Title- Tuberculous Abdominal cocoon in childerns– A Single Centre Study in a remote village

Abstract

Background -Amongst the numerous causes of intestinal obstruction listed in the literature, Sclerosing Encapsulating Peritonitis also called Abdominal Cocoon (AC) is one of the rarest entities. Its characteristic feature is a thick fibrotic membrane encasing varying lengths of the small and large gut in a cocoon. In India, there is an increasing incidence of tuberculosis more so in the rural areas.

Aims and objectives- The aim of this study was to investigate the clinical presentation and evaluate the operative findings. We also evaluated the outcomes and response to ATT in all the patients diagnosed with this condition.

Material and Methods - This study was carried out at M.M. Institute of Medical Sciences and Research, Mullana, from Aug 2013 –July 2016 in the departments of Paediatric and General Surgery. This is a retrospective study. A total of 17 patients were included in the study diagnosed as abdominal cocoon.

Results – total 17 patients presented with features of acute intestinal obstruction. The average age of patients was 15.3 years (range 9 years to 16 years). There were 14 females and 3 males. All patients presented with pain abdomen, bilious vomiting, constipation and abdominal distention. All patients were operated in our hospital. In follow-up all patients are without any recurrence of tuberculosis or intestinal obstruction.

Conclusion:- A strong clinical suspicion, sonological and computed tomography scan findings helps in arriving at a preoperative diagnosis Tuberculous AC has a strong preference to females. Surgery is the mainstay of treatment followed by Antitubercular drugs.

Key words- Intestinal obstruction; cocoon abdomen; Sclerosing Peritonitis; Computed tomography scan; perforation; Surgery

Introduction –

In the western developed world, it is rare to have peritoneal tuberculosis. This disease is generally found in those who have migrated from countries either rampant in tuberculosis or having a high incidence of AIDS. Abdominal tuberculosis (TB) is the sixth commonest extra-pulmonary TB. The gastrointestinal mesenteric lymph nodes, blood and the fallopian tubes are the sites of spread of the disease to the peritoneum. [1] Varying lengths of small and large gut loops get entrapped in a thick fibrocollagenous membrane with inflammatory infiltrate called a cocoon. This results in an acute or chronic intestinal obstruction in the patient. [2] Sclerosing encapsulating peritonitis can be primary (idiopathic) or secondary to various causes like CAPD, TB or use of practolol. Various hypotheses have been proposed to explain the reason for the primary form, most likely it is due to subclinical peritonitis that results in a cocoon formation. [3] It is very common to find loops of small gut encased in a thick whitish membrane. Sometimes other parts are also involved like the ascending colon, caecum, appendix and the ovaries. [4] Because of the nonspecific features at time of presentation, it is difficult to make a definitive diagnosis of abdominal cocoon pre-operatively. The diagnosis is generally missed and is only done at the time of laparotomy. [5] Surgery is the treatment of choice. Freeing the encased bowel along with release of any adhesions present relieves the obstruction in the patient. The aim of this study was to investigate the clinical presentation, evaluate the operative findings and see the response to ATT in all the patients who were diagnosed with this condition

Material and methods:-

This study was conducted in MMIMSR, Mullana. Haryana from August 2013 to July 2016 for a period of 3 years. The average age of patients was 15.3 years (range 9 years to 16 years). All patients diagnosed with AC clinically, by radiological imaging (ultrasound abdomen/ CECT abdomen), intraoperative findings and histopathological confirmation were included. All patients were operated after adequate preoperative resuscitation with intravenous fluids and correction of electrolyte imbalances. CECT was possible in only 1 out of 17 patients as this female patient presented with features of SAIO and was operated only when conservative management failed to relieve her obstruction after 4 days.

Results:-

The total number of patients was 17, out of them, 14 were females and 3 were males. All patients were from rural area predominately from low socio economic strata of society. 4 patients were already on treatment for pulmonary Koch’s ranging from 3weeks to 4 months but were not regular. Monteux test was performed on all patients based on high index of suspicion and was positive in 8 out of 17 patients (47.1%) ESR was high in all patients. Contact with TB was present in 3 out of 17 patients (17.6%)

All patients presented with features of small bowel obstruction. One had evidence of SAIO and was conservatively managed for 4 days but later was operated as CECT diagnosed a cocoon abdomen(figure-1 and 2). In 2 out of 17 patients (11.8%) an abdominal lump was palpable in the central abdomen. Abdominal x-rays showed multiple air fluid levels and sonography showed thickened and matted gut loops. The operative findings showed Type 1 AC (partial encasement of small bowel) in 9 females and 2 male patients, a total of 11 out of 17 patients (64.7%) and Type 2 (complete encasement from DJ flexure to IC junction) in 5 females and 1 male patient, a total of 6 out of 17 patients (35.3%) (figure- 3-5). 6 females and 3 male patients, a total of 9 patients had enlarged mesenteric lymph nodes (52.9%) and 4 female patients had tubercles all over the omentum and mesentery (23.5%). 1 female patient had a stricture at terminal ileum (5.8%) for which a strictroplasty was done.1male patient had gangrene of terminal ileum (5.8%) because of thick membrane compromising the blood supply, for which ileostomy was done which was closed after 3 months on anti tuberculous therapy.

Ascetic fluid ADA was elevated in 5 patients (29.4%) and it confirmed tuberculosis. Omental and lymph node biopsies were taken and sent for histopathological examination along with the fibrotic membrane (figure- 6). After histopathological confirmation of tuberculosis, all patients were started on ATT postoperatively for duration of 1 year. None of the 17 patients operated for cocoon abdomen had any post operative complications like faecal fistula, adhesive intestinal obstruction and wound dehiscence. All had a smooth recovery. Periodic follow up was satisfactory. Patients gained weight and no recurrence was seen in any patient.

DISCUSSION:-

Gastrointestinal tract TB affects the ileum and the caecum in 75% of cases. Ingestion of infected sputum of pulmonary tuberculosis causes intestinal tuberculosis. The other organs that can get affected include omentum, liver, spleen, pancreas, adrenals and the female genital tract. A review of the literature shows that the abdomen is the sixth commonest site of extra-pulmonary TB tuberculosis. Pasteurization of milk has considerably reduced the occurrence of primary intestinal TB and made it a rare disease in the western world. [6]

The common causes of intestinal obstruction include intestinal adhesions, bands and obstructed hernia. Abdominal cocoon also known as SEP is a known cause of intestinal obstruction although it is a rare condition. Encasement of varying lengths of small bowel in a fibrocollagenic cocoon like sac is a pathognomonic feature of this condition. [6] 3 types of abdominal cocoon are described depending on the extent of the encasing membrane covering the bowel loops. Type 1 abdominal cocoon is partial encapsulation of the intestine, Type 2 is complete encapsulation of the entire intestine and Type III cocoon syndrome is the encasement of the entire intestine along with other intra abdominal organs like the appendix, caecum, ascending colon and ovaries. The clinical presentation of these patients may be acute, sub acute, or chronic with signs and symptoms suggestive of small bowel obstruction like colicky pain abdomen bilious vomiting, abdominal distention and obstipation. Sometimes there is also a palpable abdominal mass. Malnutrition and weight loss is also evident in most cases. [7]

The different modes of presentation closely resemble the presentations reported in other series. Apart from presenting to the surgical emergency with features of intestinal obstruction, most of them also had complaints that suggested tuberculosis like low grade fever, weight loss, anorexia and menstrual abnormalities ranging from several weeks to months. Symptoms and signs have been reported similarly by other authors with variable percentages of prevalence. [8] Literature reviewed in adults revealed 118 cases of SEP. Majority were male (68%) and the mean age of these patients was 39 years. Abdominal pain was the chief complaint in 72.0%, abdominal distention in 44.9% and a palpable abdominal lump in 30.5%. The encasing membrane was excised surgically in almost all the patients (99.2%). There were no post op complications (88.1%). [9] Without doubt the CT scan of the abdomen is currently the most useful radiological method for diagnosis of SEP. The obstruction is due to extensive fibrosis that will cause shortening and retraction of the root of the mesentery. The bowel loops get clumped together. This in turn causes gut dysfunction. The “gingerbread man” sign on CT is due to retraction and clumping of the bowel loops. A combination of clinical presentations and CT abdomen will further enhance the chances of a preoperative diagnosis. Small bowel loops conglomerating in the midline with a dense mantle encasement without peripheral contrast uptake is a characteristic. [10]

Multi detector CT scan has played a vital role in detecting minute perforations of the gut in blunt injury abdomen. [11] Recently many reports have emphasized the role of MDCT in the diagnosis of AC. Coronal, saggital and axial reconstructions have shown upto 3mm thick sac surrounding the small bowel loops suggestive of a cocoon. MDCT also helps in telling the extent of the disease thereby helping the surgeon to plan better. [12]

There is a lot of debate regarding the management of SEP. Many series have reported successful conservative management in mildly symptomatic cases. In severe cases of SEP, or where a cocoon is encountered at laparotomy, it is necessary to resort to surgical treatment. Depending on the condition of the patient various techniques like membrane excision + adhesiolysis, resection + anastomosis, resection + anastomosis + protective enterostomy can be used alone or in combination. As the operation can be quite a challenge, it is important to be slow and meticulous. Whenever possible, it is better to avoid doing an intestinal resection. This not only increases the morbidity and mortality but also increases the chances of dreaded complications like anastomotic leaks and short bowel syndrome. [13]

Laparoscopic management is still unclear. Very few cases have been reported in literature regarding the role of laparoscopy in the management of SEP. Few authors have performed successful membrane excision and adhesiolysis by laparoscopy. A definite advantage of laparoscopy is that it can be used for diagnostic purposes. [14] We did not attempt any laparoscopic management in our centre as most all our patents were young and sick children. All except 1 patient presented to the surgical emergency in total intestinal obstruction. We also have to admit that we do not have adequate experience in the laparoscopic management of such cases.

Histopathological examination of the membrane showed thickened vascular fibrocollagenous tissue with chronic inflammatory reaction evidenced by lymphocytic and plasma cell infiltrates and sometimes even epithelioid cell granulomas. In our study, 2 patients had a positive biopsy for TB of greater omentum and 3 patients had positive evidence of TB on mesenteric lymph node biopsies.

Key points –

* AC secondary to TB does not generally respond to conservative treatment.
* Surgery is the preferred treatment. Lysis of the encasing membrane and freeing the bowel loops is required.
* Other additional procedures like strictroplasty, simple closure of perforation and an enterostomy may be needed.
* Additionally, presence of tubercles over omentum and gut and caeseating mesenteric lymph nodes, mesenteric abscess may be encountered suggesting a tubercular etiology.

Conclusion- tuberculosis as a cause of AC is becoming a rather common entity. It is mostly seen in females coming from rural areas. It is difficult to diagnose preoperatively. A strong clinical suspicion is important. Surgery is the preferred choice. ATT for 1 year is prescribed after histopathological confirmation. The condition is unlikely to occur thereafter.

References:-

1. Singal R, Gupta S, Gupta S. Primary abdominal tuberculosis presenting as peritonitis in a young child-managed surgically. Asian Pac J Trop Med 2012; 5: 413-15.
2. [Sharma D](http://www.ncbi.nlm.nih.gov/pubmed/?term=Sharma%20D%5BAuthor%5D&cauthor=true&cauthor_uid=24055916), [Nair RP](http://www.ncbi.nlm.nih.gov/pubmed/?term=Nair%20RP%5BAuthor%5D&cauthor=true&cauthor_uid=24055916), [Dani T](http://www.ncbi.nlm.nih.gov/pubmed/?term=Dani%20T%5BAuthor%5D&cauthor=true&cauthor_uid=24055916), [Shetty P](http://www.ncbi.nlm.nih.gov/pubmed/?term=Shetty%20P%5BAuthor%5D&cauthor=true&cauthor_uid=24055916). Abdominal cocoon-A rare cause of intestinal  obstruction. [Int J Surg Case Rep](http://www.ncbi.nlm.nih.gov/pubmed/?term=Abdominal+cocoon%E2%80%94A+rare+cause+of+intestinal+obstruction)  2013; 4(11):955-7.
3. Akbulut S. Accurate definition and management of idiopathic sclerosing encapsulating peritonitis. World J Gastroenterol 2015; 21:675–87.
4. [Yang CS](http://www.ncbi.nlm.nih.gov/pubmed/?term=Yang%20CS%5BAuthor%5D&cauthor=true&cauthor_uid=27073795), [Kim D](http://www.ncbi.nlm.nih.gov/pubmed/?term=Kim%20D%5BAuthor%5D&cauthor=true&cauthor_uid=27073795). Unusual intestinal obstruction due to   idiopathic sclerosing encapsulating peritonitis: a report of two cases and a review. [Ann Surg Treat Res](http://www.ncbi.nlm.nih.gov/pubmed/?term=Unusual+intestinal+obstruction+due+to+idiopathic+sclerosing+encapsulating+peritonitis%3A+a+report+of+two+cases+and+a+review)  2016; 90(4):231-4.
5. Li N, Zhu W, Li Y, Gong J, Gu L, Li M, et al. Surgical treatment and perioperative management of idiopathic abdominal cocoon: single-center review of 65 cases. World J Surg 2014; 38(7):1860–7.
6. Singal R and Mittal A. Plasted abdominal tuberculosis presenting as an obstruction. J Infect Dev Ctries 2013; 7(7):561-63.

# [Wei B](http://www.ncbi.nlm.nih.gov/pubmed/?term=Wei%20B%5BAuthor%5D&cauthor=true&cauthor_uid=19217609), [Wei HB](http://www.ncbi.nlm.nih.gov/pubmed/?term=Wei%20HB%5BAuthor%5D&cauthor=true&cauthor_uid=19217609), [Guo WP](http://www.ncbi.nlm.nih.gov/pubmed/?term=Guo%20WP%5BAuthor%5D&cauthor=true&cauthor_uid=19217609), [Zheng ZH](http://www.ncbi.nlm.nih.gov/pubmed/?term=Zheng%20ZH%5BAuthor%5D&cauthor=true&cauthor_uid=19217609), [Huang Y](http://www.ncbi.nlm.nih.gov/pubmed/?term=Huang%20Y%5BAuthor%5D&cauthor=true&cauthor_uid=19217609), [Hu BG](http://www.ncbi.nlm.nih.gov/pubmed/?term=Hu%20BG%5BAuthor%5D&cauthor=true&cauthor_uid=19217609), [Huang JL](http://www.ncbi.nlm.nih.gov/pubmed/?term=Huang%20JL%5BAuthor%5D&cauthor=true&cauthor_uid=19217609). Diagnosis and treatment of abdominal cocoon: a report of 24 cases. [Am J Surg](http://www.ncbi.nlm.nih.gov/pubmed?cmd=Retrieve&list_uids=19217609) 2009; 198(3):348-53.

1. [Mukhopadhyay A](http://www.ncbi.nlm.nih.gov/pubmed/?term=Mukhopadhyay%20A%5BAuthor%5D&cauthor=true&cauthor_uid=25177599), [Dey R](http://www.ncbi.nlm.nih.gov/pubmed/?term=Dey%20R%5BAuthor%5D&cauthor=true&cauthor_uid=25177599), [Bhattacharya U](http://www.ncbi.nlm.nih.gov/pubmed/?term=Bhattacharya%20U%5BAuthor%5D&cauthor=true&cauthor_uid=25177599). Abdominal tuberculosis with an acute abdomen: our clinical experience. [J Clin Diagn Res](http://www.ncbi.nlm.nih.gov/pubmed/25177599) 2014; 8(7):NC07-9.
2. Machado NO. Sclerosing Encapsulating Peritonitis: Review. Sultan Qaboos University Medical Journal 2016; 16(2):e142-e51.
3. Tannoury JN, Abboud BN. Idiopathic sclerosing encapsulating peritonitis: abdominal cocoon. World J Gastroenterol 2012; 18(17):1999–04.
4. [Singal R](https://www.ncbi.nlm.nih.gov/pubmed/?term=Singal%20R%5BAuthor%5D&cauthor=true&cauthor_uid=23542824), [Gupta R](https://www.ncbi.nlm.nih.gov/pubmed/?term=Gupta%20R%5BAuthor%5D&cauthor=true&cauthor_uid=23542824), [Mittal A](https://www.ncbi.nlm.nih.gov/pubmed/?term=Mittal%20A%5BAuthor%5D&cauthor=true&cauthor_uid=23542824), [Gupta A](https://www.ncbi.nlm.nih.gov/pubmed/?term=Gupta%20A%5BAuthor%5D&cauthor=true&cauthor_uid=23542824), [Singal RP](https://www.ncbi.nlm.nih.gov/pubmed/?term=Singal%20RP%5BAuthor%5D&cauthor=true&cauthor_uid=23542824), [Singh B](https://www.ncbi.nlm.nih.gov/pubmed/?term=Singh%20B%5BAuthor%5D&cauthor=true&cauthor_uid=23542824), et al. Delayed presentation of the traumatic abdominal wall hernia; dilemma in the management - review of literature. [Indian J Surg](https://www.ncbi.nlm.nih.gov/pubmed/?term=Delayed+Presentation+of+the+Traumatic+Abdominal+Wall+Hernia%3B+Dilemma+in+the+Management+%E2%80%93+Review+of+Literature)  2012; 74(2):149-56.
5. [Gadodia A](http://www.ncbi.nlm.nih.gov/pubmed/?term=Gadodia%20A%5BAuthor%5D&cauthor=true&cauthor_uid=21212192), [Sharma R](http://www.ncbi.nlm.nih.gov/pubmed/?term=Sharma%20R%5BAuthor%5D&cauthor=true&cauthor_uid=21212192), [Jeyaseelan N](http://www.ncbi.nlm.nih.gov/pubmed/?term=Jeyaseelan%20N%5BAuthor%5D&cauthor=true&cauthor_uid=21212192). Tuberculous abdominal cocoon. [Am J Trop Med Hyg](http://www.ncbi.nlm.nih.gov/pubmed/?term=tuberculous+abdomen+cacoon+%2C+gadodia+a)  2011;84(1):1-2.
6. Xu P, Chen LH, Li YM. Idiopathic sclerosing encapsulating peritonitis (or abdominal cocoon): a report of 5 cases. World J Gastroenterol 2007;13(26):3649–51.
7. Ertem M, Ozben V, Gok H, Aksu E. An unusual case in surgical emergency: Abdominal cocoon and its laparoscopic management. J Minim Access Surg. 2011;7(3):184–6.

Legends-

Figure-1 - Contrast enhanced computed tomography(CECT) – it revealed an encapsulated form and loops are encircled inside of it

Figure-2 – CECT is showing dilated loops and in between there are formations of encapsulation

Figure-3 –gross operative picture showing fibrosis whitish in colour forming abdominal cacoon

Figure-4 - gross picture revealed separation of the bands from the interloops

Figure-5 – gross thickened omentum and whitish nodules seen; b – operative area showed dilated and plasted intestinal bowel loops

Figure-6- histopathology picture showing tuberculosis of the omentum ; b- on histopathology of the mesenteric lymph node revealed tuberculosis cells