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Editorial

Abdominal cocoon (Sclerosing Encapsulated Peritonitis): A rare entity

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Sclerosing encapsulated peritonitis (SEP) or abdominal cocoon is life threatening condition presented with acute or chronic intestinal obstruction with abdominal pain. It's a rare entity with a prevalence rate of 0.5 to 0.9 %.[1,2] In the literature two main types of SEP are described as primary or idiopathic called as abdominal cocoon with unknown cause and 2nd type as secondary due to previous surgical interventions or operations, medical illness like tuberculosis or sarcoidosis or by some drugs.[1] It's a curious disease and high index of suspicion is required for its diagnosis. Preoperative diagnosis are impossible in last[2,3] decades but due to imaging advances its diagnosis in today's era were done but still it's difficult for the operating surgeon preoperatively.[1,2] SEP is most common in countries like China, Malaysia, South Africa and Turkey with high prevalence in young girls.[2]

We encountered it as abdominal cocoon or primary sclerosing encapsulated peritonitis in a 32 years male patient presented with severe abdominal pain in our surgical department. Computerized tomography abdomen was done showed clumped up bowel loops. Emergency laparotomy was done and excision of terminal ileum, IC junction and caecum was done and end to end anastomosis was done and specimen of clumped bowel loop was sent for histopathological examination.

We received excised terminal ileum, IC junction and caecum totally measuring 24 cms. External examination showed coiled bowel loops and adherent with blind ends and showed multiple grey white nodular areas, largest nodule measure 3x2 cms with elevated regions. On cutting open showed multiple thickened parts with many whitish membranous parts dividing the bowel loops into different segments with blind areas. Large areas of fibrosis with adhesions and membranous areas noted throughout the bowel.(**Figure 1**) Both peripheral margins are grey brown in color with fecal matter.

Light microscopy shows small intestinal mucosa with mild diffuse infiltration by lymphocytes throughout the mucosa. Submucosa showed edema and congested blood vessels. Muscular hypertrophy is evident in all sections with hypertrophy of both muscles (**Figure 1**). Large fibrocollageneous tissue encasing upto serosa is noted with large areas of fibrosis up to serosal layers. Serosa showed diffuse lymphocytic infiltration. Both the peripheral margins showed diffuse mononuclear cells infiltration.No evidence of tuberculosis or malignancy in the sections studied noted. Final histopathological impression was given as Sclerosing encapsulated peritonitis - type II- s/o abdominal cocoon.

It's a rare entity comprising of fibrocollageneous membrane ensheathing the small intestine leading to intestinal obstruction.[3] Etiopathogenesis of SEP is complex and less understood. Many authors thinks that its mutlifactorial. Repeated and chronic intra-abdominal inflammation leading to formation of fibrous tissue bands and sheaths covering bowel loop leading to obstruction as its motility is compromised. These repeated episodes leading to formation of whitish, leathery thick fibrocollageneous sheet like structure around small intestine giving rise to cocoon like appearance.[3,4] Primary and secondary cases justifies its pathogenesis. Many cases of SEP were presented with nonspecific symptoms mainly to severe abdominal pain

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depending on four phases and three types of sclerosing encapsulated peritonitis as per Anandi A et al.[4]

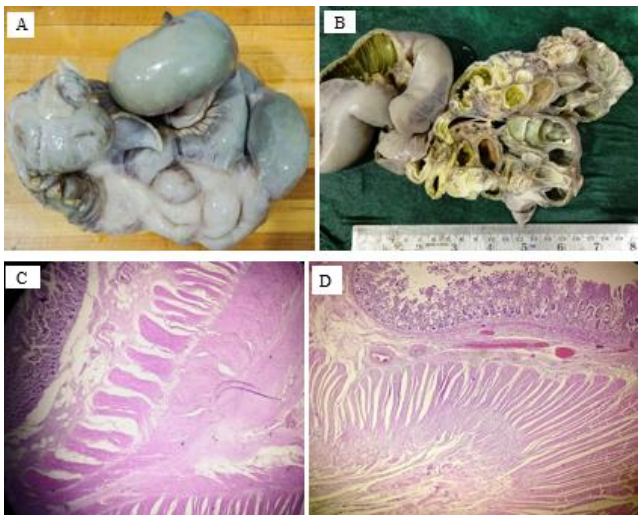

Figure 1: A: Gross examination showed coiled and clumped up bowel lops with multiple whitish nodules; **B:** Cut section showed multiple thickened parts with many whitish membranous parts dividing the bowel loops into different segments with fibrosis with adhesions; **C:** Light microscopy showed small intestinal mucosa inflammation and submucosa showed edema and congested blood vessels. (H&E,x400); **D:** Muscular hypertrophy and large fibrocollageneous tissue encasing upto serosa is noted with large areas of fibrosis up to serosal layers. (H&E,x100).

Due to its rare and complex presentation, diagnosis is very challenging to operating surgeon in SEP. No specific markers are available of this disease. High degree of suspect with aid from imaging modalities like CT scan and USG may be helpful most of the times. Complete surgical excision sac with bowel loops is the mainstay of the treatment. Prognosis after the surgery is favorable in most of the cases, hence proper diagnosis and timely treatment is main crux in the management of cocoon syndrome patients.

The aim of this editorial to highlight the rarity and complex presentation of abdominal cocoon. High index of suspiciousness, timely management by surgical excision is crucial for better, favorable outcomes to SEP patients.

Happy and Prosperous Diwali to all academic readers.

Regards,

Dr. Dhiraj B Nikumbh

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