Gastrointestinal imaging findings in collagen vascular diseases

Poster No.: C-452
Congress: ECR 2009
Type: Educational Exhibit
Topic: Abdominal and Gastrointestinal
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Keywords: ct, SLE, collagen disease, SSc
DOI: 10.1594/ecr2009/C-452

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Learning objectives

To illustrate abdominal imaging features in collagen vascular diseases on CT and barium study.

Background

Collagen vascular diseases are known to present with various gastrointestinal manifestations classified as 1) gastrointestinal damage due to the collagen vascular disease itself; 2) adverse events caused by pharmacotherapies; or 3) gastrointestinal infections secondary to corticosteroid administration. The first group includes lupus enteritis in systemic lupus erythematosus, pneumatositis cystoides intestinalis in scleroderma, and ileocecal ulcer in Bechet's syndrome.

Imaging findings OR Procedure details

1. Gastrointestinal damage due to the collagen vascular disease itself

Systemic lupus erythematosus (SLE)\textsuperscript{1-5)}

SLE is a complex autoimmune disease with multisystem involvement. It can affect any portion of the gastrointestinal tract, which is especially susceptible to small vessel vasculitis). In addition, about 35% of patients with SLE have an antiphospholipid syndrome that places them risk for development of mesenteric arterial or venous thrombosis. Inflammation of the small blood vessels of the gut produces a spectrum of complications, including intestinal ischemia, hemorrhage, ileus, ulceration, infarction, pseudo-obstruction and perforation. Vasculitis is characterized by fibroinoid necrosis with marked wall thickening and minimal infiltration by inflammatory cells). The end result of the vasculitis or thrombus formation is bowel ischemia. The diagnosis of bowel ischemia is thickening of bowel wall with \textit{thumb-printing} appearance on plain radiograph and barium study (Fig. 1 on page ). CT findings of ischemic bowel disease are symmetric bowel wall thickening and mesenteric vascular engorgement and haziness (Fig. 2 on page ). Bowel wall thickening is multifocal, with variable length, and do not appear to be confined to a single vascular territory. Common CT findings of small bowel ischemia are dilated bowel wall thickening with \textit{target sign} such as edema, intramural hemorrhage, ascites and lymphadenopathy. (Fig. 3 on page ) Indeed, the radiologic findings may be indistinguishable from those of ischemia, hemorrhage, or inflammatory bowel disease. In more advanced cases, pneumatositis or mesenteric venous gas is seen on CT. Pneumatositis cystoides intestinalis (PCI on page ) is a relatively uncommon condition characterized by the presence of multiple gas-filled mucosal, submucosal, or subserosal cysts located throughout the colon and/
or small intestine. PCI on page ___ is occasionally associated with collagen vascular diseases, particularly with systemic sclerosis, but rarely with SLE. In patients with SLE, PCI is though to result from ischemic necrosis of the bowel wall due to vasculitis or antiphospholipid antibody disease. PCI in SLE as well as in other condition, is as a benign disorder, especially in adult.

The most serious complication of bowel ischemia is perforation, which is common in sigmoid colon or rectum (Fig. 4 on page ___). CT is useful for detecting of perforation and monitoring for infarction. Often, SLE patients are on multiple drug regimens, including nonsteroidal anti-inflammatory drugs, which can contribute to the development of gastritis and peptic ulcer.

The kidney is usually affected in SLE. However the imaging findings in SLE renal disease are generally nonspecific and similar to those in other causes of medical renal disease. CT can only demonstrate severe renal disease, i.e. diffuse enlargement or diminution of renal size or potentially treatable causes of deteriorating renal function, such as renal vein thrombosis, renal infarcts, urinary obstruction or abscesses. After interstitial cystitis related to SLE is named "lupus cystitis" and it strongly associated with gastrointestinal involvement and CT is a useful radiological tool to investigate them (Fig. 5a-c on page ___)(Fig. d,e) on page ___

Tissue deposition of immune complexes have been reported in the gastrointestinal wall, as well as in bladder wall, which suggests that common autoantigen of both urinary bladder and gastrointestinal tract might play an important pathologic role. Lupus cystitis is frequently associated with hydroureteronephrosis, which is usually due to fibrosis of the ureterovesical junction or detrusor muscle spasm resulting in vesicoureteral reflux.

Pancreatitis is seen in 8-28% of patients with SLE and can be focal and diffuse. Pancreatitis in SLE is due to vasculitis, ischemia of small pancreatic vessels, immune complex deposition, or a combination of these entities.

Systemic sclerosis: SSc, Scleroderma 6-8)

Gastrointestinal involvement occurs in 80% of patients with SSc. Sings and symptoms include dysphagia, regurgitation due to gastroesophageal reflux, and constipation. The distal two thirds of the esophagus is dilated, peristalsis is absent on barium study (Fig 6 on page ___), and the gastroesophageal sphincter is flaccid, resulting in gastroesophageal reflux. On CT, the normal esophagus is ovoid, with a greater coronal than sagittal luminal diameter, not exceeding 10 mm. Intraluminal air in the cervical esophagus is a normal finding on CT. except for scattered small pockets of intraluminal air, however, the presence of fluid, an air-fluid level, or an air-filled lumen exceeding 10 mm in coronal diameter, especially below the level of the aortic arch, is considered abnormal and indicative of obstruction or dysmotility (Fig 7a b on page ___)(Fig 7c, d on page ___).

Sometimes barium study of small bowel shows marked dilatation of second and third parts of duodenum, and jejunum with hide-bound appearance of patients with SSc. On double contrast study using barium, dilatation of small bowel is seen as coiled-spring appearance.
Colonic involvement in SSc occurs less frequently than is observed in the esophagus and small bowel. Symptoms from colonic involvement are non specific, but decreased motility causes constipation and may result in impaction of fetal contents.

PCI is associated collagen vascular diseases such as SLE, mixed connective tissue disease, dermatomyositis, juvenile rheumatoid arthritis, particularly with SSc. In patients with SSc, PCI is a late complication of the disease thought to be due hypoxemia, hypomobility of the leading to a blind loop syndrome with bacterial overgrowth and elevated intraluminal pressure. About 1/3 of affected patients had received steroid treatment before PCI was dissolved. The diagnosis of PCI is established using abdominal radiograph, US and CT (Fig. 8 on page ). CT is most useful of demonstrate of gas in the intestinal wall with highest sensitivity. However, PCI with patients SSc is distinguish from idiopathic PCI on CT (Fig. 9a, b on page ) (Fig. 9c, d on page ).

Rupture of the subserosal cysts of PCI results in pneumoperitoneum without clinical peritonitis, so-called "benign pneumoperitoneum". Benign pneumoperitoneum is seen in patients of SSc with or without PCI and is asymptomatic for years. The cause is show passage of bowel gas through minute or micro-perforation in colonic or small bowel sacculations.

**Dermatomyositis (DM) and Polymyositis (PM)**

Dermatomyositis (DM) is an idiopathic inflammatory disease of unknown etiology which manifests itself with symmetrical proximal muscle weakness and typical heliotrope shin rash. The common gastrointestinal symptom in DM is dysphagia. Distal esophageal dysmotility is reported in up to two thirds of adults with DM, and delayed gastric emptying and dysmotility are commonly seen. The thickening of the small bowel wall, or "stacked coin" appearance, is reported. Occasionally, ulceration and perforation of gastrointestinal tract and PCI is seen in juvenile DM. In children, lesions can develop on the walls of the muscular vessels leading to thrombosis and infarction. Widespread vasculitis is shown to produce intestinal necrosis, ulceration, perforation and hemorrhage. Vasculitis reduces effectiveness of the mucosal barrier and leads to PCI in the DM of childhood. DM of adult differs from the juvenile form. PCI associated with adult patients with DM is very rare and benign condition (Fig. 10a, b on page ) (Fig. 10c, d on page ).

DM has been linked to internal malignancy in adult patients. Associated malignancies are respiratory tract carcinoma in males, genital and breast carcinomas in females. Gastric, esophageal, hepatic and colon cancers are frequent in Japan (Fig. 11 on page ). DM associated with photoallergy has a higher incidence of complications with malignant disease than ordinary DM.

**Polyarteritis Nodosa: PN**

PN is a fibrinoid necrotizing vasculitis that mainly involves small and medium sized arteries of the muscle. Gastrointestinal involvement is most common and occurs in up to 50% of patients, followed kidney, liver, spleen, and pancreas. The small intestine is the most commonly affected portion of the gastrointestinal tract, followed by the mesentery and colon. Two-thirds of patients with PN have abdominal pain, nausea, vomiting due to ischemia and infarction. The diagnosis
of PN is suggested by angiographic findings of aneurisms up to 1 cm in diameter within renal, splenic, mesenteric, and hepatic vasculature; however, this finding is not always pathognomonic for PN (Fig. 12 on page ). Other angiographic findings are vascular ectasia and occlusive vascular disease manifesting as luminal irregularity, stenosis, or occlusion of small and medium-sized arteries of the viscera. Recent MDCT and breath-hold MR imaging may allow early detection of vasculitic changes and may read such noninvasive imaging techniques replacing invasive angiography for that diagnosis. Other imaging techniques such as CT, MR imaging, sonography, and barium studies usually result in nonspecific findings of ischemic injury (Fig. 13 on page ).

Henoch-Schönlein Syndrome

Henoch-Schönlein (HS) Syndrome, or anaphylactoid purpura, is a systemic vasculitis of unknown cause that affects small vessels and mainly involves the skin, joints, gastrointestinal tract, and kidneys. The diagnosis is based on such characteristic clinical signs and symptoms as skin rash, arthritis involving the large joints, colicky abdominal pain, gastrointestinal bleeding, and hematuria. However, in the absence of characteristic features, especially cutaneous involvement, the diagnosis is massed. HS syndrome spares the very young and is most common in children between the ages of 3 and 10 years, occurring with a slight male predominance. It occurs more often in the winter than in other seasons. Gastrointestinal involvement occurs in more than half of affected patients; abdominal pain is the most frequent complain. When gastrointestinal symptoms predominate or precede the appearance of skin lesions. Gastrointestinal bleeding occurs in about one half of the pediatric patients but is unlikely to require transfusion. Bleeding is less common in older patients. Pathologically the most common abnormality in the gastrointestinal tract is submucosal and mural infiltration of the bowel by blood or edema fluid causing an intense scarlet color. Microscopically, changes of a vasculitis are seen, with endothelial proliferation and thrombosis of small arterioles. The gross appearance may closely resemble Crohn disease. The intestinal reaction usually resolves spontaneously, but perforation or intussusceptions and obstruction may occur.

Radiographically HS syndrome usually causes small bowel involvement manifested as fold thickening which is smooth or irregular, “thumb-printing”, pseudotumors, and intestinal hypomotility. These changes are reversible. No characteristic radiological findings are seen. The CT features of HS syndrome include multifocal area of wall thickening, luminal narrowing, fold thickening, and ulceration. Enlargement mesenteric nodes, small bowel dilation, engorged mesenteric vasculature, ascites, and mesenteric stranding have also been seen (Fig. 14 on page ).

Beçhet's Syndrome

Beçhet's syndrome is a well-recognized, nonspecific necrotizing vasculitis involving multiple organ systems with well-known clinical manifestations including orogenital ulceration, uveitis, arthritis, and neurologic and gastrointestinal involvement. The gastrointestinal tract is involved in 10%-40% of patients with Beçhet's syndrome; the usual site is the ileocecal area, especially the terminal ileum. The presence of deep, penetrating ulcers results in a high frequency of
complications, such as perforation, bleeding, and peritonitis. Barium study shows a large ovoid or irregular ulcer with marked thickening of the surrounding intestinal wall at sites of involvement (Fig 15 on page ). Multiple ulcers and skip lesions are typical. The ulcers in Bechet's syndrome tend to be larger and deeper than those in Crohn disease.

CT shows a polypoid mass bowel thickening (Fig 16 on page ) (Fig 17 on page ). Occasionally, central ulceration of polypoid lesions is demonstrated at CT. Polypoid lesions with central ulceration can easily be mistaken for a concentrically thickened bowel wall. Marked contrast enhancement of involved bowel segment is presumed to be stasis of blood in association with vasculitis and perivasculitis involving primary the veins and venules of the submucosal around the ulcers. The presence of diffuse vascular dilation with perivascular lymphocytic infiltration at microscopic examination supports this assumption. Although CT findings alone are not specific for the diagnosis of Bechet's syndrome in patients without a history of the disease. CT has a primary role in patients with a history of Bechet's syndrome in detection of the primary lesion and exclusion of extraluminal complications. Because of the presence of deep, penetrating ulcers, a high of complications, such as perforation, fistula, hemorrhage, and peritonitis has been reported; the perforating ulcers tend to occur at multiple sites. Differentiation of Bechet's syndrome from Crohn disease and intestinal tuberculosis without evidence of diffuse peritoneal changes may be difficult or impossible when bowel wall thickening is localized to the ileocecal region, but the polypoid type of bowel involvement is rare in the latter two diseases.

2. Adverse events caused by pharmacotherapies

Adverse events caused by pharmacotherapies include lesions in gastrointestinal tract due to nonsteroidal anti-inflammatory drugs (NSAID) and corticosteroid. Multiple gastric ulcers in antrum are common in patients receiving NSAID (Fig. 18 on page ). As gastrointestinal perforation in steroid-treated patients displays relatively high incidence with poor prognosis, careful attention to such complications is needed.

3. Gastrointestinal infections secondary to corticosteroid administration

Gastrointestinal infections secondary to corticosteroid administration include candidal esophagitis and cytomegalovirus enteritis. Prompt diagnosis is required to prevent colonic bleeding and perforation due to cytomegalovirus.
Fig.: Small intestinal wall thickening in a 42-year-old woman with SLE. a. Barium study showed wall thickening with thumb-printing appearance in ileum (red arrow). After steroid therapy, intestinal wall thickening was improved.
Fig.: Large bowel wall thickening in a 44-year-old woman with SLE. Contrast-enhanced CT shows transverse and ascending colon wall thickening with luminal narrowing (pink arrows) and ascites.
Fig.: Bowel perforation in a 52-year-old woman with SLE Un-enhanced CT shows extraluminal abnormal gas in front of sigmoid colon (yellow arrows). Necrosis and perforation of sigmoid colon were confirmed at operation.
Fig.: Lupus cystitis and peritonitis in a 43-year-old woman. a-c Contrast-enhanced CT shows small intestinal wall thickening (pink arrows), ascites and trabeculated bladder with wall thickening (yellow arrow).
**Fig.**: d. Axial image of contrast-enhanced CT shows bilateral hydronephrosis. e. 3D-CTU shows marked hydronephrosis and hydroureter bilaterally.
Fig.: Dilatation of esophagus in a 39-year-old woman in SSc a,b Esophagography shows dilatation of esophagus and poor peristalsis.
Fig.: Dilatation of esophagus and chronic interstitial pneumonia in a 45-year-old woman in SSc a. b Chest radiographs show dilatated esophagus clearly (white arrows).
Fig.: c CT shows dilatation of esophagus with air-fluid level (white arrow) in left side of trachea. d CT on lung window shows interstitial changes in both lower lobes. Red arrow: dilatation of esophagus
Fig.: CT colonography of idiopathic PCIa, b CT colonography shows multiple intraluminal air cysts.
Fig.: c,d Virtual encsopic image of idiopathic PCI is similar to that of conventional endoscopy.
**Fig.**: PCI in a 46-year-old woman in DMa, b CT on lung window shows extensive gas within and around the wall of ascending and transverse colon.
**Fig.** c,d Chest CT shows pneumomediastinum (white arrows) and interstitial change.
Fig.: Gastric cancer a 52-year-old woman in DMa Contrast-enhanced CT shows abnormal gastric wall thickening (yellow arrows). b Barium study shows pyloric stenosis constant with gastric cancer.
Fig.: Intestinal involvement in a 51-year-old man in PNa,b. Contrast-enhanced CT shows wall thickening of terminal ileum and mild mesenteric vascular engorgement.
Fig.: Small intestinal involvement in a 40-year-old woman in Henoch-Schönlein syndrome. b Contrast-enhanced CT shows multifocal intestinal wall thickening (white arrows) and small amount of ascites.
**Fig.**: Mass formation with ulcer in a 43-year-old woman in Beçhet's syndrome. Contrast-enhanced CT shows mass with ulcer in terminal ileum.
**Fig.** Multiple small intestinal ulcers in a 45-year-old woman in Beçhet's syndrome. a, b Barium enema shows multiple ulcers in terminal ileum (red arrow).
**Fig.**: Infiltration of terminal ileum in a 31-year-old woman in Beçhet's syndrome. Contrast-enhanced CT reveals focal wall thickening with enhancement in terminal ileum (red arrow).
**Fig.:** Multiple gastric ulcers in patient receiving NSAID. Endoscopic image shows multiple ulcers in gastric antrum.
Thumb-printing appearance on Plain radiograph or barium study

- Thumb-printing is the most diagnostic finding of small and/or large bowel ischemia
- Wide transverse band associated with haustral fold thickening at colon
- Other conditions: Infectious enterocolitis, Pseudomembranous colitis, IBD, hemolytic-uremic syndrome, et. al.

Fig.: D.D.
Target sign on CT

- Or bull’s eye appearance on CT
- Symmetrical bowel wall thickening
- An enhancing central higher density mucosal layer is surrounded by a water density submucosa that in turn is surrounded by a higher density muscularis propria.

Fig.: CT
Pneumatosis intestinalis (PI)

PI is an uncommon but important condition in which gas is found in a linear or cystic form in the submucosa or subserosa of the bowel wall. In adult 85% of cases there is coexistence of gastrointestinal, respiratory or collagen vascular disease.

Fig.: PCI
Conditions associated with PI

GI condition
ulcer, cancer, inflammatory bowel disease,
necrotizing enteritis, infectious colitis, et. al.
Pulmonary condition
chronic obstructive lung disease
Pneumomediastinum
Collagen vascular disease
SSc, Mixed connective tissue disease, SLE

Fig.: D.D.
Fig.: a.b. Contrast-enhanced CT shows small bowel wall thickening with target sign (red arrows), mesenteric vascular engorgement and ascites.
Fig.: PCI in a 39-year-old woman in SSc. a. Abdominal radiograph shows multiple air cysts in transverse colon. b, c Unenhanced CT reveals intraluminal air cysts (yellow arrow) and pneumoperitoneum (pink arrows).
**Fig.**: Splenic aneurisms in a patient of PN. Angiography shows intrasplenic multiple aneurysms.
Conclusion

To recognize image findings on CT and barium study is important in correct diagnosis and treatment in patients with collagen vascular diseases.

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