Primary encapsulating peritoneal sclerosis in a tuberculosis-endemic region

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Key words
cocoon, intestinal obstruction, peritonitis, tuberculosis.

Accepted for publication 23 January 2019.

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Declaration of conflict of interest: None.

Introduction
Encapsulating peritoneal sclerosis (EPS), also known as abdominal cocoon, is characterized by the encasement of variable length of intestine by a dense, fibrous, membrane-like sac, giving rise to a cocoon appearance and ultimately leading to intestinal obstruction. It is of two types—primary or idiopathic and secondary.1 The primary form is of unknown origin and has been classically described in young adolescent females from the tropical and subtropical countries. Secondary EPS has been reported in association with abdominal tuberculosis; beta-blockers (Practolol); chronic ambulatory peritoneal dialysis; liver cirrhosis; sarcoidosis; systemic lupus erythematosus; ventriculoperitoneal, peritoneovenous shunts; prior abdominal surgery; intraperitoneal chemotherapy; endometriosis; and gastrointestinal malignancy. EPS is a rare cause of intestinal obstruction in patients with abdominal tuberculosis.2 Therefore, in a region where tuberculosis is endemic, even primary EPS is frequently misdiagnosed as abdominal tuberculosis, and such patients are given antituberculous treatment (ATT) empirically. Here, we report a series of six patients who presented to our hospital over a period of 5 years (2007–2012) with acute intestinal obstruction because of EPS of primary origin and review the relevant medical literature.

Case series
The median age (range) of patients (n = 06) was 26 (13–62) years. There were 4 females and 2 males. On presentation, all patients had evidence of acute intestinal obstruction. All of them had a history of long-standing abdominal pain associated with early satiety, bloating, and recurrent episodes of vomiting. The median (range) duration of symptoms was 26 (7–62) months.

Discussion
Primary EPS presenting with recurrent intestinal obstruction is a rare occurrence. This condition typically presents in young
adolescent girls in tropical and subtropical countries, although many cases have also been reported from temperate zones, and it can occur in other age groups and genders, as found in our study. To the best of our knowledge, this is the first case series of this rare condition from the eastern part of India. The cause of EPS is still speculative. Although many cases are diagnosed incidentally at surgery, better awareness of this condition and multidetector CECT abdomen may aid in preoperative diagnosis. Based on the imaging findings of this series, we suggest that the diagnosis of idiopathic cocoon be strongly considered if the CECT exhibits clustered bowel loops, sac-like membranous structure encasing the small bowel loops, and no distinctive feature suggestive of tuberculosis, such as enlarged abdominal lymph nodes, significant ascites, or mesenteric thickening. Three types of EPS have been described in the literature depending on the extent of organ involvement: type I, if the membrane involves only a part of the small intestine; type II, if the entire small bowel is involved; and type III, if the colon or any visceral organs are also involved. Sometimes, EPS may be confused with internal hernia, which, like the abdominal cocoon, also depicts abnormal clustering of small bowel loops. Other conditions that may mimic EPS on imaging are pseudomyxomaperitonei, peritoneal mesothelioma, peritoneal carcinomatosis, sclerosing lymphoma, and mesenteric neoplasm.

Although multiple reports of EPS have been described in the literature, its occurrence creates a diagnostic dilemma and confusion, particularly with abdominal tuberculosis in a region where tuberculosis is endemic. Because the clinical features of EPS mimic tuberculosis, and radiological findings are subtle, this condition is frequently misdiagnosed as tuberculosis in endemic areas. Therefore, four of six patients of primary EPS in our study were given empirical ATT. The practice of empirical use of ATT is not always safe because of its propensity to cause hepatotoxicity and even death due to acute liver failure (ALF). The reported rates of ATT-induced hepatotoxicity are higher among the Indian population than the Western population. The mortality rate from ATT-induced ALF was 63% in a study from India in which 67% of the study cohort had received ATT empirically. In our study, none of the patients was found to have tuberculosis on histopathological examination of the membrane. Thus, an indiscriminate use of ATT seems unjustified in patients with EPS. On the other hand, because patients with tubercular EPS may respond to nonsurgical treatment, every attempt should be made to rule out tuberculosis in such patients. Whenever applicable, these patients should be evaluated with chest imaging, sputum for acid-fast bacilli, tuberculin skin test, erythrocyte sedimentation rate, and ascitic fluid testing including adenosine deaminase, etc. The presence of loculated ascites, matted bowel loops, mesentric thickening with nodules, and characteristic mesentric lymphadenopathy on computed tomography (CT) scan may suggest a diagnosis of abdominal tuberculosis. Following surgery, a pathological examination of membrane showing a dense fibrocollagenous tissue without any neoplastic or granulomatous change establishes the diagnosis of primary EPS.

Although many cases of tubercular EPS may respond to ATT only, surgical treatment is often required in cases of primary EPS. In our study, as all patients had presented with acute abdomen, we performed open laparotomy. A laparoscopic approach has also been suggested in the literature, but one has to be careful when establishing a pneumoperitoneum to avoid injury to the bowel loops. A simple surgical removal of the fibrotic capsule to release the encased bowel is required. The long-term prognosis after surgery is good, and a cocoon rarely recurs if completely removed.

All our patients who underwent surgical intervention and subsequent appropriate medical therapy remained asymptomatic during follow up. With an aim to reduce inflammation and peritoneal fibrosis, many pharmacological agents such as steroids,
Primary encapsulating peritoneal sclerosis in India

Table 1 Summary of demographic data and clinical findings

| Patient | Age (years) | Gender | BMI (kg/m²) | Presenting symptoms | Duration of symptoms (months) | History of empirical ATT | CT findings | Intraoperative findings | Type of Cocoon (I, II, III) | Follow-up recurrence |
|---------|-------------|--------|-------------|---------------------|-------------------------------|-------------------------|-------------|------------------------|-------------------------|----------------------|
| 1       | 62          | Male   | 19.7        | Recurrent bilious vomiting, abdominal fullness, weight loss | 7                             | Yes, for 3 months         | Membrane-like structure encasing bowel loops with minimally dilated bowel | Entire small bowel from terminal ileum to stomach encased in sac. Minimal free fluid, no stricture or tubercles | II                    | No recurrence          |
| 2       | 20          | Female | 16.5        | Recurrent pain abdomen with on and off vomiting, constipation | 62                            | Yes, 9 months             | Dilated small bowel loop with stricture in terminal ileum with mesentric lymphadenopathy | Around 30 cm of terminal ileum encased in sac with part of caecum. No stricture of bowel and free fluid | I                     | No recurrence          |
| 3       | 22          | Female | 17          | Dyspeptic symptoms, weight loss, vomiting                     | 40                            | Yes, 6 months             | Left paraduodenal hernia                                                   | Entire ileum and half of jejunum encased in sac without free fluid. Minimal lymphadenopathy present | II                    | No recurrence          |
| 4       | 13          | Female | 18.5        | Abdominal mass asymptotic, dyspeptic symptoms                 | 22                            | No                         | Dilated small bowel loops. Colon collapsed                               | Entire ileum up to duodenojejunal flexure involved in sac without free fluid and stricture | II                    | No recurrences         |
| 5       | 30          | Male   | 16.4        | Intestinal obstruction                                        | 20                            | No                         | Membrane-like structure encasing bowel loops with minimally dilated bowel | Entire small bowel up to and part of stomach involved in sac without free fluid and stricture | II                    | No recurrence          |
| 6       | 40          | Female | 17.3        | Recurrent vomiting with pain abdomen and abdominal mass       | 30                            | Yes, 3 months              | Minimal dilatation of small bowel loops with mesentric lymphadenopathy     | Entire small and large intestine with lesser sac and right lobe of liver involved within sac | III                   | No recurrence          |

ATT, antituberculosis treatment; BMI, body mass index; CT, computed tomography.

immunosuppressive agents, and tamoxifen have been used in patients with secondary EPS; however, there is no evidence for their use in patients with primary EPS.  

EPS should be suspected in patients who present with intestinal obstruction without a definite cause; with a history of recurrent episodes of obstruction; and presenting with a centrally located, ill-defined, nontender abdominal lump. Although EPS may be diagnosed incidentally at surgery, better awareness of this condition and the characteristic findings on CECT of abdomen may help in preoperative diagnosis in most cases. The CECT of abdomen is the most important modality that may help not only in diagnosing this condition but also in defining the extent of involvement and differentiating it from other conditions. The definite risk factors for primary EPS is not known; however, in the absence of risk factors for secondary EPS, such as tuberculosis, peritoneal dialysis, beta-blocker, intraperitoneal chemotherapy, previous abdominal surgery, endometriosis etc., histopathology of encasing membrane showing nonspecific fibrocollagenous tissues virtually establishes a diagnosis of primary EPS.

In conclusion, primary EPS is an enigmatic entity that may be erroneously diagnosed as abdominal tuberculosis in an endemic region of tuberculosis. In patients with a history of recurrent intestinal obstruction, a high index of suspicion along with characteristic images of a CT scan can help in diagnosis. It is potentially a curative condition by surgery. Unless there is strong supportive evidence preoperatively, an empirical ATT should be avoided till corroborative intraoperative findings or histopathological evidence of tuberculosis on membrane biopsy are obtained.

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