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Review Article

Abdominal cocoon: a surgical challenge

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ABSTRACT

Encapsulating peritoneal sclerosis (EPS) or sclerosing peritonitis best described as abdominal cocoon is a complex syndrome characterized by formation of a thick fibrocollagenous membrane that totally encloses the small intestinal loops. The etiology is varied and multifactorial. This complex disease is usually associated with continuous peritoneal dialysis (PD), tuberculosis, post renal transplant, or with long term use of beta blockers. A few cases may even be idiopathic in origin. The pathogenesis is attributed to conversion of epithelial cells from the peritoneal lining to mesenchymal cells leading to a formation of a thick membrane. The clinical features range from abdominal pain due to altered gut motility to frank features suggestive of intestinal obstruction. Making a pre-operative diagnosis is the biggest challenge in abdominal cocoon. However computerized tomography provides enough information to arrive at a diagnosis. Surgical intervention continues to remain the final option in an unremitting intestinal obstruction despite the role of conservative therapy using drugs proposed for treatment. The focus of management is early diagnosis and treatment in order to halt the progression to a frank cocoon. The paper reviews the etiopathogenesis and management of this intricate condition.

Keywords: Cocoon management, Encapsulating, Peritoneal sclerosis

INTRODUCTION

Encapsulating peritoneal sclerosis (EPS) or abdominal cocoon is one of the uncommon cause for intestinal obstruction. It is a fibrotic encapsulation of the small bowel loops in majority of cases. It was first described by Owtschinnikow in 1907 as peritonitis chronic fibrosa encapsulate. It was later termed as abdominal cocoon by Foo in 1978.[1,2]

As the incidence of renal failure requiring peritoneal dialysis has increased over a period of time, this condition now is commonly encountered by the surgeon.[3] Awareness of this condition is therefore essential for a prompt diagnosis and optimum management.

ETIOPATHOGENESIS

The pathogenesis of this condition is complex. The two-hit hypothesis is the most commonly accepted theory for EPS seen in a PD patient.[3-5] The first hit comprises of pre-disposing factors such as PD.[5,6] This leads to two sets of changes in the peritoneal lining. Morphologic changes comprising of mesothelial denudation, angiogenesis, interstitial fibrosis and vascular sclerosis. The functional changes comprise of reduced absorptive surface area of the peritoneal lining. The net result is increased fibrin production and transformation of myofibroblast from the mesothelial cells leading to increased fibrosis. The second hit comprises of events such as recurrent peritonitis and bleeding from the PD catheter.[6,7]

Various inflammatory cytokines are produced which lead to amplification of fibrin production and reduced breakdown. The net result is fibroblast proliferation, peritoneal fibrosis, and intestinal adhesions eventually leading to capsule or cocoon formation (Figure 1).[7,8]

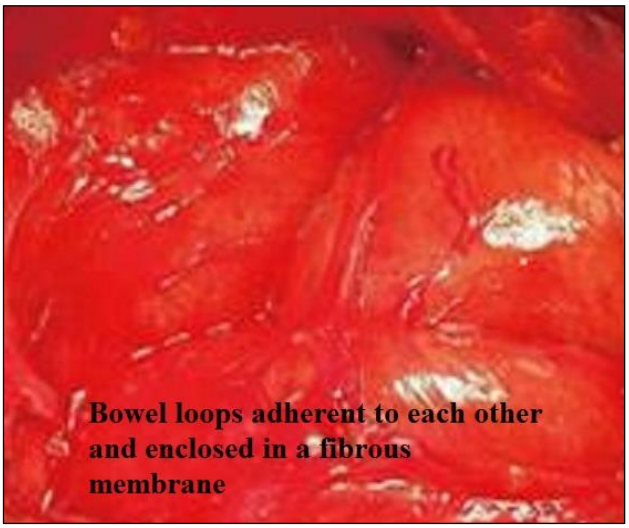

Figure 1: Intraoperative appearance of an abdominal cocoon.

A variety of causes have been attributed to the etiology. The commonest are peritoneal dialysis related, abdominal tuberculosis, post renal transplantation, use of povidone iodine, long term use of beta blockers and malignancies such as GIST or neuroendocrine tumors. Surprising associations of abdominal cocoon are cryptorchidism, omental hypoplasias, malrotation of the gut and Treitz's hernia have also been described.[7,8]

CLASSIFICATION

Two variants are usually seen, primary and acquired (secondary).[9] Primary are seen in adolescent girls in tropical and subtropical countries. It is attributed to retrograde menstruation.[10] Secondary is the commonest effectively causing peritoneal irritation and inflammation leading to peritoneal fibrogenesis. Abdominal cocoons are also classified on the basis of their contents.[3]

Type one which contains a part of the small intestine. Type two contains the entire small intestine. Type three contains colon or viscera. Recently type 4 has been described which is associated with neuroendocrine tumors which envelope the entire peritoneal lining of the peritoneal cavity.

Histopathology of the membrane shows a lamellar matrix of fibrin suggestive of its origin from fibrin deposition which took place during the first hit. Blood and inflammatory cell infiltrate when seen is suggestive of recurrent peritoneal irritation best described by the second hit.[3,4]

CLINICAL PRESENTATION

The hall mark of EPS is the intermittent nature of the presentation itself.[11,12] The initial symptoms are due to alteration in the gut transit and motility. These usually give rise to intermittent episodes of severe colicky pain devoid of features of infection. As the disease progresses, more intestinal loops get involved thereby causing dense adhesions leading to multiple segments of obstruction. The eventual clinical picture is typical of small intestinal obstruction. A careful history is therefore pivotal in assessing causes of suspected small intestinal obstruction. A history of previous PD or transplantation associated with obstructive symptoms should clinch the diagnosis of EPS. However, on the Asian subcontinent abdominal tuberculosis is a very common cause for a similar pathology.[12,13] Therefore a previous history of tuberculosis associated with signs of obstruction are highly suggestive of EPS of tuberculous origin. This should raise caution on the part of the surgeon with respect to complex anticipated intra-abdominal filings at laparotomy rendering limited options for surgical treatment.[12]

DIAGNOSIS

A variety of radiological investigations can aid in the diagnosis of EPS. Plain abdominal X-ray will reveal dilated small bowel loops, multiple air fluid levels, clumping of bowel loops, peritoneal and bowel wall calcifications.[13,14]

USG

Ultra-sonography will reveal clumped bowel loops and ascites with loculations. A variety of patterns have been described in EPS. Clumped bowel loops attached by a narrow mesentery appearing in a cauliflower like fashion is a typical finding. Sand-witch sign due to presence of an echogenic membrane around the bowel loops may also be seen. However, the radiologist needs to be fully aware of this peculiar condition in order to propose a provisional diagnosis of EPS on ultra sound.[14,15]

Contrast enhanced computerized tomography (CECT)

CECT is the diagnostic investigation for EPS. It continues to be the gold standard for diagnosis.[15-18]

Findings on CECT are

- Ascites.
- Loculated fluid collections.
- Peritoneal and mesenteric thickening.
- Small bowel thickening.
- Lymphadenopathy.
- Mural or peritoneal calcification especially around the capillaries extending into the visceral and muscular layers.

- Ginger and bread man sign which comprises of tethered small bowel loops because of the retraction of the mesentery.
- Cauliflower sign similar to that seen on ultra sound due to clumped bowel loops attached to by a narrow mesentery.

CECT findings in EPS secondary to chronic PD are more typical.[17,18] They include:

- Peritoneal thickening.
- Located fluid collection.
- Calcification.
- Congregated small bowel loops in the center of the abdomen
- Peritoneal enhancement
- Helical disposition of small bowel loops displaced anteriorly suggestive of the typical helix sign.

CECT has a distinct advantage of ruling out majority of other differential diagnosis.[19]

MDCT

MDCT is more advantageous as it shows the extent of the disease along with subtle radiologic findings as well as helps in formulating a plan for surgery by virtue of contribution of coronal and sagittal reformatted images.[19,20]

MRI

There is no significant difference between the advantages of MRI and CT.[21] However the encasing membrane may be more obvious with an MRI with a definitive advantage of no exposure to ionizing radiations.

Management

Management of EPS is a complex issue which requires a proper understanding of the etiopathogenesis and stage at presentation of the patient.[22]

With respect to chronic PD as the etiology, a number of risk factors have been identified which point towards the development of EPS. These include age, repeated attacks of peritonitis, type of peritoneal fluid used, faster peritoneal membrane transport, age at which PD was started, diminishing ultra-filtration and renal transplantation.[22]

The aims of treatment in such cases are:

- Cessation of peritoneal dialysis
- Shifting to hemodialysis
- Removal of the PD catheter which may perhaps slow down the process of EPS.

Medical treatment comprises of three group of drugs:

Steroids

These have immunosuppressive and anti-inflammatory properties. When combined with an immunomodulatory agent such as tamoxifen it has benefial effects in few patients especially in post renal transplant EPS.[23]

sTOR (e.g: Everolimus)

This comprises of inhibition of mechanistic targetting of rapamycin receptors. The mechanism of EPS in post renal transplant patients is by virtue of calcineurin inhibitors such as cyclosporin and tacrolimus. Therefore, use of everolimus may be beneficial especially in post renal transplant EPS patients. Mycophenolate mofetil and azathioprine may be useful in a few cases.[23]

Tamoxifen

Belongs to the group of SERMs. It is specifically effective in PD related EPS. The mechanism of action is mediated by non-estrogen receptor dependant mechanisms comprising of modulation of beta TGF pathways. The main advantage of tamoxifen is that it has no immunosuppressive effect.[24]

Medical management has a very limited role in non-peritoneal dialysis EPS patients. Cases related to long term beta blocker usage can be managed by cessation of beta blocker therapy.[22]

However, on the Asian subcontinent abdominal tuberculosis continues to be the commonest cause for EPS in a few cases. Commencement of anti-tuberculosis treatment therefore is the main stay of management of such cases.[27] However quite a few of these patients may progress to unremitting obstruction necessitating surgical intervention.

SURGICAL TREATMENT

This is the biggest challenge in EPS. It has two-fold purpose viz. diagnostic, where diagnosis of a cocoon is made and therapeutic where in resection of membrane and adhesiolysis can be done.[28] Irrespective of the etiology of EPS, adhesiolysis continues to pose the biggest surgical challenge to the operating surgeon.[29,30] Attempts at separating the membrane of the adherent loops may cause serosal damage and many a times may cause perforations in the bowel.

Despite meticulous technique, perforations are a common occurrence during the course of adhesiolysis in EPS.[31,32] Exteriorization is the best and the safest option in such cases in order to avoid the development of anastomotic leakage and septicemia. No heroic attempt at resection and anastomosis should be contemplated during such cases. This approach is especially relevant in abdominal cocoons due to tuberculosis.[32]

CONCLUSION

EPS continues to be the biggest challenge to the gastrointestinal surgeon. Carefully weighted history and proper interpretation of the radiological images can help in diagnosing EPS at an early stage. A trial of conservative management can be offered to patients who present with mild symptoms especially in post PD cases.

Surgical intervention is limited to intractable obstruction. Meticulous removal of the fibrous membrane, with adhesiolysis is the main stay of surgical intervention. In the event of bowel perforation, exteriorization should be the standard practice.

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