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MIMICS OF ABDOMINAL COCOON

Radiology

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KEYWORDS

INTRODUCTION-

Abdominal cocoon is a rare condition of unknown cause in which intestinal obstruction results from the encasement of variable lengths of bowel by a dense fibrocollagenous membrane that gives the appearance of a cocoon. This condition is not often suspected preoperatively, and therefore the diagnosis is usually made at laparotomy.

Definition-

Abdominal cocoon a condition in which a fibrous cocoon has surrounded the bowel loops is an uncommon but devastating complication of chronic peritoneal dialysis (PD). Long PD duration and chronic exposure to dialysis solutions are considered risk factors for its development.

The diagnosis of EPS is based on clinical symptoms in combination with pathological findings and abdominal imaging.

Etiology

The incidence varies across the globe. It has been reported to vary between 2.1% to19.4% for patients maintained on PD for 5-8 years. Factors that may contribute to this variation in the published literature include: patient numbers in the study, single center vs registry data, retrospective vs prospective, prevalent vs incident patients, criteria to establish the diagnosis and potential true differences in incidence. However, the observed incidence of abdominal cocoon increases across all studies with length of time on PD.

Clinically presentation

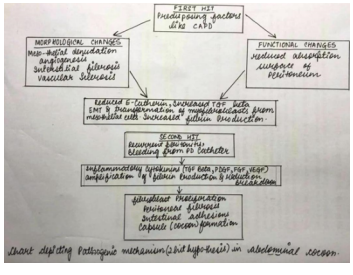
Patients can present with symptoms of abdominal pain, nausea, vomiting, repeated bowel obstruction, blood-stained effluent and Loss of ultra filtration capacity.

Common	Uncommon	Associations
Idiopathic	Cirrhosis	Cryptorchidism
Peritoneal Dialysis related	Endometriosis	Omental hypoplasia
Tuberculosis	Hyper-thermic peritoneal perfusion	Peritoneal encapsulation
Malignancy (Neuroendocrine tumours, Ruptured tumours like GIST or dermoids)	Hemodialysis	Mal-rotation
Post-operative (Renal transplantation, Liver transplant, laparotomy)	Abdominal trauma	Treitz's hernia
Drugs (Beta-blockers, Ergot)	Mycobacterium bovis	
	HIV	
	Whipple's disease	

Pathogenesis

The peritoneal stromal and mesothelial reaction to the various etiologic factors for EPS results in inflammation giving way to peritoneal sclerosis or fibrosis that finally leads to cocoon formation. Chronic irritation of the peritoneum whether chemical or infective, disrupts the mesothelial cell junction damaging the subserosal tissue setting an inflammatory response ending in fibrosis. Analogously recently the pathogenesis of EPS has been explained by the "Two Hit hypothesis" that states that two factors are required for onset of EPS. Pathogenic mechanisms which contribute to genesis of EPS are as described below.

A predisposing factor that causes functional and morphological damage to the peritoneum like in peritoneal dialysis, chronic exposure to the dialysate results in mesothelial disruption (first hit) setting the ground for further development of sclerosis. An initiating factor in the form of an inflammatory stimuli superimposed on the damaged peritoneum like in recurrent peritonitis. (second hit) in the setting of PD. The characteristics of the dialysate like acidity, high glucose and advanced glycation end products, hyper-osmolarity etc cause the first hit by causing mesothelial denudation and subsequent upregulation of profibrotic growth factors like TGF-beta, platelet derived growth factor (PDGF), tumour necrosis factor (TNF)-alpha setting the stage for fibrosis. Also under the influence of transforming growth factor (TGF)-beta, mesothelial cells express plasminogen activator inhibitors (PAI) 1 and 2 leading to decrease in fibrin degradation. Also, exposure to drugs like practolol has been shown to cause mesothelial cell disruption (first hit) that leads to the cascade of events leading to EPS. The damage to mesothelium causes down-regulation of E-cadherin with loss of cell to cell contact and apical basal polarity and with the upregulation of TGF-beta there is epithelial to mesenchymal transformation with mesothelial cells getting converted to myofibroblasts with increased production of fibrin. The second hit in the form of recurrent peritonitis cause upregulation of pro-inflammatory cytokines Interleukin 1, 6, 18, TNF-alpha which along with TGF-beta further expand the process of fibrosis. TGF-beta also causes over-expression of matrix metalloproteinase 2 and tissue inhibitor of matrix metalloproteinase 1 that inhibit fibrin degradation and along with vascular endothelial growth factor (VEGF), PDGF, fibroblast growth factor (FGF) lead to cocoon formation.



Short depicting Pathogenic mechanism (2 hit hypothesis) in abdominal cocoon

Stages of Abdominal cocoon

Based on the clinical presentation, Nakamoto categorized EPS in to four groups(Nakamoto,2005). The following are the proposed clinical stages:

Stage 1 (Pre-EPS stage)	**Asymptomatic with mild ascites and no inflammation.**
Stage 2 (Inflammatory stage)	Patients are symptomatic with nausea and diarrhea consistent with partial encapsulation of the bowel and intestinal swelling. Mild inflammation with fibrin exudation is present.
Stage 3 (Encapsulation)	Symptoms of bowel obstruction due to the formation of the fibrous cocoon causing encapsulation .It can be associated with mild to severe inflammation.
Stage 4 (Chronic stage of ileus)	Patients have absolute bowel obstruction caused by thickening of the encapsulating fibrous cocoon. There is little if any, inflammation at this stage.

Radiological evaluation

Abdominal radiography- Plain abdominal radiography can show air–fluid levels and signs of bowel dilation, indicating obstruction. Another common feature is the presence of peritoneal calcification. However, plain abdominal X-ray films can appear normal. Although it is readily available and helpful in establishing bowel obstruction and peritoneal calcifications, it does not provide conclusive or sometimes not even additional information on the presence or absence of Abdominal cocoon.

Ultrasonography (USG)- USG has been used in the past when EPS was suspected. USG characteristics of EPS are best appreciated with peritoneal fluid in situ. In one study, USG was performed of 1 patient showing presences of Ascites, Abnormal small bowel, Membrane formation. USG is non-invasive and has no radiation burden. A major limitation is that the interpretation of the images is very dependent on the radiologist. There are no data on sensitivity, specificity and reproducibility. Calcifications can also be detected with USG.

Abdominal CT scans- Patients with a clinical suspicion of EPS revealed loculated ascites, adherent bowel loops, narrowing of bowel lumen and a thickened peritoneum. Several other case reports described similar CT findings and other features such as bowel dilation and the presence of peritoneal calcification. In this report we have performed CT scan of all the 3 patients, In all cases, signs of disturbed motility indicated by dilated bowel loops and air–fluid levels were seen and in half of the cases, loculated fluid.

AIMS-

- To highlight the similarities between EPS and its mimics.
- To differentiate EPS from its mimics.

METHODS-

Informed consent was acquired from patients. A retrospective observational study was performed by including all patients who presented with abdominal pain and underwent contrast-enhanced CT scan revealing imaging findings of EPS.

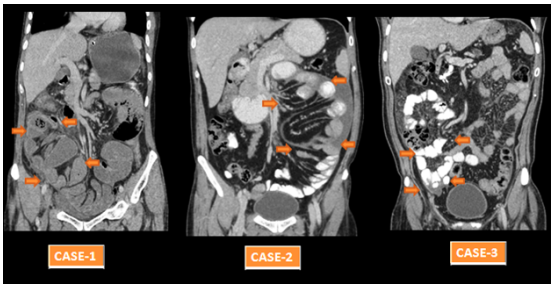
CT imaging was performed at 128 slice GE Optima CT machine by using a multiple phase protocol (plain, arterial, venous and delayed phase).

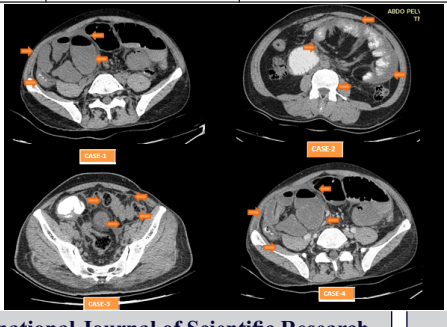
RESULTS-

All patients had peritoneal thickening with variable stages of cocoon formation. One patient had imaging evidence of cirrhosis and thus was labeled to have cirrhosis related disease. Second patient underwent operation to relieve obstruction and was found to have tuberculosis. The 3rd patient was found to have recurrent pancreatitis. None of the patients were on peritoneal dialysis.

	Case 1	**Case 2**	**Case3**	**Case4**
AGE / SEX	51 yrs, Male	53 yrs, Male	41 yrs, Male	60 yrs, Female
CLINICAL PRESENTATION	Abdominal pain since 4 days.	Chronic diabetes leading to endocrinal failure with uncontrolled diabetes	Abdominal pain since 15 days.	Abdominal pain since 1 month. uncontrolled diabetes
USG PERFORMED	NO	YES	NO	NO
Findings of USG	NA	Central clumping of bowel loops without alteration of arrangement on external compression.	NA	NA
CT FINDINGS				
Peritoneal thickness	4.2 mm	2.4 mm	2.9 mm	3 mm
Bowel Dilatation	Involving small and large bowel loops	Involving small bowl loops with clumping.	Involving small bowl loops with clumping.	Absent
Features of inter bowel adhesion	Yes	No	No	No
Features of bowel Gangrene or perforation	No	no	No	No
Peritoneal Calcification	Absent	Absent	Absent	Absent
Vascular pedicle	Normal	Normal	Normal	Normal
Intra abdominal fluid collection	Minimal ascites.	Minimal loculated ascites within sac.	Mild to moderate amount of loculated fluid collection within the sac.	Minimal in the forming sac
Imaging Findings supportive of diagnosis	Pleural thickening with calcification Calcified lymph nodes	Chronic pancreatitis	Portal hypertension secondary to cirrhosis	Cirrhosis of liver, Portal hypertension
Cocoon stage	Stage 2 (Inflammatory stage)	Stage 2 (Inflammatory stage)	Stage 3 (Encapsulation)	Stage 1 (Pre-EPS stage)

IMAGES-





CONCLUSIONS-

None of the patients was suspected for a case of abdominal cocoon or considered to be at an increased risk and non of the patient was on PD.

Early identification of EPS and its mimics is important in patient's management as EPS entails suspension of PD where as diagnosis of its mimic's entails treatment of the underlying etiology.

In view of rapid progression leading to fatal outcome in 50% of cases early diagnosis is critical for patient management.