Abdominal Cocoon Syndrome-Enigmatic and Unconventional Entity of Intestinal Obstruction

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Received: 04 Mar 2020 Accepted: 15 Mar 2020 Published: 20 Mar 2020

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1. Abstract

Abdominal cocoon syndrome, the idiopathic form of sclera sing encapsulating peritonitis, is a rare condition of unknown etiology that results in an intestinal obstruction due to total or partial encapsulation of the small bowel by a fibro collage nous membrane (cocoons) [1]. A major challenge faced by clinicians while dealing with this entity is that the early clinical features are nonspecific and often not recognized, making the pre-operative diagnosis difficult. In this case we aimed to present a patient diagnosed with abdominal cocoon syndrome peri-operatively.

2. Keywords: Abdominal cocoon syndrome; Peritonitis chronic fibrosain capsulate; Sclerosing encapsulating peritonitis; Primary capsulating peritonitis.

3. Introduction

It was first described by Owtschinnikow in 1907 as "peritonitis chronica fibrosa incapsulata" [2] and termed "abdominal cocoon" by Foo in 1978.

It was first described by Owtschinnikow in 1907 as "peritonitis chronica fibrosa incapsulata" [2] and termed "abdominal cocoon" by Foo in 1978 [2, 3]. The etiology of abdominal cocoon is unknown and most often it is found in adolescent girls from tropical or subtropical countries [4]. Abdominal cocoon syndrome is also known as Peritonitis chronic fibrosain capsulata, sclerosing encapsulating peritonitis and Primary sclerosing peritonitis

Most patients present with features of recurrent acute or chronic small bowel obstruction secondary to compression of the intestines within the constricting cocoon [5]. Excision of the thick sac with the release of the small intestine leads to complete recovery in the vast majority of cases.

4. Case Report

A 21 year old female who is a diagnosed case of portal vein thrombosis presented to the OPD with chief complaints of diffuse pain in abdomen, multiple episodes of vomiting and abdominal distention. Abdominal pain was insidious in onset, gradually progressive and localized to epigastric and umbilical region. Patient also gives history of significant weight loss. No history of bladder complaints.

General examination revealed no significant findings and on palpation, a mass was felt in the hypogastric and left iliac region. Bowel sounds were sluggish.

5. Ascitic Fluid Findings

5.1. Physical Examination

Quantity: 3ml

Color: Reddish

Appearance: Turbid

Coagulum: Absent

Clot/Cobweb: Absent

ADA Level: normal

5.2. Microscopic Examination

Total nucleated cells: 800/cmm

RBC's: present

Impression: Lymphocytes: 70%

Macrophages: 23%

Neutrophils: 6%

Eosinophils: 1%

Gram stain: No pus cells seen. No organism seen.

ZN Stain: Acid Fast Bacilli not seen

GeneXpert: MTB not detected

Fluid Culture and Sensitivity: No growth seen.

5.3. Radiology Findings

X-ray Abdomen (Figure 1)

5.3.1. Ultrasonography: Jejunal loops appear clumped and are seen in left para umbilical region, normal in diameter with sluggish peristalsis. Mild ascites present.

5.4. Intraoperative Findings and Further Investigations

- Severe inter bowel and peritoneal adhesions.
- Supracolic area could not be entered due to dense adhesions.
- Intracolic loculated cyst formation in the peritoneal sac with cocooneal small bowel mass (bowel wall thickened) with multiple cyst.
- Uterus and ovary palpable inside the cyst. (S/O loculated ascitis) left ovarian cyst

5.5. Histopathology

5.5.1. Gross: Specimen of capsule covering the bowel consists of single fibrofatty piece of tissue measuring 2x1x0.5cm

Specimen of omentum consists of single rediish white flap of tissue measuring 2x1x0.3cm

5.5.2. Microscopy: Section of capsule covering the bowel showed showed fibrofatty tissue along with lymph node showing features of reactive lympadenitis. No evidence of granuloma/atypia

Section of omentum showed large number of congested blood vessels and chronic non-specific inflammatory infiltrate and mild fibrosis.

6. Discussion

Bowel obstruction is a common surgical emergency. However, at times, unusual cases of bowel obstruction such as Abdominal Cocoon (AC) may be encountered. In this condition, a whitish, thickened membrane encapsulating the small bowels as a 'cocoon' is seen on laparoscopy (Figure 2 and 3). Early postoperative small bowel obstruction is a potential complication of extensive adhesiolysis. Therefore, special intra operative care including gentle tissue dissection, careful haemostasis and thorough peritoneal cleansing should be taken. The long-term prognosis of abdominal cocoon syndrome is usually favorable [4]. In our case, laparoscopy was a useful tool both for the definitive diagnosis and the treatment of this condition, leading to a favorable postoperative outcome.

7. Conclusion

A contrast based computed topography or a diagnostic laparoscopy along with clinician awareness especially in adolescent females proves to be a helpful aid for pre-operative diagnosis. Since etiology of this disease is unknown and it is found mostly in adolescent girls it must always be kept as a possible diagnosis.



Figure 1: Xray abdomen was unremarkable



Figure 2: small bowels as a 'cocoon'



Figure 3: small bowels as a 'cocoon' is seen on laparoscopy

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