



CASE SERIES

Agensis of the greater omentum with primary abdominal cocoon: not an acquired condition

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Background and aims: Agensis of the greater omentum and the primary abdominal cocoon is rare. It is characterized by total or partial encasement of the small bowel inside a membranous bag. We report four cases of primary cocoon-causing intestinal obstruction. *Materials and methods:* Primary abdominal cocoons are the cause of intestinal obstruction. In this condition, the small intestine, in part or whole, is encased inside a glistening membranous sac. The first case we encountered in 1998 was at the Gauhati Medical College and Hospital (GMCH), Guwahati, Assam. It was presented at a local conference and in the Journal of the Association of the Surgeons of Assam. Long 21 years passed before we faced another case of agensis of the greater omentum with primary abdominal cocoon causing acute dynamic small bowel obstruction in the Jorhat Medical College and Hospital (JMCH) in Jorhat, Assam. Later in quick succession, we had two more cases; one was diagnosed preoperatively by the Contrast Enhanced Computerized Tomography (CECT) abdomen. All these cases were explored for unresolved obstruction, and on the table, findings suggested the primary abdominal cocoon. The findings were compared with the first case encountered in 1998. The other cases and the peculiarities were noted, especially the agensis of the greater omentum and the presence of free peritoneal bodies. Photographs were taken. *Results:* The patients were operated upon as emergency exploratory laparotomy, and the Sac was excised. All recovered well. The one patient diagnosed by the CECT awaits intervention as she improved on conservative therapy. *Conclusion:* The literature reports a primary abdominal cocoon with agensis of the greater omentum. We suggest the condition is not acquired as the agensis of the greater omentum association in all cases proves that this syndrome is congenital. The presence of the Sac without any cellular component is an enigma.

Keywords: Intestinal obstruction; agensis; hypoplasia; greater omentum, primary cocoon.

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INTRODUCTION

Agensis of the greater omentum along with primary abdominal cocoon is a rare condition that refers to the absence of the greater omentum from the transverse colon

and along with total or partial containment of the small bowel inside a fibro-collagenous membrane bag leading to acute or chronic bowel obstruction. The cocoon is described by various terms like 'peritonitis chronica fibrosa

incapsulata' and sclerosing encapsulating peritonitis.¹ We hereby present four cases with features of acute intestinal obstruction, were found to have, and three were treated successfully by surgery. These cases had agensis of the greater omentum. Abdominal cocoon syndrome was first defined in 1978 and is a relatively rare condition, limited to a few case reports. In this syndrome, a portion or all of the small intestine is encased inside a sac lined by a fibro collagenous membrane without the mesothelium.^{2,3} It is a rare entity whose clinical findings are non-specific, so it is mainly diagnosed during surgery. However, contrast-enhanced abdominal computerized tomography (CECT) can be used to diagnose these days during the preoperative period.⁴ Treatment is the removal of the membranous Sac and release of adhesion.^{6,7}

We present a series of four patients with agensis of the greater omentum with abdominal cocoon; three were successfully and surgically treated.

MATERIALS AND METHODS

We present our findings based on the four cases encountered over 20 years. The first one, a female in her teens, was admitted with acute dynamic intestinal obstruction. A plain X-Ray of the abdomen showed multiple gas fluid levels in the pattern of small bowel obstruction. It did not resolve on active conserve treatment. Emergency exploratory laparotomy revealed a thick glistening membrane resembling a bag of peritoneum inside the peritoneal cavity. The bag contained the whole small intestine inside. The terminal ileum was obstructed by the edge of the bag, which acted like a sharp band. The large gut was empty, and no obstruction was found. The greater omentum was non-existent. It was hypoplastic to the amount of almost agensis. The sac was surgically excised totally. No resection and anastomosis were needed. Recovery was smooth and uneventful. No photographs could be taken during the midnight emergency surgery, but operation records were kept.

The second patient, a 40 years old healthy young adult male, who presented with acute dynamic intestinal obstruction of a few days duration, was taken up for emergency exploratory laparotomy. The patient recovered from being discharged usually. Here also, no resection of

the bowel was necessary. Ultrasonography and CT scan of the whole abdomen failed to detect the cocoon before surgery.

The following case was a middle-aged female with recurrent intestinal obstruction of various degrees. CECT Scan diagnosed her, and the finding corroborated the presence of a cocoon and not detectable greater omentum.

The fourth case was a middle-aged male with the same complaints, and he also had the same shiny bag-like structure and agensis of the greater omentum. He was operated upon and had a turbulent post-operative recovery but was later discharged healthy.

Biopsy reports from the membranous sac in all resected specimens showed a fibrous structure in a collagenous background and no evidence of any epithelium or mesothelium. The peritoneal mice and the membrane showed Fibro-collagenous tissue with occasional acute inflammatory cells without evidence of any mesothelium. The pathologist's opinion in all resected cases was that of sclerosing encapsulating peritonitis.

Ethical committee clearance was obtained. Informed consent was also obtained before the data collection.

RESULTS

The presentation was an intestinal obstruction in all the patients. Out of four, three cases were diagnosed with exploratory emergency laparotomy. In these cases, it was found that the entire small bowel was encapsulated in a dense, glistening, smooth, white, fibrous cocoon, which looked very much like a bag of peritoneum inside the peritoneal cavity (Fig.1). The greater omentum was found to be almost non-existent all cases (Fig. 2, 3, 4). Present-day CECT abdomen can detect the condition if the radiologist is aware of the condition. In our experience, the discussion with the radiology faculty in the second case probably led to the detection of the cocoon in the third case by the radiology in the preoperative CECT abdomen. Excision of the membrane and release of adhesions were carefully performed to free the small intestine. Post-operative recovery in these operated cases was smooth and uneventful. The sac was excised in toto in all cases operated upon (Fig. 5).



Figure 1 The sac showing the glistening wall of the cocoon

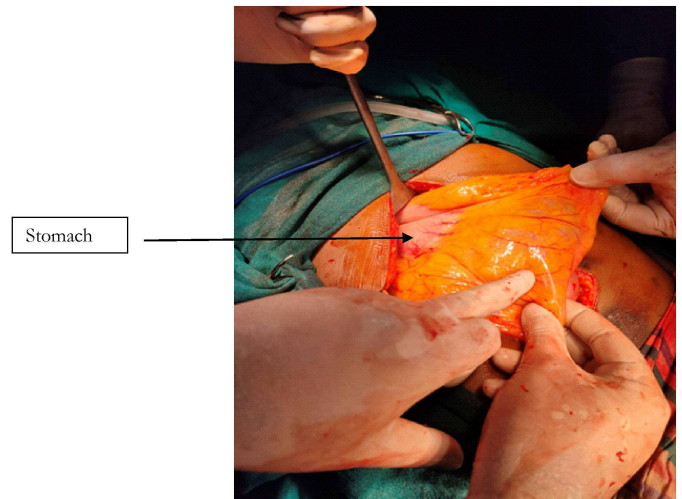


Figure 4 Agnesis of the gastrocolic omentum beyond the transverse colon

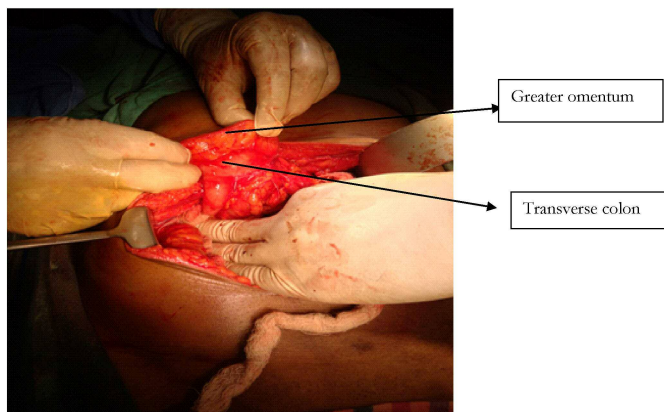


Figure 2 Almost non-existent greater omentum

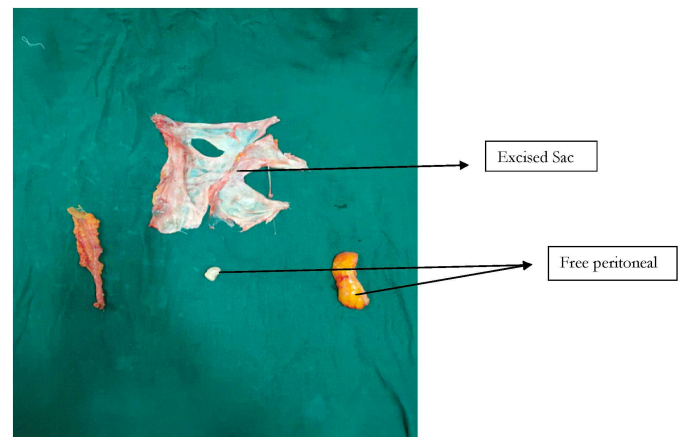


Figure 5 Case two-excised sac and removed free peritoneal bodies (Peritoneal Mouse/ Mice)

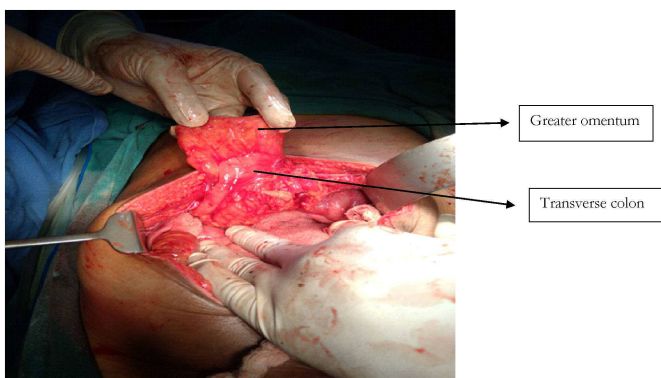


Figure 3 The second case (hypoplastic greater omentum)

DISCUSSION

The agnesis of the greater omentum to the level of severe hypoplasia beyond the free border of the transverse colon could not be explained by the available embryological concepts. Abdominal cocoon, although it is classified as primary and secondary based on whether it is idiopathic or has a definite cause. The origin of the primary form is uncertain, with various hypotheses. It is probably caused by subclinical peritonitis leading to the formation of a cocoon.^{1,4,5} Patients with abdominal cocoons present with features of recurrent acute small bowel obstruction secondary to kinking and compression of the intestines within the constricting cocoon.^{2,3} Foo et al. detected the condition in 10 young girls with symptoms of intestinal obstruction two years post menarche.^{4,6} It was

suggested that chemical peritonitis might be caused by retrograde Menstruation leading to the formation of a cocoon.⁴ Secondary causes include the placement of Le Vein shunts for refractory Ascites, continuous ambulatory peritoneal dialysis, erythematosus, and the use of povidone-iodine for abdominal wash-out. The α adrenergic blocker Practolol is noted to have caused a cocoon. Practolol has hence been withdrawn from use because noted to cause the formation of a peritoneal membrane just like the cocoon.

Presentation as an abdominal mass or lump may also be found due to an encapsulated cluster of dilated small bowel loops.

Abdominal X-ray findings are non-specific and suggest fluid air level in the gut suggesting intestinal obstruction of variable degrees. CECT of the abdomen is now considered valid for the preoperative diagnosis of the abdominal cocoon. The imaging features are, however, not confirmatory. CT findings of a membrane-like structure enveloping small bowel loops were seen in some para-duodenal hernias, an abdominal cocoon, and peritoneal encapsulation.⁵ The clinical and pathological features of these entities are, of course, different.

The final diagnosis of the abdominal cocoon is usually based on intra-operative and histopathology findings. A significant number present for emergency treatment without any imaging being performed. In all the reported patients, portions of the small bowel were encased within a fibrous cocoon.⁷⁻¹⁰

Differential diagnosis includes a condition called "peritoneal encapsulation", a developmental anomaly where the small bowel is encased in a thin accessory peritoneum-like membrane. In peritoneal encapsulations, the patients are mostly asymptomatic, the findings are incidental, and the patients present late in life. Treatment, as in the present case, consists of excision of the accessory peritoneal Sac with adhesiolysis of the inter-loop adhesions. Bowel resection is not necessary unless a gangrenous segment is found.

In our cases, the hypoplasia or agensis of the greater omentum was the striking feature, along with the free peritoneal bodies. Other authors have also mentioned this additional finding.⁷ The absence of the greater omentum conclusively proves that the condition is not an acquired condition, and its origin must be some congenital accidents not yet identified.

CONCLUSION

The findings of agensis of the greater omentum associated with a primary abdominal cocoon are a mystery. We don't know the reasons for this association yet, and it can't be explained with the presently available data. Primary Abdominal cocoon otherwise is a specific entity and is always present with intestinal obstruction of various stages. Preoperative diagnosis is difficult because of the rarity of the condition. The agensis of the greater omentum remains a mystery. The cocoon treatment is surgery, and the recovery of our patients was also satisfactory. As primary abdominal cocoon is a rare condition, diagnosis requires a high index of suspicion.

From our experience, it can be suggested that the agensis of the greater omentum is a congenital anomaly, not an acquired condition. Hence this condition of primary abdominal cocoon associated with agensis of the greater omentum be considered a congenital anomaly rather than an acquired condition.

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