatomy; Vasculitis; Bowel ischemia; Mesenteric vasculitis

J Ayub Med Coll Abbottabad 2017;29(1):139–41

INTRODUCTION

Vasculitis syndromes usually result in noninfectious inflammation of blood vessels. Various syndromes have predilection for different sized blood vessels and although rare in incidence, many of them can present with a wide spectrum of symptomatology. Vasculitis syndromes can involve any part of the body with classic and rare presentations. Autoimmune disorders particularly systemic lupus erythematosus (SLE) can lead to vasculitis of gastrointestinal tract with presentations like abdominal pain, nausea, vomiting and loose stools. The associated findings could include severe serositis, enteritis, and pancreatitis and bowel ischemia leading to perforation.[1,2] The unusual presentations of ischemic bowel changes as a result of mesenteric vasculitis pose a diagnostic challenge. In patients with already known underlying autoimmune disease, a presentation of acute abdomen with or without signs of peritonitis should always raise concern of bowel ischemia secondary to mesenteric vasculitis. The role of computed tomography (CT scan) has recently been highlighted in literature as an efficient tool in diagnosis of mesenteric vasculitis and resulting inflammatory & ischemic changes.[3] In this work we would like to describe a series of patients with acute abdomen secondary to mesenteric vasculitis and resultant changes in bowel.

CASE 1

A 32-year-old lady presented to emergency room (ER) with complains of severe abdominal pain and loose stools for past 6 days. Her past medical history was positive for Systemic Lupus Erythematosus and seronegative polyarthritis. She was not taking any active medications at time of presentation. On examination, she was afebrile with a blood pressure of 114/70 mm Hg, Pulse rate of 140 beats per minute and a respiratory rate of 24 breaths per minute.

Systemic examination revealed distended abdomen with rigidity, rebound tenderness in epigastrium and absent bowel sounds. A contrast enhanced CT scan of the abdomen and pelvis was performed which revealed diffusely thick walled, oedematous and mildly dilated small bowel along with surrounding fat haziness and peripheral enhancement. The findings were giving characteristic target appearance without evidence of intramural air. The top differential of this appearance was bowel ischemia however there was no evidence of thrombus in superior mesenteric artery or vein (Figure-1). The patient was started on pulse dose of Methyl prednisolone and responded well to it. The complaints of abdominal pain and rigidity resolved and patient was discharged home on Immunosuppressive treatment.

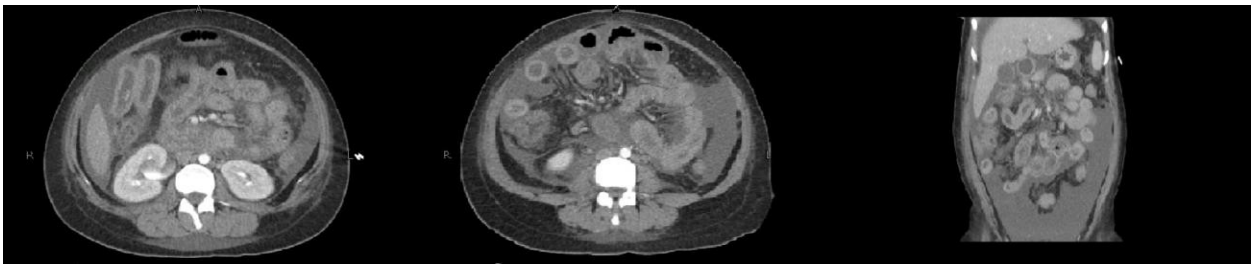

Figure-1: Thickened and enhancing small bowel loops with target appearance. Associated serositis has resulted in ascites

CASE 2

A 16-year-old lady presented to emergency room with complaints of severe abdominal pain and vomiting for past 15 days. Her past medical history was positive for Systemic Lupus Erythematosus and before presenting to our institute she was conservatively managed in a peripheral hospital with provisional diagnosis of intestinal obstruction. She was not on any active treatment for SLE at time of presentation. On examination, she was afebrile with a blood pressure of 128/80 mm Hg, Pulse rate of 89 beats per minute and a respiratory rate of 20 breaths per minute. Systemic examination revealed diffusely tender abdomen with hyper resonant note on percussion and sluggish gut sounds. An initial impression of sub-acute intestinal obstruction was made and management started on its lines.

A contrast enhanced CT scan of Abdomen was ordered which revealed; abnormal differential enhancement of almost the entire small bowel loops without any significant dilatation. Normal mesenteric vasculature was seen without evidence of thrombosis; the findings were consistent with bowel ischemia (Figure-2). The patient was administered methyl prednisolone as pulse therapy and responded well to it. The complaints subsided, she was started enteral feeding which she tolerated well and subsequently was discharged from hospital.

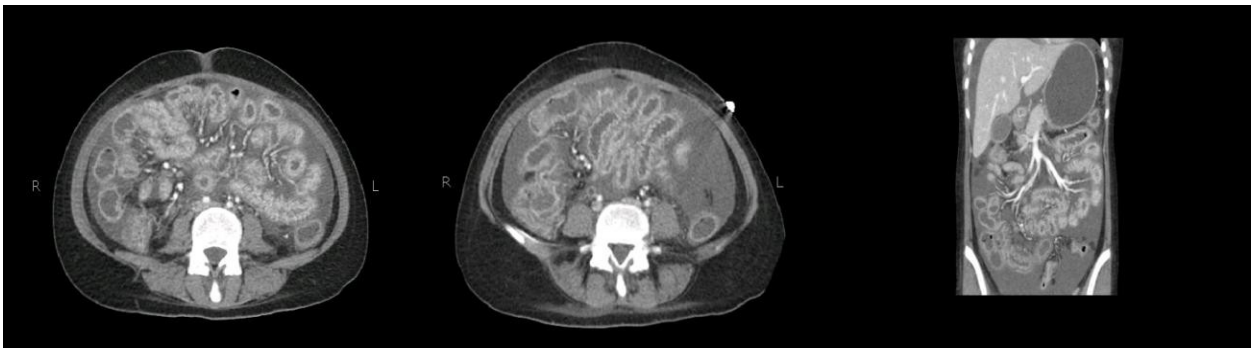

Figure-2: Oedematous small bowel loops with differential enhancement. Associated serositis has resulted in ascites

CASE 3

A 55 years old gentleman presented to emergency room with history of abdominal pain, purpura and pruritus for 15 days. He had no previously known comorbidities. On examination, he was afebrile with a blood pressure of 130/80 mm Hg, heart rate of 110 beats per minute and respiratory rate of 18 breaths per minute. Systemic examination revealed a soft, diffusely tender abdomen. After initial resuscitation contrast, enhanced CT scan of abdomen was performed to evaluate the abdominal pain. The findings of this scan revealed abnormal wall thickening with dilatation of the proximal jejunum, distal ileum and terminal ileum with associated involvement of caecum.

The caecum was giving a targeted appearance. Surrounding fat stranding with significantly increased vascularity of the arterial arcades (vasa recta) was also noted giving appearance of comb sign. Patient was administered methyl prednisolone as pulse therapy along with azathioprine and responded well to it. The complaint subsided and was subsequently discharged from hospital

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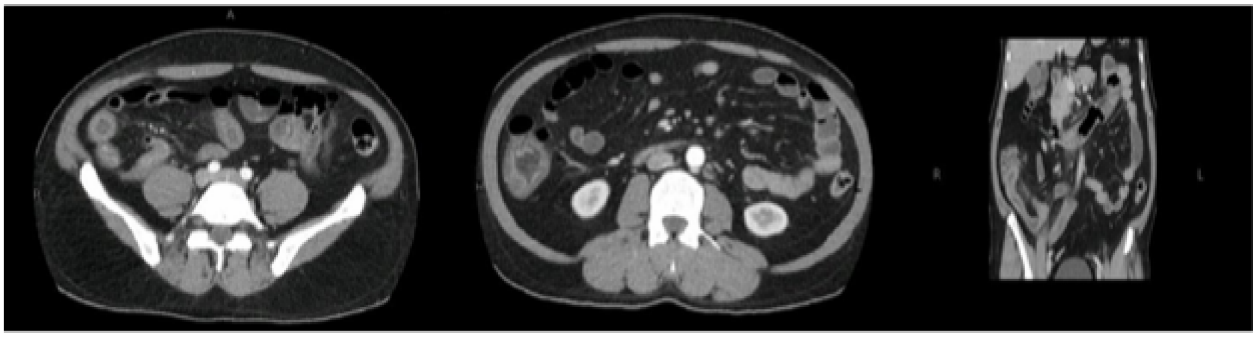

Figure-3: Abnormal wall thickening of distal small bowel loops including terminal ileum. Note the target appearance of caecum

DISCUSSION

Mesenteric vasculitis is one of the most serious complications of autoimmune disease particularly systemic lupus erythematosus. It results in associated changes of bowel ischemia and mucosal inflammation leading to enteritis. In severe cases this may result in bowel perforation and subsequent peritonitis. The incidence of mesenteric vasculitis in patients with systemic lupus erythematosus is reported to be in range of 0.2–53%. A variety of symptoms have been observed in patients presenting with underlying autoimmune process however acute abdominal pain remains to be the most common presenting feature with reported frequencies of 33–62%. The cause of acute abdominal pain can be secondary to involvement of central nervous system, serositis, enteritis, pancreatitis, uraemia and primary peritonitis however mesenteric vasculitis leading to bowel ischemia remains to be the most serious.[1,2,4]

Computed Tomography (CT scan) remains to be the most valuable modality in early diagnosis of bowel ischemia. The strength of CT scan lies in its ability to demonstrate bowel wall and mesenteric vasculature simultaneously. Typically, CT findings in mesenteric ischemia include abnormal dilatation of loops with or without wall thickness (this can be focal or diffuse). Abnormal enhancement which are classically describe by double halo or target signs. Mesenteric oedema with prominent vasculature. Although these signs are sensitive in detection of bowel ischemia, the downside is their lack of specificity as they can be seen in a variety of other disease processes as well. Although intramural air is a classic finding in bowel ischemia due to thrombotic episode, it is rare in mesenteric vasculitis. The same was observed in our patients as well. Recent literature has also emphasized on the importance of initial mesenteric vascular prominence (Comb sign) as an early sign of mesenteric vasculitis. Similar picture was observed in one of our patients as well.[3,5]

Other finding seen on imaging in autoimmune processes as described in literature include lymphadenopathy, hepatosplenomegaly, inflammatory changes of the pancreas, ascites and renal parenchymal changes. Ascites and pancreatitis was observed as associated findings in our patients as well.[3]

According to findings of established literature patients usually respond well to high dose prednisolone therapy with resolution of symptoms. The same was observed in this series as well. A surgical intervention is rarely required. Most of the cases later need to be started on immunosuppressant therapy by use of agents like Azathioprine and Cyclophosphamide as relapse is common.[2] The limitation of our series include lack of follow up scans to assess the extent of resolution of disease process on imaging.

CONCLUSION

Early diagnosis and management by high dose steroids is the key to prevent complications in cases of mesenteric vasculitis. The diagnosis should be considered in any patient with history of autoimmune disorder presenting with acute abdomen or having Imaging features of bowel Ischemia.

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Received: 14 August, 2015	*Revised: 17 May,2016*	*Accepted: 26 December, 2016*

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