Sclerosing encapsulating peritonitis in a young male secondary to Adenocarcinoma of the gastrointestinal tract

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ABSTRACT

Sclerosing encapsulating peritonitis or abdominal cocoon is a rare cause of small bowel obstruction. In the tropical countries, infectious etiologies like tuberculosis predominate, whereas in the Western world, gastrointestinal tract malignancies are more common and that too, in the elderly population. Here we present a case of a young male from the tropics, who presented to us with an abdominal cocoon, in whom an initial consideration of intestinal tuberculosis led to anti tubercular therapy which proved futile. A diagnosis of adenocarcinoma was eventually made on histopathology of omental tissue.

Keywords: Adenocarcinoma, Abdominal cocoon, Small intestinal obstruction, Tuberculosis

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INTRODUCTION

Sclerosing encapsulating peritonitis (SEP) or abdominal cocoon is a very rare cause of small intestinal obstruction. It is considered to be idiopathic or secondary to many other causes. The secondary causes most commonly in developing countries is due to tuberculosis, continuous ambulatory peritoneal dialysis, recurrent peritonitis, post liver transplantation, beta blocker usage, peritoneal shunts and the rarest being related to malignancies.(1) Malignancies that lead to SEP include mostly mucinous tumors of the ovary, thecomas and gastric adenocarcinomas. The disease mostly comprise of a thick fibrotic rind of peritoneal encasement across the visceral and parietal peritoneum that decreases the small bowel volume leading to features of obstruction. This condition is mostly described in young adolescent girls living in tropical regions, but can be seen rarely in males in non tropical regions also. (2)

This condition is different from pseudomyxoma peritonei in which copious

amounts of mucinous ascites is seen covering the peritoneal cavity leading to massive abdominal distension and features of obstruction. This presentation can be secondary to two types; the first in which a low grade mucinous tumor produces characteristic peritoneal involvement without underlying sub peritoneal invasion and the second in which a high grade poorly differentiated tumor produces characteristic pattern of disease with underlying subperitoneal invasion has been documented. [(3)] Patients of end stage liver disease in whom refractory ascites develops with recurrent spontaneous bacterial peritonitis intervenes also are predisposed to this condition. [(4)] The disease seldom affects males, that too, young males from the tropics. The disease presentation secondary to gastrointestinal tract malignancy is even unreported in young males. Adenocarcinomatous cause of SEP has not been reported in a young male from the tropics previously.

CASE REPORT

We report the case of a 29 year old male who presented to us with painful progressive abdominal distension for a period of one month associated with low grade fever. This was associated with severe anorexia and loss of appetite and a significant loss of weight of over 14 kilograms in the preceding month. The patient was a teetotaler with no history of past surgeries, dialysis procedures or chronic drug intake. He has never had exposure to tuberculosis in the past. There was no history of bilateral leg swelling, chest pain or palpitations, orthopnea or paroxysmal nocturnal dyspnea. The patient also denied long standing exposure to chemical and toxins. On examination, the patient was found to be conscious and alert. His blood pressure in the right brachial region in the supine position was 122/68 mm of Hg and the heart rate was 88 per minute. There was pallor, no icterus, cyanosis, clubbing or lymphadenopathy. The abdomen was distended, tender to palpation and tympanitic, with the flanks being dull. There was no appreciable organomegaly and the bowel sounds were hyperactive in the lower left quadrant and the infra umbilical region. The cardiovascular, pulmonological, central nervous system and reticuloendothelial and musculoskeletal system examination was essentially normal.

The investigation revealed the presence of anemia with hemoglobin of 10.8 g/dL

(normal), a total count of 11200 cells per cubic mm (normal), platelet count of 6 lacs per cubic mm (normal) and a total bilirubin of 1.0 mg/dl (normal) and a direct fraction of 0.2 (normal) mg/dl. There was no transaminitis and the serum albumin level was 3.8 g/dl (normal). Ultrasonography of the abdomen revealed the presence of moderate ascites and thickening of bowel loops and omentum. A subsequent ascitic fluid study revealed the presence of low SAAG, high protein ascites with a total cell count of 650, with lymphocytic predominance and an adenosine deaminase level of 8.3 U/L (normal < 33). An abdominal X ray revealed the presence of multiple air fluid levels and a subsequent contrast enhanced computed tomography (CT) of the abdomen revealed the presence of small bowel loops, from the duodeno-jejunal flexure till the ileocecal junction which were closely placed, abutting each other longitudinally in an 'accordion' like fashion (Figure 1). These loops had edematous walls and were surrounded by thick enhancing membrane consistent with cocoon formation (Figure 1). Few of the bowel loops were prominent and showing the presence of particulate (colon-like) feculent matter mingled with gas bubbles in the lumen of dilated loops of the small intestine, the small bowel 'feces' sign (Figure 1).

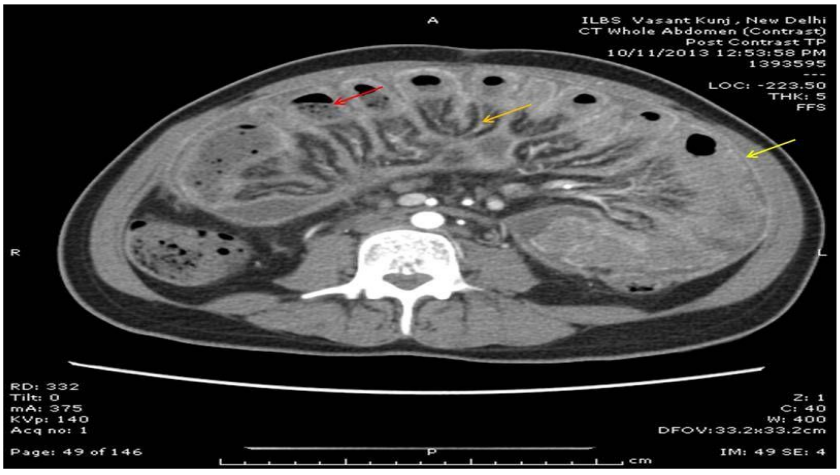

Figure 1: Contrast enhanced computed tomography of the abdomen showing the presence of enhanced peritoneal thickening forming a cocoon (yellow arrow); the 'small bowel feces sign' (red arrow) and the 'accordion sign' (orange arrow).

The peritoneum and omentum were enhancing with presence of nodular haziness suggestive of diffuse ometal involvement. Enhancing abdominal lymph nodes were seen associated with these findings. The liver was also enlarged in side with mild serrated outlines and heterogenous attenuation with areas of infiltration associated with hypertrophy of the left lobe (Figure 2).

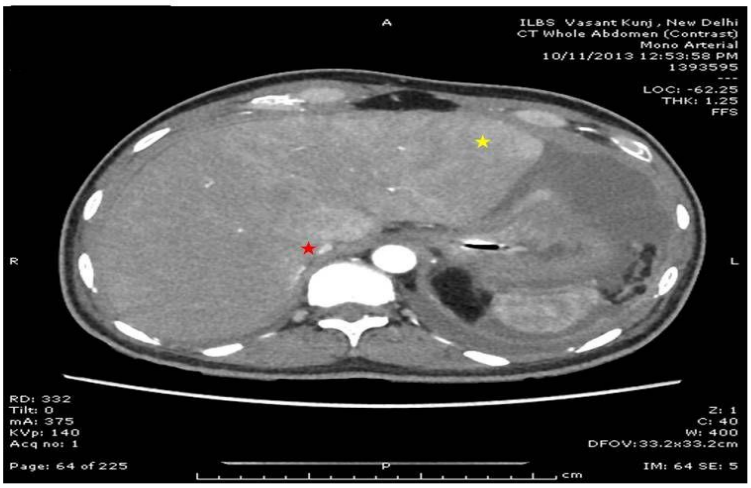

Figure 2 : Contrast enhanced computed tomography of the abdomen showing the presence of heterogenous enhancement of the liver (yellow star) secondary to passive congestion of the inferior vena cava by the cocoon (red star) .

The intrahepatic and infra hepatic inferior venacava upto the level of the left renal vein was collapsed. Collaterals were are seen at G-E juncton, periportal, perigastric, gastroepiploic, pericholedochal, splenic hilar and mesenteric regions and the large bowel loops were relatively spared with stomach showing reactive edematous wall thickening. Considering the possibility of extensive abdominal tuberculosis, the patient was started on four drug anti tubercular therapy. His condition worsened during the course in hospital as the small bowel obstructive features recurred with prolonged courses towards the later part of the hospital stay. At this juncture, the tumor markers, carcinoembryonic antigen, CA19-9, alfa feto protein and tumoral beta- HCG all came back negative. A surgical consultation was opined and the patient was taken up for exploratory laparatomy. During the exploratory laparatomy, it was noticed that the liver was

enlarged and congested with the omentum being thickened and hard. Dense adhesions were noticed between the stomach, transverse colon and small intestine – features suggestive of a frozen abdomen. The stomach, transverse colon and the small intestine were hard to palpation. Frozen section analysis of the omental biopsy was taken and it was suggestive of infiltration by malignant epithelial cells arranged in acini into the surrounding stroma. These malignant cells showed large pleomorphic and hyperchromatic nuclei with high N:C ratio. At places, signet ring cells were also seen, which was suggestive of positive for metastatic adenocarcinoma. The subsequent histopathological examination of the peritoneal biopsy revealed the presence of signet ring type of metastatic adenocarcinoma (Figure 3). Further surgical intervention was withheld and the abdomen closed. The patient was then managed conservatively with palliative symptomatic therapies. He eventually left against medical advice and was lost to follow up.

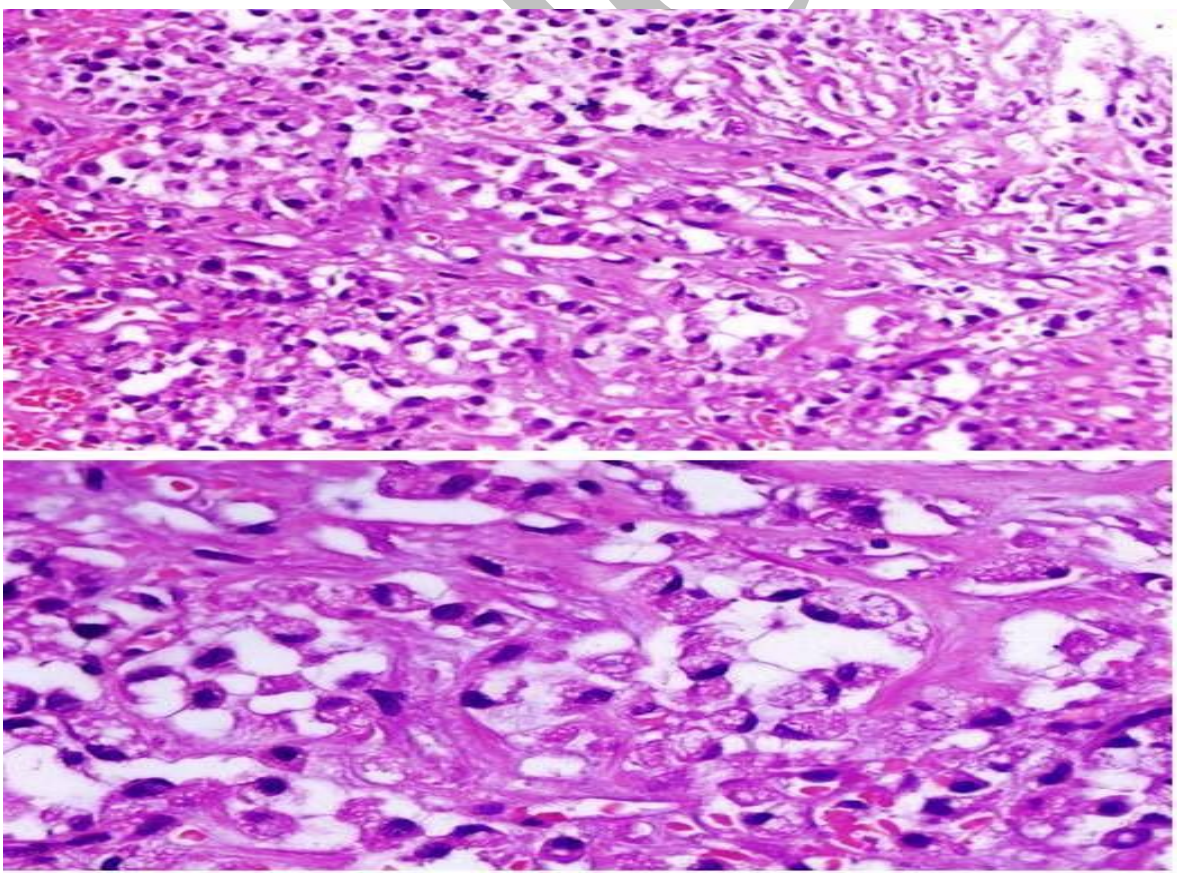

Figure 3: Frozen section from the omentum (above, Light Microscopy, H&E Stain, 100X) showing malignant epithelial cells arranged in acini formation with pleomorphic and hyperchromatic nuclei. The histopathological examination of peritoneal biopsy (below, Light Microscopy, H&E stain, 400X) showing few signet ring cells interspersed between malignant epithelial cells.

DISCUSSION

SEP is a rare and devastating condition that can be idiopathic or secondary to a variety of causes. It has been classically described in adolescent women hailing from tropical regions. In countries like ours, abdominal tuberculosis is one of the commonest causes for cocoon formation. [5] The other causes include ovarian malignancies in women.

The occurrence of abdominal cocoon is a rare finding in young men and that too in the tropical countries. SEP has also been described in the literature as peritonitis chronic fibrosa incapsulata and its severe form, the abdominal cocoon. Prolonged treatment with beta adrenergic blockers, continuous ambulatory peritoneal dialysis, peritoneal shunts, intraperitoneal instillation of drugs, connective tissue diseases and malignancies have been reported to be some of the causes. It usually presents with recurrent episodes of intestinal obstruction and abdominal pain and distension/mass. Ascites is much rare in the setting of abdominal cocoon. [6] Our patient presented uniquely in the sense that he was a young male from the tropics, without any predisposing factors and presented initially with ascites that rapidly progressed to abdominal cocoon formation. The idiopathic condition was first described by Foo et al in 1978. [7] Grossly, the condition is characterized by a thick, fibrotic cocoon like membrane partially or totally encasing the small bowel thereby presenting with features of small bowel obstruction/abdominal mass. Most cases are diagnosed intraoperatively at laparotomy or by means of a barium follow through which shows the classical 'concertina pattern or cauliflower sign'. The CT of the abdomen revealed the presence of segregated small bowel loops towards the center of the abdomen, encased in a thick ring of peritoneal and omental tissue. The other CT features include signs of obstruction, as seen by the 'small bowel feces sign' in our patient, adherent bowel loops, bowel wall thickening, localized fluid collections and peritoneal thickening and enhancement and reactive adenopathy which was also evident in our case. [8] Ascites is usually an uncommon

feature in abdominal cocoon, which was actually the presenting feature in our patient. [9] The 'accordion sign' is classical of Clostridium difficile colitis in which there is oral contrast material trapped in between colonic folds thickened by submucosal edema. In our patient there was an 'accordion' like phenomenon seen in the small bowel region, secondary to submucosal edema of the small bowel in association with contrast trapping in the enhanced edematous small bowel walls. [10] In an idiopathic case scenario of cocoon formation, the histopathological evaluation reveals only thickened vascular fibrocollagenous tissue with chronic inflammatory reaction with plasma cell and lymphocytic infiltrates. The surrounding enhancing nodes may reveal the non specific reactive hyperplasia. [11]

Vorhauer et al in 1976 had first demonstrated the presentation of gastric adenocarcinoma leading to cocoon formation. [12] Yanagi et al in 1999 demonstrated the occurrence of membranous encapsulation of small bowel loops in early gastric cancer which was eventually surgically managed and medically managed post surgically with trimebutine maleate. [13] Recently, Reyna Pamo et al published a series on abdominal cocoon in which two out of three cases were secondary to gastric adenocarcinoma and one related to ovarian the coma. [14] Luteinizing ovarian carcinomas have been shown to cause SEP in women more frequently than has malignancy in men. [15]

CONCLUSION

To our knowledge the occurrence of SEP secondary to metastatic gastrointestinal malignancy in a young male from the tropics, without any predisposing factors to cocoon development, mimicking abdominal tuberculosis, with a speedily progressive course has not been documented erstwhile. This also warrants an astute clinical acumen to consider devastating malignant processes in young males who present with abdominal cocoon in tropical areas and in whom infective causes should not be the only biased diagnosis.

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