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ABDOMINAL COCOON: A CASE PRESENTATION

Nursing

Mrs. Hilda Mercy Priyadarsini M. Sc (N), Junior Lecturer, College of Nursing, Christian Medical Collge, Vellore

Mrs. Mary Jancy Joy M. Sc (N), Lecturer, College of Nursing, Christian Medical Collge, Vellore

Mrs. Meenakshi Sekar* M. Sc (N), Professor, College of Nursing, Christian Medical Collge, Vellore *Corresponding Author

ABSTRACT

Abdominal cocoon also known as encapsulating peritoneal sclerosis (EPS) is a clinical syndrome characterized by the formation of cocoon like fibro-collagenous membrane covering the small intestinal loops, resulting in intestinal obstruction. Although the condition is of idiopathic origin in majority of the cases, there are some identified risk factors such as peritoneal dialysis, abdominal tuberculosis etc. Despite radiological advancements, many cases are diagnosed at the time of surgery and the role of conservative therapy remains uncertain. Patients suffer from varied intensities of physical illness and psychological distress associated with the disease. Early identification and timely management may halt the progression of frank cocoon formation and holistic Nursing care will aid in physical and psychological recovery of patients. This article highlights on etiology, pathogenesis, clinical presentation, stages, diagnosis and management of patients with this enigmatic condition.

KEYWORDS

Abdominal cocoon, encapsulating peritoneal sclerosis and intestinal obstruction.

Case history

Mr. K, a 52 years old gentle man presented to emergency department with complaints of obstipation and bilious vomiting for past 2 days with mass felt per abdomen. No past history of tuberculosis, renal failure or abdominal surgeries. Emergency CT abdomen was performed in suspicion of intestinal obstruction. Findings revealed multiple jejunal and ileal loops being encapsulated by enhancing membrane. Multiple small bowel loops were intermittently dilated with air-fluid level and collapsed. Large bowel was collapsed with diffuse peritoneal thickening. Moderate loculated ascites was seen in anterior abdomen with features suggestive of encapsulating peritoneal sclerosis with subacute small bowel obstruction. Peritoneal biopsy showed extensive fibrosclerosing lesion with moderate lymphoplasmacytic chronic inflammation with no evidence of malignancy. This article presents the etiology, pathogenesis, clinical presentation, stages, diagnosis and management of patients like Mr. K who suffer with this condition.

Introduction

Abdominal cocoon or encapsulating peritoneal sclerosis (EPS) or sclerosing encapsulating peritonitis (SEP) is a chronic clinical syndrome of insidious onset, which presents at a later stage with signs of as chronic malnourishment and acute or sub-acute gastrointestinal obstruction (Moinuddin, Summers, Van Dellen, Augustine and Herrick, 2015). It was first described in 1907 as "peritonitis chronica fibrosa incapsulata". In 1978, the International Society for Peritoneal Dialysis (ISPD) has proposed the current term "abdominal cocoon" in patients undergoing peritoneal dialysis. Later the current name was generalized to other conditions, where chronic fibro-inflammatory process results in formation of thickened fibro-connective tissue sheath like structure that envelopes and constricts the small intestine like coccon compromising its motility (Mandavdhare, Kumar, Sharma & Rana (2017).

Etiology

In majority of the cases, abdominal cocoon are of idiopathic in nature otherwise known as primary cocoon. In cases where underlying causes are identified, they are said to be secondary EPS or secondary abdominal cocoon (Moinuddin et al., 2015).

Table 1: Causes and risk factors of abdominal cocoon

Causes		Risk factors
Common	**Rare**	
Idiopathic Continuous Ambulatory Peritoneal Dialysis (CAPD) Recurrent- bacterial peritonitis (bacterial or chemical) Intraperitoneal chemotherapy Peritoneal tuberculosis Malignancy (Neuroendocrine tumors, ruptured tumors or dermoid cysts) Postsurgery (Renal or liver transplantation, laparotomy) Medications (e.g Beta blockers, methotreaxate, povidine-iodine) Ruptured dermoid cysts	Endometriosis Mycobacterium bovis infection Systemic Lupus Erythematosus Abdominal trauma Ventriculo peritoneal/ peritoneo Venous shunts Human Immuno Deficiency Virus infection Protein S deficiency Whipple's disease	Peritonitis (pseudomonas, staphylococcus aureus and certain fungus) Prolonged duration of peritoneal dialysis Organ transplantation Immunosuppressive medications Composition of dialysate (Low pH, high glucose) Ultrafiltration failure Exposure to antiseptics like chlorhexidine

Pathogenesis

Pathogenesis of EPS has been explained by the "Two Hit hypothesis".

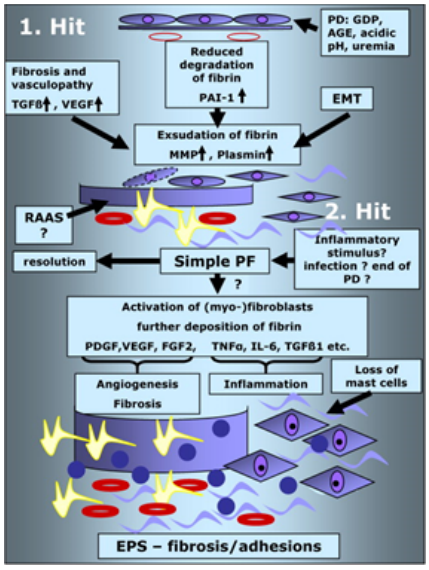

Fig 1: "Two hit hypothesis: Braun, Alscher, Kimmel, Amann & Buttner (2011).

Predisposing factors such as chronic exposure to dialysate acidity, high glucose, advanced glycation end products cause mesothelial disruption, upregulation of profibrotic growth factors like transforming growth factor (TGF-beta), platelet derived growth factor (PDGF), tumor necrosis factor (TNF)-alpha and sets the stage for *fibrosis*. Under the influence of TGF-beta, mesothelial cells express plasminogen activator inhibitors (PAI) leading to decrease in fibrin degradation of the peritoneum setting the ground for further development of *sclerosis.* An initiating factor in the form of an inflammatory stimuli (recurrent peritonitis) superimposed on the damaged peritoneum forms the second hit (Moinuddin et al., 2015).

Clinical presentation

Patients present with the following symptoms Abdominal symptoms like early satiety, anorexia, nausea, vomiting, and altered bowel habits (constipation or diarrhea).

Signs of inflammation (pyrexia and raised CRP) and/or blood stained ascites (in early stages).

Signs of intestinal obstruction, abdominal mass, malnutrition, infection and death (later stages).

Intermittent nature of its presentation- Hall mark of presentation (Brown et al., 2009).

Stages

Based on the clinical presentation, the following clinical stages of abdominal cocoon are proposed (Nakamato, 2005)

Table 2: Stages of abdominal cocoon

Stages	Clinical manifestations
Stage I (Pre EPS stage)	Ascites with hypoproteinemia (Increased peritoneal permeability)
Stage II (Inflammatory stage)	Bacterial or chemical peritonitis (Fever, loss of appetite, loss of weight, fatigue, ascites increase in inflammatory markers- ESR and CRP.
Stage III (Encapsulation)	Signs of progressive encapsulation with small intestinal obstruction and malnutrition.
Stage IV (Chronic stage of ileus)	Formation of cocoon, complete ileus and abdominal mass.

Diagnosis

Diagnosis of cocoon/ EPS is based on history, presenting complaints, physical examination findings, blood, radiological and histological and operative findings.

A) ***History:*** Risk factors and presenting complaints: Nausea, anorexia, early satiety, weight loss, altered bowel habits, malnutrition and ascites (Nakamoto, 2005).

B) ***Physical examination findings:*** Abdominal distension, abdominal mass, ascites, fever and cachexia.

C). ***Blood investigations:*** High CRP, low albumin, leukocytosis and anemia (Yokoyama et al., 2008).

D). ***Radiological investigations:*** Abdominal X-rays- Dilated small bowel loops, multiple air fluid levels, clumping of bowel loops in center of abdomen, peritoneal and bowel wall calcification.

Ultrasonography (USG) of abdomen: Dilated small bowel loops fixed to posterior abdominal wall, clumped bowel loops in "concertina shape" with a narrow mesentery giving appearance like a "cauliflower" or "sandwich".

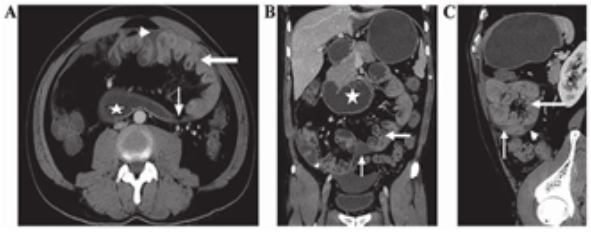

Fig 2: CT findings of abdominal cocoon disease.

CT of the (A) abdomen axial, (B) coronal reformatted and (C) sagittal reformatted show the small bowel contained within a sac. The thickened enhancing membrane (short and thin arrows) covering congregated small bowel loops (thick white arrows) are visible and ascites (white triangle) around loops and the dilated loop of duodenum with fluid (stars) are also visible due to intestinal obstruction. The second and third part of the duodenum mimic the Bottle Gourd appearance (stars).

(Yu, R., Ya, Y., Ni, X., & Fan, G. (2020).

Intestinal barium series: Small intestine loops clumped towards center of abdomen like a cauliflower.

Computed tomography (CT) of abdomen: Small bowel tethering (Ginger bread man sign), thickened peritoneum, peritoneal calcifications and dilated loops of bowel

Cine MRI: Bowel movements confined to some areas of the abdomen.

Management

A) Nutritional Support

Total Parenteral Nutrition (TPN) is indicated as an adjunct treatment in patients with advanced EPS and who has undergone peritonectomy and enterolysis until the intestinal function recovers (Moinuddin et al., 2015). Good nutritional support (enteral or parenteral) reduces post-operative complications, hospital stay and promotes recovery (Mandavdhare et al., (2017).

B) Treatment of underlying etiology

In case of abdominal cocoon in patients undergoing peritoneal dialysis, regular peritoneal lavage helps in mesothelial cell recovery and resolution of EPS (Nakamoto, 2005). However, cessation of peritoneal dialysis with withdrawal of peritoneal catheter and initiation of hemodialysis should be considered in these patients (Moinuddin et al., 2015). Anti-tubercular therapy or surgery may help in resolution of cocoon formation in tubercular abdominal cocoon (TAC). In drug related EPS, discontinuation of drug may be beneficial (Mandavdhare et al., 2017).

C) Pharmacological management

Immunosuppressants

Corticosteroids are the drugs of choice in the management of early inflammatory stages of EPS. Steroids possibly act by suppressing inflammation, preventing fibrin deposition and collagen synthesis and maturation (Habib, Betjes, Fieren, Boeschoten, Abrahams, Boer and Sevaux (2011).

Immunomodulators

Immunomodulators such as selective estrogen receptor modulators (e.g tamoxifen) in combination with steroids are effective in the management of abdominal cocoon related to peritoneal dialysis. Its action may be mediated by non-ER dependent mechanisms including modulation of TGF-beta related pathways. Tamoxifen monotherapy is administered at doses of 10-40 mg/day (Mandavdhare et al., (2017).

Inhibitors of mechanistic target of rapamycin- mTOR were suggested to benefit patients who had developed EPS after renal transplant. However, these drugs are usually used in combination with steroids and/or surgery (Mandavdhare et al., (2017).

D) Infrared therapy

The far-infrared therapy, through anti-inflammatory action and stimulation of neo angiogenesis and improved endothelial function may improve EPS.

E) Surgical treatment

In case of abdominal cocoon or EPS, surgery plays both diagnostic and therapeutic role. The most widely used surgical procedure is complete resection of membrane (peritonectomy) and adhesiolysis.

Intestinal resection is performed in non-viable intestine but there are high chances of post-operative fistula formation (Mandavdhare et al., 2017).

Prevention

Prevention of epithelial-to-mesenchymal transition of the mesothelial cells of the peritoneum helps in prevention of EPS. Agents with anti-

fibrotic properties such as melatonin, rapamycin, pirfenidone, rosiglitazone, N-acetyl cysteine, colchicine, angiotensin inhibition, thalidomide, biocompatible PD solutions may help in prevention of EPS but require experimental evidence for clinical use. Anti tubercular therapy in patients with tubercular abdominal cocoon helps to avoid surgery (Mandavdhare et al., 2017).

Nursing management

Nursing management of patients with intestinal obstruction secondary to abdominal cocoon includes gastric decompression, measurement of nasogastric output, assessing fluid and electrolyte balance, replacement of depleted fluid and electrolytes, maintaining nutritional status, institution of antibiotics and meeting their psychological and informational needs. Any imbalances in intake and output, worsening of pain or abdominal distention, and increased nasogastric output are notified immediately for timely interventions. If the patient's condition does not improve, patient is prepared for surgery.

Mr. K who was admitted with symptoms of subacute intestinal obstruction secondary to abdominal cocoon was kept on nil per oral (NPO) and nasogastric tube connected to dependent drainage during which large volume of fecal contents were drained. He was started on parenteral (intravenous) fluid, antibiotic therapy and was conservatively managed. Nursing management of Mr. K is discussed using nursing process approach.

Nursing process:

1. Nursing diagnosis

Acute pain (abdomen) related to inflammation and constriction of the diseased bowel.

Expected outcomes

Patient uses appropriate pain management strategies and reports satisfactory pain control with decrease in pain intensity and increased comfort such as baseline levels for pulse, blood pressure, respirations, relaxed muscle tone or posture.

Nursing interventions

- Assessed patient's intensity of pain, aggravating and relieving factors
- Auscultated for return of bowel sounds and reduction in ascites (abdominal girth) after gastric decompression.
- Encouraged patient to engage in familiar diversional activities, relaxation techniques or psycho social support systems as tolerated.

2. Nursing diagnosis

Imbalanced nutrition less than body requirement related to therapeutic restriction of dietary intake and loss of nutrients through gastric decompression.

Expected outcomes

Patient expresses understanding of the required therapeutic restriction and willingness to comply with the restrictions.

Nursing interventions

- Inserted nasogastric tube (NG tube) for GI tract (decompression) and connected to dependent drainage.
- Maintained precautionary measures to prevent displacement of the tube.
- Administered parenteral fluids to meet the nutritional requirements.
- Maintained accurate record of fluid intake, urine output and drainage of gastric contents was and reported to physicians for necessary interventions like parenteral fluid therapy.

3. Nursing diagnosis

Risk for deficient fluid volume related to restricted intake and loss of fluids through decompressive therapy.

Expected outcomes

Patient maintains euvolemic status as evidenced by systolic blood pressure > 90 mm Hg or patient's baseline of pulse rate and strength, normal skin turgor, urine output > 30 ml per hour and moist mucous membranes.

Nursing interventions

- Assessed the patient's hydration status (Skin turgor, mucous membrane, weight, blood pressure and pulse rate and strength).
- Observed him for orthostatic changes.
- Anticipated the need for intravenous fluid therapy and advocated to the physician about patient's fluid needs.
- Administered intravenous fluids as per plan and monitored intake and output.

4. Nursing diagnosis

Constipation related to absence of intestinal peristalsis secondary to cocoon constricting the bowel.

Expected outcomes

Patient maintains passage of soft, formed stool at a frequency perceived as "normal" by the patient and states relief from discomfort of constipation.

Nursing interventions

- Inserted nasogastric tube to remove fluid and gas from the upper GI tract and connected to dependent drainage for decompression.
- Implemented precautionary measures to avoid accidental displacement of the tube and ensured patency of the nasogastric tube.
- Recorded the amount, color, and type of nasogastric drainage and assessed for decrease in abdominal girth.
- Assessed improvement (eg, return of normal bowel sounds, decreased abdominal distention, subjective improvement in abdominal pain and tenderness, passage of flatus or stool).

5. Nursing diagnosis:

Impaired oral mucous membrane related to restricted oral intake and dehydration

Expected outcomes

Patient's mucous membrane is maintained moist as evidenced by lack of dryness, discomfort and bad odor.

Nursing interventions

- Provided regular oral and nasal hygiene to avoid discomfort and irritation.
- Inspected daily for skin irritation, and the nasal tape is changed periodically to prevent skin excoriation.
- Provided water for gargles whenever he reported dryness of mouth and throat.
- Suggested use of chewing gum or sugarless candies to assist in relieving dryness of oral mucosa.

6. Nursing diagnosis

Anxiety related to sudden change in health status characterized by increasing sense of powerlessness and expressed concerns.

Expected outcomes

Mr. K uses appropriate relaxation techniques and exhibits decrease in anxiety as manifested by relaxed posture and expression of psychological relief.

Nursing interventions

- Assessed his level of anxiety and coping abilities.
- Developed and maintained therapeutic relationship with him and spent time to listen to his expression of anxious feelings.
- Maintained a quiet environment with privacy, respecting his need for dignity and confidentiality.
- Suggested diversion through television, radio, games to divert the mind from stress and anxiety.
- Provided explanation about disease condition and explained about ongoing treatment to involve him in decision making and to promote a sense of control.

CONCLUSION:

After 2 days of decompression and Nursing care aimed at providing optimal relief of his physical and psychological symptoms, his signs of intestinal obstruction got resolved and expressed relief from his anxiety pertaining to onset of unknown condition. His nutritional status, fluid and electrolyte levels, signs of recovery from intestinal obstruction were continuously monitored. He was started on oral feeds and was discharged with medical advice for follow up on OPD basis.

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