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Idiopathic Sclerosing Encapsulated Peritonitis- Case Series and Review of Literature

Authors
Gulab Dhar Yadav[1], Ramendra Kumar Jauhari[2], Anurag Singh[3], Ujjwal Pal Singh[4], Amulya Singh[5]
[1]Professor, [2]Associate Professor, [3]Assistant Professor, [4]Junior Resident, [5]Senior Resident
Department of General Surgery, G.S.V.M Medical College, Kanpur, India

Abstract
Introduction: *Idiopathic Sclerosing encapsulated peritonitis (SEP) is a rare cause of recurrent intestinal obstruction mainly in young and adolescent females from tropical and subtropical regions. SEP can be primarily classified as idiopathic or secondary-due to some known etiology.*
Case Summary: *During a period of eight years we encountered five cases of idiopathic SEP. All patients presented with recurrent intestinal obstruction and managed operatively.*
Discussion: *SEP is a rare condition in the clinics. Etiology is relatively unknown. Most cases are diagnosed incidentally at laparotomy, and a better awareness of this entity and the imaging techniques may facilitate pre-operative diagnosis.*
Conclusion: *Primarily the diagnosis is made incidentally at laparotomy. Management of SEP is debated. But most clinicians agreed that surgical treatment is required in symptomatic patients.*
Keywords: *Idiopathic, Sclerosing, Encapsulating peritonitis, fibrous capsule.*

Introduction

Sclerosing encapsulating peritonitis or abdominal cocoon is a rare condition which refers to partial or total encapsulation of abdominal viscera within a dense fibrous membrane. It is an uncommon cause of subacute small bowel obstruction. It has been referred to as peritonitis chronic fibrosaincapsulata by Owtschinnikow, 1907and first described in detail by Foot et.al. 1978. Abdominal cocoon is predominantly reported among female from tropical and subtropical regions, however cases of the adult male were also reported. It can be classified as primary (idiopathic) or secondary. Correct diagnosis is not often made as CT scan and MRI finding are non-specific but they facilitate preoperative diagnosis.

Cases summary

In our study five patients presented to our hospital over a period of 2010 to 2018 with acute, subacute and chronic intestinal obstruction. All of them underwent surgery in our institute. This case study was done retrospectively for clinical presentation, operative findings and postoperative outcome. The average age of the patients was 28.8 years range 15 to 50 year, with three female and two male. All patients presented with classical signs and symptoms of intestinal obstruction-abdominal pain, vomiting, abdominal distension and constipation varying over duration of 6 months to 5 years. Plain abdominal X-ray was insignificant. CT scan ABDOMEN showed small

bowel dilatation and clumping in center of the abdomen as shown in figure 1.

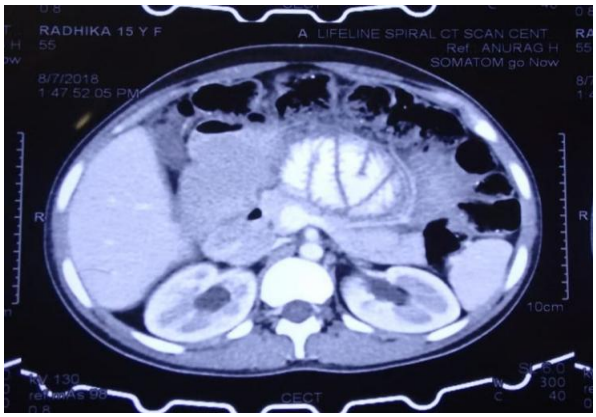

Figure 1(a)

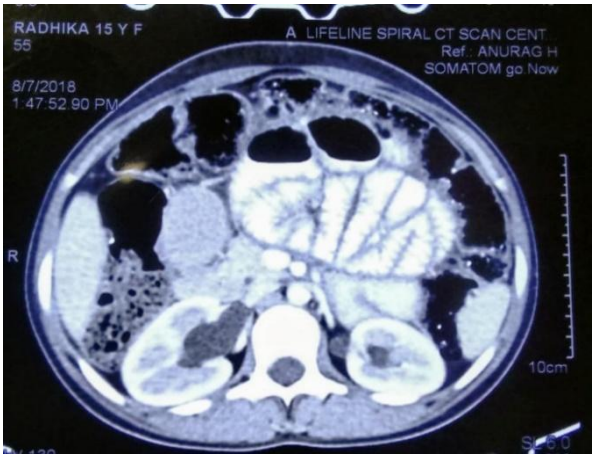

Figure 1(b)

Figure 1. Axial views of contrast-enhanced CT Scanabdomen shows small bowel loops are dilated and clumped in center of the abdomen

All other causes of similar pathology have been ruled out. Exploratory laparotomy was done as symptoms did not resolve even after conservative treatment. Intraoperative findings were almost similar in all cases, thick white capsule of size 4mm to 10mm seen encasing small bowel. In three cases complete encasement of small bowel seen, in one case partial encasement of small bowel seen and in one case encasement seen all over small bowel, ceacum and appendix. Greater omentum hypoplasia seen in all cases (Figure 2).

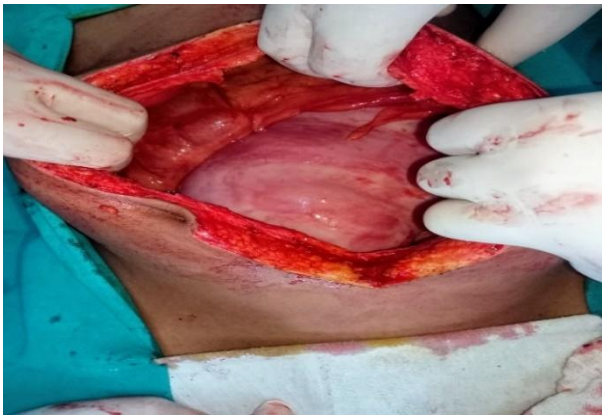

Figure 2. Laparatomy showed thick white fibrous membrane encasing whole small bowel

During surgery excision of the thick fibrous layer and inter-loop adhenolysis done to relieve the small bowel obstruction. In case of extended encasement of ceacum and appendix prophylactic appendectomy was done. In one case ileostomy done due to small bowel perforation during adhenolysis. All other patient's surgery went uneventfully except for some minor serosal tear in all cases (Figure 3 and 4).

Three cases have an uneventful postoperative period and passed stool spontaneously on postoperative day 4 to 5. Ileostomy become functional on postoperative day 2. During follow up one patient develop Enterocutaneous fistula which was managed conservatively due to which patient postoperative stay prolonged. All other patient discharged from 8 to 12 postoperative day. Histology of all cases shows report of thick fibro collagenized tissue bundles with hyalinization, focal fat necrosis surrounded by the cluster of foamy macrophages and chronic inflammatory cells are seen the area of edematous and vascularised tissue with the mixed inflammatory infiltrate also seen which are suggestive of inflammatory fibrosclerosis.

Ileostomy closure done after 3 months for one patient which was uneventful. On further follow-up no patient presented with recurrence of symptoms till now.

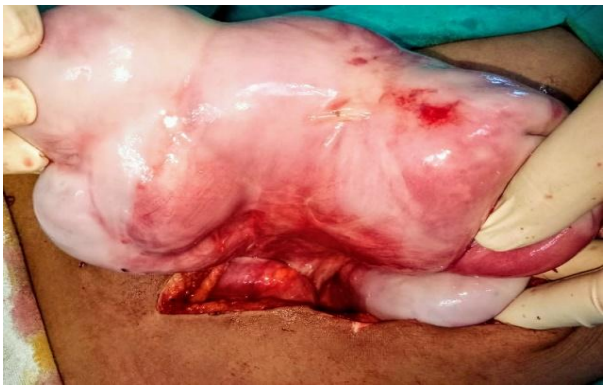

Figure 3. Intraoperative findings whole of small bowel encased in thick white fibrous sac appendix was inside the sac

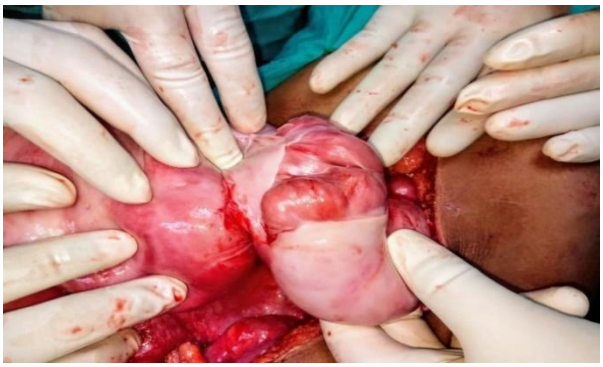

Figure 4. On excision of thick fibrous membrane small bowel loop seen inside with interbowel adhesions

In one case appendix histopathology shows mixed inflammatory infiltration of mucosal surface and underlying all the layers up to serosa with large hypertrophied lymphoid follicle seen suggestive of acute or chronic appendicitis.

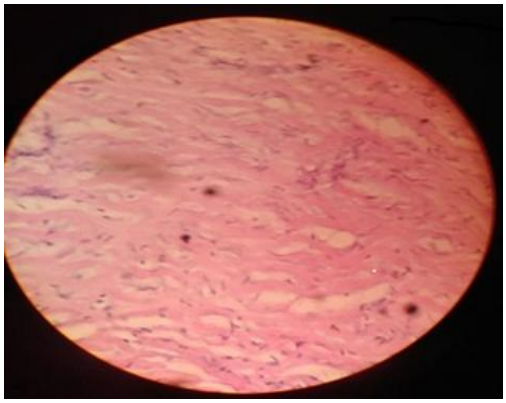

Figure 5. Histology shows thick fibro-collagenized tissue bundles with hyalinization, focal fat necrosis surrounded by the cluster of foamy macrophages and chronic inflammatory cells are seen suggestive of sclerosing mesentritis

Discussion

Idiopathic sclerosing encapsulating peritonitis (SEP) is a rare condition in the clinic. Etiology is relatively unknown. It is characterized by a thick grayish-white fibrotic sac type membrane, partially or totally encasing the small bowel[1, 2, 3, 4] the thick fibrous layer can extend to involve other organs like the ceacum, appendix, large intestine, liver, and stomach. In our study, one patient has a thick fibrous layer involving small bowel with caecum and appendix. Clinically, it presents with recurrent episodes of abdominal pain, subacute or chronic small bowel obstruction, weight loss, nausea, and anorexia, with a palpable abdominal mass, but some patients may be asymptomatic[1, 2, 5- 8]. In the current study all patients present with symptoms of recurrent bowel obstruction. SEP can be classified as primary (idiopathic) or secondary the etiology of the primary form is of uncertain[9-12].

The secondary form of SEP is more common has been reported in association with continuous ambulatory peritoneal dialysis. Other rare causes include abdominal tuberculosis systemic lupus erythematosus gastrointestinal malignancy sarcoidosis, Beta-blocker practolol intake, ventriculo-peritoneal and peritoneovenous shunts, orthotopic liver transplantation and recurrent peritonitis[11, 13, 14]. All of these etiology were absent in our patients. The idiopathic form also known as abdominal cocoon has been classically described in young and adolescent females from the tropical and subtropical countries[1, 2, 5, 8, 16, 18, 21]. In the present study there are 3 females who are of young or adolescent age group and from north India. The etiology of this entity has remained relatively unknown. To explain the etiology, a number of hypotheses have been proposed. These include retrograde menstruation with a superimposed viral infection, retrograde peritonitis and cell-mediated immunological tissue damage incited by gynecological infection[5,12,14,15]

Sr.no	Age (Yrs)	Sex	Clinical Presentation	Intra-Operative Findings	Procedure Done	Histology Finding	Outcome
1	18	F	Intestinal obstruction	Complete encasement of small bowel	Excision of the fibrous layer with adhenolysis	sclerosingmesenteritis (inflammatory fibrosclerosis)	Uneventful
2	37	M	Intestinal obstruction	Complete encasement of small bowel	Excision of the fibrous layer with adhenolysis with ileostomy	sclerosingmesenteritis (inflammatory fibrosclerosis)	Ileostomy closure after 3 months and no recurrence of symptoms till now
3	24	F	Intestinal obstruction	Complete encasement of small bowel with caecum and appendix	Excision of the fibrous layer with adhenolysis	sclerosingmesenteritis (inflammatory fibrosclerosis)	Uneventful
4	15	F	Intestinal obstruction	Complete encasement of small bowel	Excision of fibrous layer with adhenolysis	sclerosingmesenteritis (inflammatory fibrosclerosis)	Enterocutaneous fistula
5	50	M	Intestinal obstruction	Partial encasement of small bowel	Excision of fibrous layer with adhenolysis	sclerosingmesenteritis (inflammatory fibrosclerosis)	Uneventful

However, since this condition has also been seen to affect males, premenopausal females and children, there seems to be little support for these theories. In our study, there are 2 males. Further hypotheses are therefore needed to explain the cause of idiopathic SEP. Since abdominal cocoon is often accompanied by other embryologic abnormalities such as greater omentumhypoplasia, and developmental abnormality may be a probable etiology[3, 5, 7, 14, 16]. In our study all patients present with greater omentum hypoplasia .To elucidate the precise etiology of idiopathic SEP, further studies of cases are necessary. Our cases are primary (idiopathic sclerosing encapsulated peritonitis) as no etiology can be identified.

Although it is difficult to make a definite pre-operative diagnosis, most cases are diagnosed incidentally at laparotomy, and a better awareness of this entity and the imaging techniques may facilitate pre-operatively diagnosis. Ultrasonography may show a thick-walled mass containing bowel loops, loculated ascites, and fibrous adhesions[2, 8,14,17,18]. In some of our cases, we successfully make probable diagnosis of abdominal cocoon by a combination of abdominal CT and clinical presentations. The characteristic findings of CT include that small bowel loops congregated to the center of the abdomen, encased by a soft-tissue density mantle which cannot be contrast enhanced, and inter-bowel ascites was demonstrated in some cases[2,8,16,18].

Management of SEP is debated. But most clinicians agreed that surgical treatment is required. In our study all cases were managed by surgery. During surgery, in addition to careful dissection and excision of the covering membrane, dense inter bowel adhesions also need to be freed for complete recovery. In order to avoid complications of postoperative intestinal leakage and short bowel syndrome, resection of the bowel is indicated only if it is nonviable[1,2,6,8,16].

In our study, no resection of bowel needed, in one case ileostomy done due to perforation of the bowel during adhenolysis. No surgical treatment is required in asymptomatic SEP. Surgical complications were reported including intra-abdominal infections, enterocutaneous fistula, and perforated bowel[1, 8,12,19,20]. In our study one case develop enterocutaneous fistula which was managed conservatively. No recurrence of

symptoms and complication were described in postoperative follow-up.

Conclusion

Idiopathic sclerosing encapsulating peritonitis (SEP) is a rare cause of recurrent intestinal obstruction. It occurs in all age group but common in young females of subtropical and tropical regions .In most cases etiology is unknown and managed by careful dissection and excision of thick sac with the release of intraloop adhesions. Clinical features and CT scan is helpful in making preoperative diagnosis. Definite diagnosis is made intraoperatively Preoperative diagnosis is helpful to reduce intraoperative complications..Prognosis after surgery is good as recurrence of symptoms is rare.

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