

**Multimodality Review of Sclerosing Encapsulating Peritonitis – A Case Series**

**Gehlot Kunal<sup>1</sup>, Choudhary Mayank<sup>2</sup>, Gehlot Kushal<sup>3</sup>, Kardam Narendra<sup>4</sup>, Ola Sandeep<sup>5</sup>, Gehani Malvika<sup>6</sup>, Jain Amit<sup>7</sup>, Garasiya Kantilal<sup>8</sup>**

<sup>1</sup>Junior Resident, Department of Radiodiagnosis & Modern Imaging, Rabindranath Tagore Medical College, Udaipur (RAJ.)

<sup>2</sup>Assistant Professor, Department of Radiodiagnosis & Modern Imaging, Rabindranath Tagore Medical College, Udaipur (RAJ.)

<sup>3</sup>Professor, Department of Radiodiagnosis & Modern Imaging, Rabindranath Tagore Medical College, Udaipur (RAJ.)

<sup>4</sup>Professor, Department of Radiodiagnosis & Modern Imaging, Rabindranath Tagore Medical College, Udaipur (RAJ.)

<sup>5</sup>Assistant Professor, Department of Radiodiagnosis & Modern Imaging, Rabindranath Tagore Medical College, Udaipur (RAJ.)

<sup>6</sup>Junior Resident, Department of Radiodiagnosis & Modern Imaging, Rabindranath Tagore Medical College, Udaipur (RAJ.)

<sup>7</sup>Junior Resident, Department of Radiodiagnosis & Modern Imaging, Rabindranath Tagore Medical College, Udaipur (RAJ.)

<sup>8</sup>Junior Resident, Department of Radiodiagnosis & Modern Imaging, Rabindranath Tagore Medical College, Udaipur (RAJ.)

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Corresponding Author: Sandeep Ola

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**Abstract:**

**Background:** Abdominal cocoon is a rare peritoneal disease. The aim of this study is to investigate the imaging features of abdominal cocoon (AC), the methods of diagnosis and treatment of AC and improve understanding of AC.

**Methods:** We have retrospectively analyzed 4 patients with AC. The study included a total of 4 patients with 3 female and 1 male with mean age of 45 years (Range-37 to 52 years). All patients were followed up (at 1, 6 and 12 months after surgery) by telephone or using the outpatient service. Data included clinical manifestations, imaging examinations, diagnoses and treatments of the patients.

**Results:** The incidence of acute abdominal pain, abdominal distension, unexplained chronic mechanical intestinal obstruction and soft abdominal mass was 100%. Two out of four patients presented with dilated small-bowel loops with air fluid levels in the small intestine. A total of two cases out of 4 presented with strangulated intestinal obstruction. The abdominal X ray examination identified that two patients (50%) showed proximal intestinal dilatation, normal or poor intestinal dilation and the distal bowel was normal or empty. The sac's position, size and the scope of involved bowel were consistent with CT findings.

**Conclusion:** Enhanced CT is an effective way to observe bowel mesenteric ischemia and necrosis and has high sensitivity and accuracy. However, combined with clinical and imaging reports, the current study performed preoperative diagnosis using CT scans, which allow for a reliable diagnostic method.

**Keywords:** Abdominal Cocoon, Imaging Features, Diagnosis Methods, Treatment Approaches, Mesenteric Ischemia, Enhanced CT Diagnostic Accuracy.

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**Introduction**

Sclerosing encapsulating peritonitis often known as abdominal cocoon is a rare entity in settings of chronic inflammatory disorders like tuberculosis. In a country like India which have cases of tuberculosis in bulk it is often a common condition. The presenting symptoms are often of intestinal obstruction pertaining to encasement of variable lengths of

bowel loop with fibro-collagenous membrane which resembles a cocoon. This condition is often missed preoperatively however with recent advancements of Computed Tomography and various other modalities it is now possible to diagnose such entities preoperatively before laparotomy. [1]

The study aims to investigate to investigate the imaging features of abdominal cocoon (AC), the methods of diagnosis and treatment of AC and improve understanding of AC. To do this, we have retrospectively analyzed three patients with abdominal cocoon. The clinical manifestations included bowel obstruction and abdominal distension. And the features included- 1) Bowel loops present in a fixed cluster; 2) bowel loops encapsulated partially or totally by a thickening fibrous membrane-like 'cocoon'; 3) bowel wall thickened in the 'cocoon', and the bowel loops and sac adhered; 4) loculated ascites in the 'cocoon'; 5) abnormal clustering of the mesenteric vasculature, hypoplasia of the omentum majus was present or absent. The features of a plain abdominal X-ray, a gastrointestinal barium meal series and ultrasonography lacked specificity. The results of CT in cases were consistent with the surgical findings. Surgery was the first choice of therapy. All cases were treated surgically, showing that the bowel loops were encapsulated partially or totally by a thickened fibrous membrane. All cases received operations including partial or total excision of the membrane and enterolysis. In conclusion, CT may be highly valuable in the preoperative diagnosis of AC. [2]

#### Materials & Methods

All patients were taken from Maharana Bhupal Hospital between March 2022 to December 2022. Data included clinical manifestations, imaging examinations, diagnoses and treatments of the patients. Patients' inclusion criteria were as follows: i) CT scan and enhanced scan completed prior to surgery; ii) Patients had no previous history of abdominal surgeries, peritonitis or peritoneal dialysis, autoimmune diseases or prolonged drug intake. Patients' exclusion criteria were: Patients who received CT scan without an enhanced CT. All patients were followed up (at 1, 6 and 12 months after surgery) by telephone or using the outpatient service.

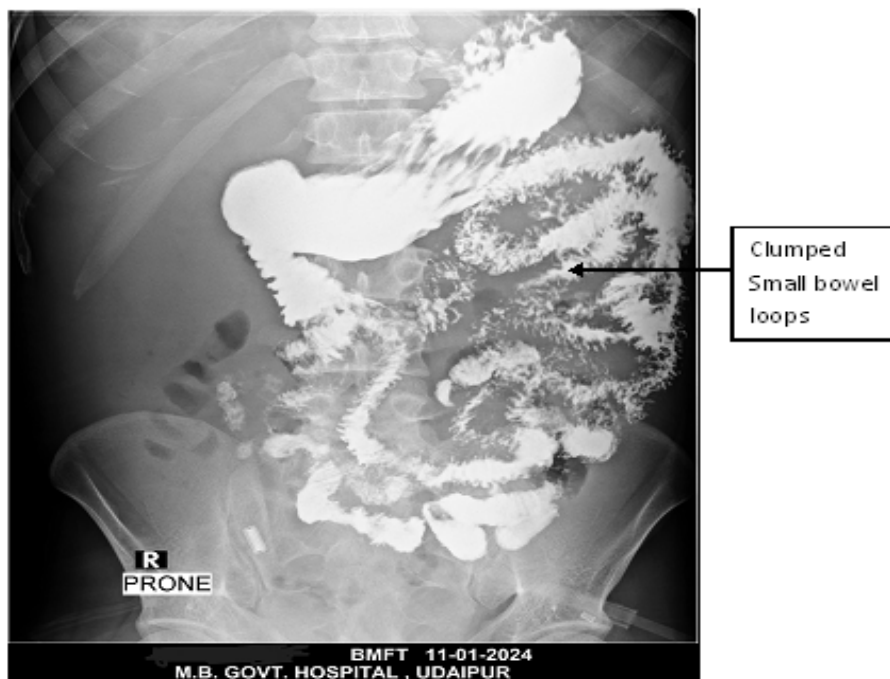
Radiological imaging included X-ray and sonographic examination. Plain abdominal X-ray and ultrasonography were performed before surgery as routine examinations. CT scans and subsequent contrast-enhanced CT scans were performed with a 64-Multidetector row helical CT scanner in all cases. For the patients with or without partial intestinal obstruction, 4% mannitol was orally administered 1 h before the examination to fill the gastrointestinal tract. A laparotomy was performed within 48 h after helical CT in all four cases. All samples were histo-pathologically examined. The pathological examination was performed under an optical microscope with a magnification of (x100). Fixation was performed using 10% formalin at room temperature for 6-8 h. Staining was performed using haematoxylin-eosin at 40°C for ~30 min. The thickness of the tissue sections was 4 µm.

#### Results

The study included a total of 4 patients with 3 female and 1 male with mean age of 45 years (Range-37 to 52 years). All patients had no previous history of abdominal surgeries, peritonitis or peritoneal dialysis, autoimmune diseases or prolonged drug intake. All patients showed recurrent abdominal pain and distention, which was more apparent after meals, all cases had episodes of intestinal obstruction, abdominal distention, colicky abdominal pain, nausea and vomiting and had a soft, smooth mass upon abdominal palpation and the boundary of the mass was not sharp.

#### X- Ray Examination

The plain abdominal X-ray examination identified that two out of four cases presented with dilated small-bowel loops with air-fluid levels in the small intestine. Following the barium meal examination revealed that one of the four patients had a cauliflower sign, which consisted of disorderly arranged and bunched bowels that congregated in a single area. When pressing the clustered bowel loops, they remained in a constant position. (Fig. 1)

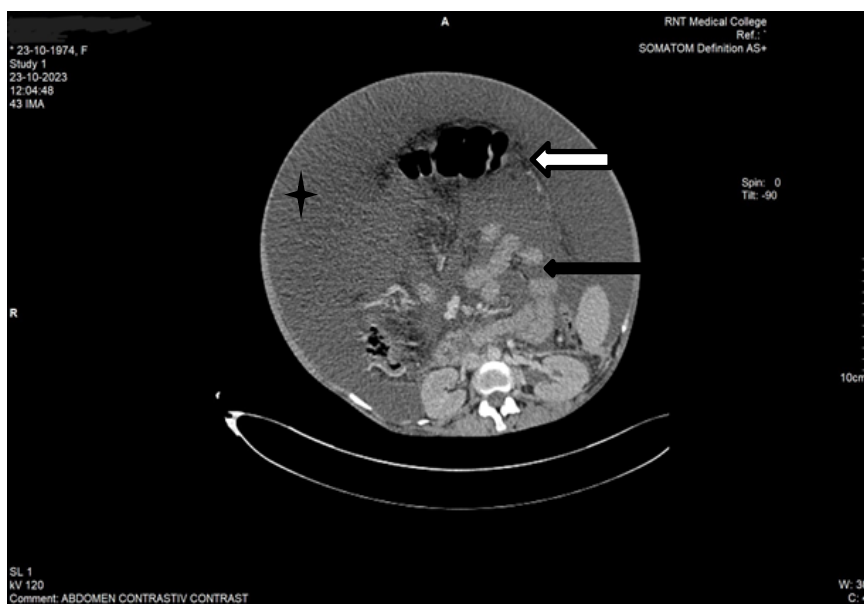


**Figure 1 X- ray AP (prone) view shows clumping of small bowel loops in left half of abdominal cavity.**

**CT interpretation:-**

An appropriate amount (1,000-1,500 ml) of oral 4% mannitol liquid was used as the contrast medium that filled the gastrointestinal tract prior to the CT scan, and helped to identify clustered intestinal loops encased by a membrane-like sac. In three cases, the small-bowel loops congregated in the middle-lower abdomen, and were encased by a soft-tissue density mantle. A small amount of fluid was identified between the sac and these

encapsulated bowel loops. A thick. (2 mm) mild-moderate enhancing membrane surrounded the bowel, (Fig. 2) and the wall of the enhancing membrane was well-distributed in three cases. A total of two cases out of four presented with strangulated intestinal obstruction. [3] The CT findings included a thickened intestine wall, which either could not be enhanced or could be enhanced slightly, and mesenteric hydrops when the mesenteric fat gained density. Ascitic fluid was identified in three of four patients.



**Figure 2: Shows Contrast enhanced axial CT scan of abdomen in which white arrow shows thick enhancing membrane surrounding bowel loops, black arrow shows clumped small bowel loops & + denotes gross ascites in abdominal cavity**



**Figure 3:** Shows sagittal contrast enhanced CT images in which the black arrow denotes clumped small bowel loops and the white arrow demarcates the thickened enhancing membrane of sclerosing encapsulating peritonitis.



**Figure 4:** Shows axial contrast enhanced CT scan of the abdomen in which the asterisk shows gross ascites and black arrow demarcates the clumped small bowel loops.

**Surgical Interpretation**

On opening the abdomen, all or part of the small bowel and the colon were revealed to be enclosed by a whitish cocoon-like sac. The small bowel was shortened and the intestine was either lacking or missing entirely, or in the abdominal cavity. During surgery it was observed that the sac's position, size and the scope of involved intestine were consistent

with CT findings. Part of the tough well-distributed membrane-like tissues adhered to the wall of abdomen, which was approx. 2-mm thick and involved encased intestine loops. The tissues congregated to the centre of the abdomen with the intestine loops adhering to each other and to the sac. A small volume of flaxen or red liquid was observed in the sac, and oedematous mesenteric

and bloody dialysis was effluent in the abdominal cavity. Homologous mesenteric vessels with preternatural distribution, were thickened and centralized. The greater omentum appeared hypoplastic or absent.

### Pathological Examination

The microbiological examination of the opalescent membrane showed proliferation of fibroconnective and adipose tissue with a chronic inflammatory reaction accompanied by degeneration or necrosis and formation of granulation tissue. The dark purple wall of the necrotic bowel appeared thickened and swelled with the haemorrhagic contents leaking into the enteric cavity. Microscopically, the bowel wall was observed to bleed, necrose and exhibit the inflammatory reaction.

### Discussion

The observation that abdominal organs were partially or totally encased in a fibrous membrane and consisted of multiple internal adhesions was first reported by Owtschinnikow [4] in 1907, in a study entitled 'Peritonitis chronic fibrosa incapsulata'. However, the condition has also been described as peritoneal fibrosis, peritoneal sclerosis, calcified peritonitis and encapsulated peritoneal sclerosis. The abdominal cocoon was first named in 1978 by Foo et al. [5] The fibrous membrane surrounds the small bowel, but it occasionally extends to include the colon, stomach or other organs. However, it rarely encases all abdominal organs. [6]

Abdominal cocoon is a rare peritoneal disease, the pathogenesis of which remains to be determined. AC can be divided into an idiopathic and a secondary form. [7] The idiopathic form has been reported to be association with congenital dysplasia. [8] Causes of the secondary form include chronic peritoneal dialysis, intra-abdomen foreign body stimulation, serious and chronic peritonitis, autoimmune disease, intake of the  $\beta$ -blocker practolol, bacteria proof filter use, liver transplantation, tuberculous inflammation, ventriculoperitoneal and peritoneovenous shunts and carcinoid syndrome. [7,9] All these factors can lead to peritonitis, leading to a decrease in mesothelial cells, a sustained expression of mesothelial metastatic growth factor and the production of a large number of extracellular matrix products, which increase the exudation of protein fibre and peritoneal fibroblasts hyperplasia, [10] and lead to the formation of the fibrous sac. Peritoneal dialysis-related AC may be due to the dialysis solution and its metabolites damaging the peritoneum, which can lead to the subcutaneous area of the peritoneal mesothelium thickening and expanding. Repeated infections can cause peritoneal damage, which can impair the normal

physiological function of the peritoneum, and undergoes three stages of turbidity, deformation and fibrosis. [1] In the current study, the four included cases did not exhibit peritonitis, peritoneal dialysis or prolonged drug intake. The patients had a wide range of age distribution and majority were male.

The clinical presentation of Abdominal Cocoon syndrome mostly occurs as acute abdominal pain. The incidence of acute abdominal pain in the current study was 100%. The main clinical manifestations included signs of bowel function and peritoneum physiological function disorder, and the fibrous sac also led to bowel function disorder, decreased reabsorption and weakened enterokinesia, which caused nausea, vomiting, abdominal distension, the disappearance of bowel tone, abdominal pain, abdomen or pelvic masses, intestine obstruction, weight loss and blunt abdominal wall trauma-induced intra-cocoon bleeding. Peritoneal fibrosis led to ascites by blocking the lymphatic vessels, which is a nonspecific symptom. The most common manifestation of the disease is small bowel obstruction, which is characterized by complete or partial obstruction and this was observed in approximately two-thirds of the cases in the current study. Signs of bowel function disorder were not only related to the encapsulated intestine but also the damaged vessels, vasa lymphatica and nerve plexus of the bowel wall muscular layer. [11] Abdominal Cocoon may lead to infertility in female patients as the fibrous membrane encapsulates the fallopian tubes, which restricts its movement, blocks the fimbriated extremities and makes it difficult for the ovum to travel the fallopian tubes, however the uterus and ovaries appeared to be normal.

Clinical diagnosis of Abdominal Cocoon is undertaken based on signs of bowel function and peritoneum physiological function disorder. The signs of Cocoon are nonspecific, so it is difficult to make a definite preoperative diagnosis. The majority of cases are diagnosed during a laparotomy. Patients presenting with recurrent episodes of abdominal pain, abdominal distension, unexplained chronic mechanical intestinal obstruction and soft abdominal mass. Weakened peritoneal transport function, anaemia and C-reactive protein levels can act as a clinical reference, but have no specific value. An elevated WBC count and C-reactive protein level, hypoalbuminemia and anaemia may be detected in patients.

Plain abdominal X-ray has been demonstrated to exhibit no diagnostic specificity, and only indicated dilated small-bowel loops with air-fluid levels and peritoneal calcification in previous studies.

In the current study, a total of two cases (50%) exhibited small intestinal air-fluid levels and there were no cases of peritoneal calcification. Barium meal examination revealed the cauliflower sign in one case (25%).

CT scans can indicate a distinctive manifestation of AC, which is of important value for diagnosis. With CT findings, a definite preoperative diagnosis is more likely. These manifestations include: 1) The small-bowel loops amassing in a certain area of the abdomen and the intestine is rarely seen in other areas of the peritoneal cavity; 2) the clustered bowel loops are surrounded by a sac-like structure in a typical cocoon pattern; 3) the wall of the sac is complete or incomplete, and well- or poorly distributed. 4) Intestine loops in the sac may adhere to each other and the wall causing it to thicken; 5) small amounts of encapsulated effusion in the sac are visible; 6) corresponding mesenteric vessels are centralized with abnormal running and distribution, mesenteric fat gains density and the greater omentum is hypoplastic or absent; 7) cocoon-like membrane and the wall of the intestine occasionally show calcification; 8) lymph nodes swell reactively; 9) CT findings of cocoon with the complication of intestinal obstruction; 10) intestine loops encased by the sac and secondary adhesion being the main cause of the obstruction. In the current study, a total of two patients (50%) showed proximal intestinal dilatation, normal or poor intestinal dilatation and the distal intestine was normal or empty. Bowel ischemia is a manifestation of a strangulated intestinal obstruction. And the Bottle Gourd sign, cauliflower sign are important radiological findings and they were identified in one of cases. Enhanced CT is an effective way to observe bowel mesenteric ischemia and necrosis, and has high sensitivity and accuracy. CT findings of bowel ischemia include: 1) Bowel wall annular thickening; 2) abnormal enhancement of the bowel wall; 3) bowel wall bleeding; and 4) mesenteric effusion, mesenteric vessels thickening and fuzziness. In the current study, characteristic CT appearances of Cocoon include clustered bowel loops encased by a thickened sac that are accompanied by the accumulation of mesenteric vessels, abnormal distribution and the hypoplasia or absent omentum majus. Stafford-Johnson et al indicated that calcification of the intestinal frenum and peritoneum were more characteristic of Cocoon, but none of the cases in the current study showed peritoneum calcification. In the past, Abdominal Cocoon could only be definitively diagnosed after surgery. However, combined with clinical and

imaging reports, the current study performed preoperative diagnosis using CT scans, which allow for a reliable diagnostic method. Using this method increases the understanding of the disease and serves an important role in assisting surgical treatments.

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