DOI: 10.15386/cjmed-725

TUBERCULAR ABDOMINAL COCOON IN CHILDREN – A SINGLE CENTRE STUDY IN REMOTE AREA OF NORTHERN INDIA

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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, especially in the rural areas.*

Aims and objectives. *The aim of this study was to investigate the clinical presentation and evaluate the operative findings of tuberculous AC. We also evaluated the outcomes and response to anti tuberculous treatment (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, Ambala, India between April 2013 – March 2016 in the Department of Pediatric Surgery. This is a prospective study. A total of 17 patients diagnosed with abdominal cocoon secondary to tuberculosis have been included in the study.*

Results. *A total of 17 patients presented to the emergency ward with features of acute intestinal obstruction. The average age was 15.3 years (range 9 years to 16 years). There were 14 females and 3 males. All patients presented with abdominal pain, bilious vomiting, constipation and abdominal distention. The patients were operated in our hospital and relieved of their obstruction. Based on their operative findings and after histopathological confirmation, patients were given ATT. In the follow-up, all patients did well, without recurrence of tuberculosis or intestinal obstruction.*

Conclusion. *Tuberculosis as a cause of childhood AC is rather common in developing countries and is potentially a fatal condition. A strong clinical suspicion, sonographic and computed tomography scan findings help establish a pre-operative diagnosis. Tuberculous AC has a strong prevalence in females. Surgery is the mainstay of treatment followed by anti-tuberculous drugs.*

Keywords: intestinal obstruction; cocoon abdomen; sclerosing peritonitis; gastrointestinal tuberculosis

Manuscript received: 22.09.2016
Received in revised form: 02.11.2016
Accepted: 08.11.2016
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Introduction

In the western developed world, it is rare to see peritoneal tuberculosis (TB). This disease is generally found in those who have migrated from countries either rampant in TB or having a high incidence of AIDS [1]. A quarter of the global burden of TB occurs in India, accounting for 20% of the 8.6 million new cases annually [2]. Almost 40% of India's population is under 15 years of age. The incidence of sputum microscopy smear positive TB in children <14 years old is about 0.6%-3.6% of all reported cases [3]. Children suffer more from extra pulmonary forms of TB than all other age groups. The high incidence in children in this country is due to poor nutrition, poor sanitation, neglect of early symptoms and signs, as well as drug resistant strains of Mycobacterium tuberculosis (10-51%) [4]. Abdominal TB is a rare manifestation of extrapulmonary TB, and a prevalence of around 3% has been noted previously in India. 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 [5]. Sclerosing encapsulating peritonitis can be primary (idiopathic) or secondary to various causes like continuous ambulatory peritoneal dialysis, TB or use of practolol. Various hypotheses have been proposed to explain the reason for the primary form. It is most likely due to subclinical peritonitis that results in a cocoon formation [6]. 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 [7]. Because of the nonspecific features at the time of presentation, it is difficult to make a definitive diagnosis of abdominal cocoon pre-operatively. The diagnosis is generally missed and is only made at the time of laparotomy [8]. 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 carried out in Department of Pediatric Surgery, M.M. Institute of Medical Sciences and Research, Mullana, Haryana, India from April 2013 to March 2016 for a period of 3 years. This is a prospective study. The average age of patients was 15.3 years (range 9 years to 16 years). All patients who presented to the emergency ward with clinical features of intestinal obstruction and diagnosed by imaging like ultrasound abdomen (USG) and contrast enhanced computed tomography (CECT) of the abdomen as AC were included in the study. Their intra-operative findings were noted and histopathological confirmation was obtained. Based on the history of chronic ill health, loss of weight and appetite, systemic symptoms of intermittent fever and cough, strong suspicion of tuberculosis was kept in mind. Moreover, all these children came from district Saharanpur, Uttar Pradesh, India, which is endemic in tuberculosis. Routine blood test like total leukocyte count, differential leukocyte count, renal function test, serum electrolytes and urine examination were performed to assess preoperative fitness for surgery. Montoux test and erythrocyte sedimentation rate (ESR) were also done keeping TB in mind. Chest X- rays were taken not only as routine preoperative measure but also to rule out or confirm associated pulmonary tuberculosis. Abdominal erect X-rays showed air fluid levels confirmatory of acute intestinal obstruction in all patients. USG and CECT were done which helped in a preoperative diagnosis. CECT of abdomen was possible in only 1 out of 17 patients as this female patient presented with features of subacute intestinal obstruction and was operated only when conservative management failed to relieve her obstruction after 4 days.

All patients were operated after adequate preoperative resuscitation with intravenous fluids and correction of electrolyte imbalances. At surgery the intra-operative findings were noted, such as presence of cocoon, type of cocoon, presence of ascitic fluid, tubercles and enlarged mesenteric lymph nodes. Ascitic fluid was sent for Adenosine deaminase (ADA) analysis. Cocoon was removed carefully and adhesions were removed. The fibrotic membrane, mesenteric lymph nodes and omental biopsy were sent for histopathological examination. The ATT regime was given for a period of 6 months – 2 months of intensive phase therapy (2 HRZE) and a maintenance phase for 4 months (4 HRE).

Results

The total no. of patients was 17, 14 females and 3 males. All patients were from a rural area predominately from low socio economic strata of society. Four patients were already on treatment for pulmonary Koch's ranging from 3 weeks 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 (100%). Contact with TB was present in 3 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 evidenced Type 1 AC, with partial encasement of small bowel in 9 females and 2 male patients, a total of 11 out of 17 patients (64.7%) (Figure 3) 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 4 and 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%). One female patient had a stricture at the terminal ileum (5.8%) for which a strictroplasty was performed. One male patient had gangrene of the 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. Ascitic 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 a duration of 6 months.

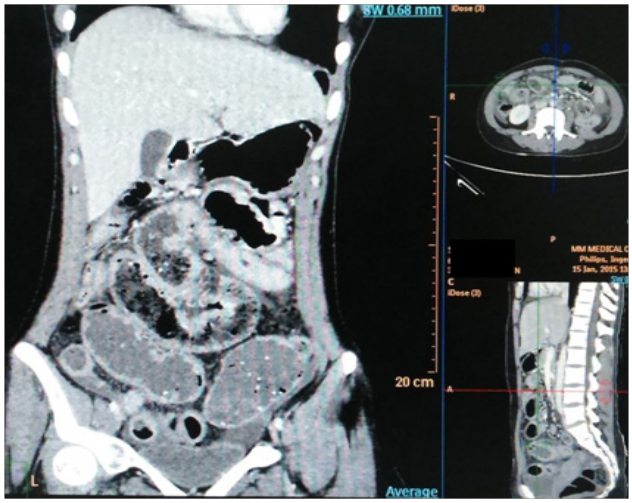

Figure 1. Contrast enhanced computed tomography (CECT) revealed an encapsulated form and loops are encircled inside of it.

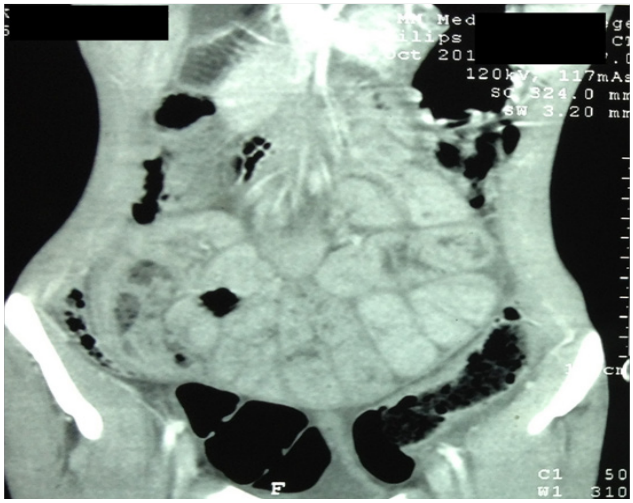

Figure 2. CECT is showing dilated loops and in between there are formations of encapsulation.

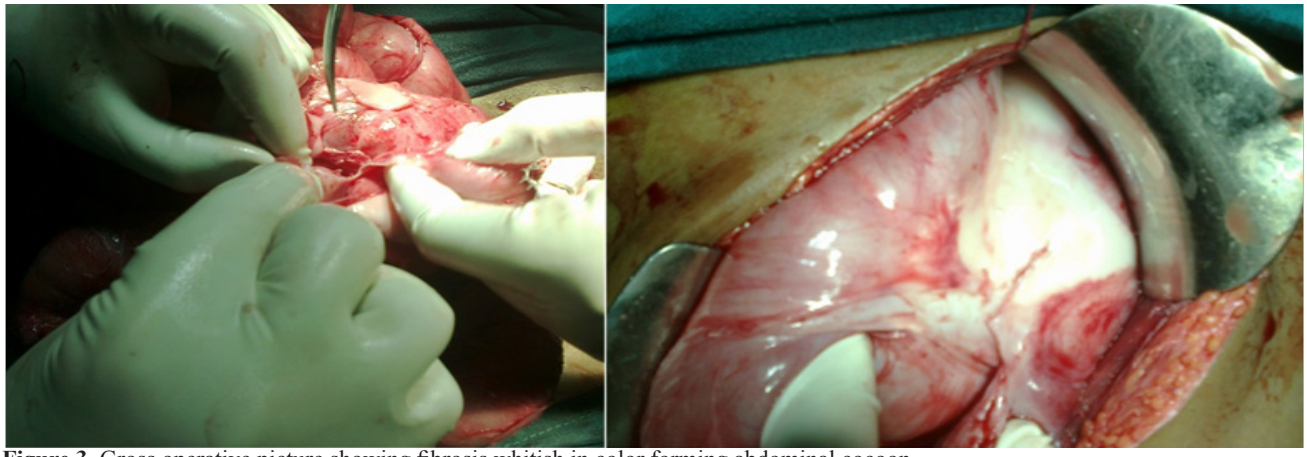

Figure 3. Gross operative picture showing fibrosis whitish in color forming abdominal cocoon.

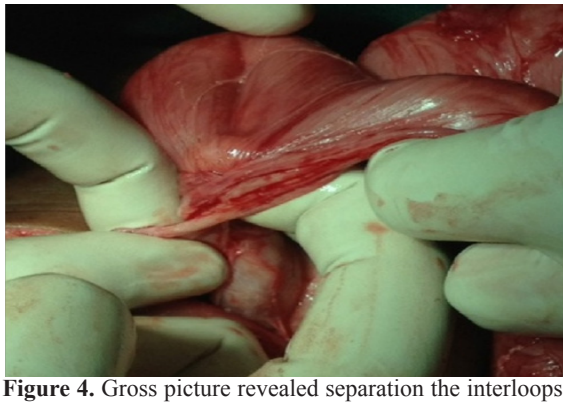

Figure 4. Gross picture revealed separation the interloops by adhesiolysis.

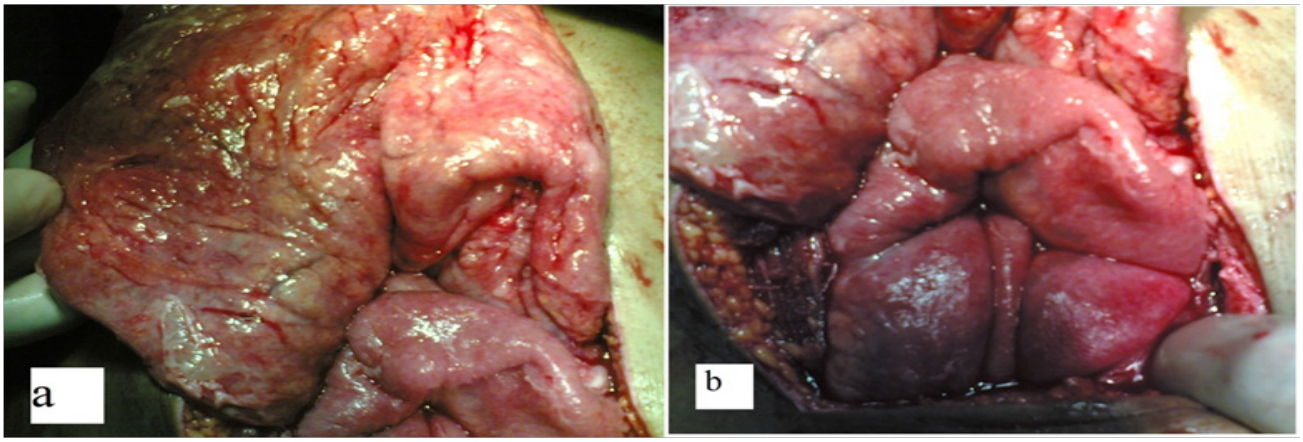

Figure 5. a) gross thickened omentum and whitish nodules seen; b) operative area showed dilated and plasted intestinal bowel loops

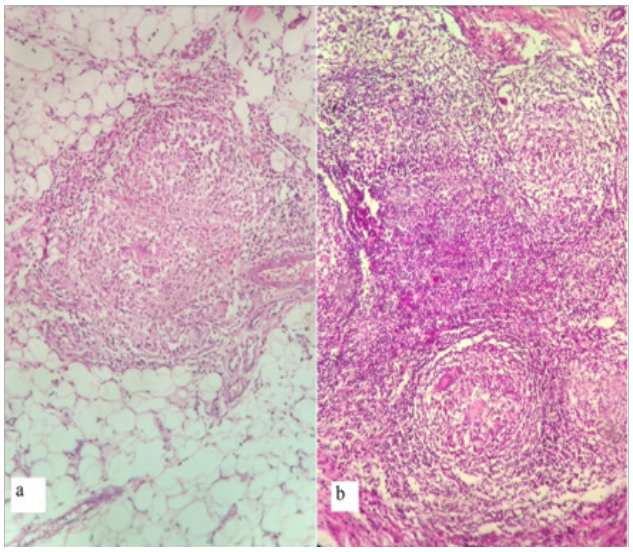

Figure 6. a) histopathology picture showing tuberculosis of the omentum; b) histopathology of the mesenteric lymph node revealed tuberculosis cells.

Evaluation and follow up

Post-operatively all the children were kept in pediatric intensive care unit (PICU) for the period of 5-7 days with intensive monitoring. I.V. fluids, blood and plasma were given as per requirements. Children were started on oral feeds on 5th post-operative day, sutures were removed on 8th post-operative day and children were discharged. Oral ATT was started in consultation with the pediatric medical team.

All the children were followed in pediatric surgical and medical OPD (Outpatient department) on a regular monthly basis for 6 months. The immediate and late complications were evaluated in the follow up. All patients were evaluated on the following:

1.Persistence of earlier symptoms of obstruction.

2.Toxaemia of TB; fever, loss of weight and loss of appetite.

3.Weight gain.

4.Investigations: ESR, CRP (C reactive protein), LFT (Liver function test).

None of the 17 patients operated for cocoon abdomen had any post-operative complications lsuch as fecal fistula, adhesive intestinal obstruction and wound dehiscence. All had a smooth recovery. Periodic follow up was satisfactory. Patients gained weight and no recurrence of TB was seen in any patient.

Discussion

TB is a public health problem worldwide. Between 19% and 43% of the world's population shows evidence of being infected with Mycobacterium tuberculosis.[9] Peritoneal TB is the most common form of abdominal TB after gastrointestinal TB and represents 0.4%-2% of cases of TB in general [1]. Abdominal cocoon also known as

SEP is a known cause of intestinal obstruction although it is a rare condition. The other organs that may be affected include the omentum, liver, spleen, pancreas, adrenals and the female genital tract. Encasement of varying lengths of small bowel in a fibrocollagenic cocoon-like sac is a pathognomonic feature of this condition [10]. Three 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, subacute, 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 [11].

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, such as 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 [12].

As per literature reviewed in pediatric age group, a total of 16 cases abdominal cocoon was revealed out of 118 cases of SEP. The 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%) [13]. Without doubt the CT scan of the abdomen is currently the most useful radiological method for the diagnosis of SEP. 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. Small bowel loops conglomerating in the midline with a dense mantle encasement without peripheral contrast uptake is a characteristic [14].

Multi detector CT (MDCT) scan has played a vital role in detecting minute perforations of the gut in blunt injury abdomen [15]. Recently many reports have emphasized the role of MDCT in the diagnosis of AC. Coronal, sagittal and axial reconstructions have shown up to 3 mm 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 [16]. In severe cases of SEP, or where a cocoon is encountered at laparotomy, it is necessary to resort to surgical treatment. The various techniques are recommended such as membrane excision + adhesiolysis, resection + anastomosis, resection + anastomosis + protective enterostomy 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 an intestinal resection [17].

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 [18]. We did not attempt any laparoscopic management in our center as our patents were young and sick children. All except 1 patient presented to the surgical emergency in total intestinal obstruction. 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.

• Other additional procedures like strictroplasty, simple closure of perforation and an enterostomy may be needed.

Conclusion

Tuberculosis as a cause of childhood AC is rather common in developing countries and is potentially a fatal condition. Diagnosing childhood TB is very challenging and current tools are inadequate. It is very difficult to make a definitive diagnosis of extra pulmonary TB in this group of vulnerable population. Radiologists should be knowledgeable about this entity which is best diagnosed on computed tomography. The final diagnosis of abdominal cocoon is usually made based on surgery and histopathology. Currently, laparoscopy is also used as a diagnostic tool in acute abdominal conditions with added advantage of tissue biopsy. The condition is unlikely to recur thereafter.

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