ANOTHER LOOK AT THE "ABDOMINAL COCOON SYNDROME"

*Ibrahim NA *Oludara MA *Adewale AO

*Department of Surgery,
Lagos State University College of Medicine/
LagosState University Teaching Hospital,
Ikeja, Lagos.
Nigeria.

Corresponding author: Dr. N.A. Ibrahim
Email: ibrahimakanmu@yahoo.com

***Dr. Ibrahim is a Consultant General Surgeon at the Lagos State University Teaching Hospital, Ikeja**

ANOTHER LOOK AT THE "ABDOMINAL COCOON SYNDROME"

Abstract

Abdominal cocoon is a rare cause of bowel obstruction. The condition was first described three decades ago and about 106 cases have so far been reported in the English literature. It is found mainly among adolescent females in tropical and sub-tropical countries. It is characterized by a thick fibrous membrane encapsulating the small intestines partially or completely. It should be differentiated from Encapsulating Peritoneal Sclerosis (EPS) and Peritoneal Encapsulation (PE) which are sometimes referred to as abdominal cocoon but are of different aetio-pathology. Patients present with features of intestinal obstruction and the diagnosis is usually made intra-operatively. Surgical excision of the membrane with adhesiolysis in an emergency exploratory laparotomy results in complete recovery. Appropriate management of this condition requires adequate knowledge about the disease. We present a review of this rare entity.

Key words: *abdominal cocoon, intestinal obstruction*

Introduction

Abdominal cocoon, first described by Foe et al.[1] in 1978 is a rare cause of intestinal obstruction. It primarily affects adolescent females. However, cases have been reported among males, [2] the elderly [3] and children. [4] This condition commonly affects residents of tropical and subtropical regions. Majority are from the continents of Asia and Africa where cases have been reported from China, [5] South Korea, [6] Japan, [7] Singapore, [1] Malaysia, [8,9] Australia, [10] India, [11,12,13] Turkey, [14] Middle East, [2,15,16] East Africa [17] and West Africa [18,19,20]. Cases found outside these regions were among those that lived in the tropics before migrating to Europe. [21, 22] Among recent reports is that of an adolescent male patient seen at the Lagos State University Teaching Hospital in Lagos, Nigeria. [20] Medline search revealed that approximately 106 cases have so far been reported in the English literature.

Etiology

The aetiology of this entity which commonly presents with features of recurrent acute or sub-acute intestinal obstruction is not fully known. This condition was found among adolescent girls in the initial reports and it was postulated that retrograde menstrual flow with super-imposed viral infection resulted into low grade peritonitis and subsequent cocoon formation. [1] The possibility of spread in low grade gynaecological infections through the fallopian tube to the peritoneum was reported by Stassewn et al. [23] However, this mechanism cannot explain the occurrence of this disease in children and male patients. Haematogenous sub-clinical peritonitis has been suggested as a possibility; however, it was observed that this type of peritonitis occurs mainly among patients with ascites from cirrhosis, nephrosis, malignancy and cardiac failure. [24] These conditions are not present in patients with this disease. Report of abdominal cocoon in identical twins lends support to the possibility of genetic susceptibility to developing the condition. [11] Abdominal cocoon is often accompanied by other embryonic abnormalities such as greater omentum hypoplasia and mesenteric vessel malformation. [5]

Abundance of collagen tissue on histo-pathological examination of excised cocoon membranes and presence of collagen balls in ascitic fluid cytology in this condition suggest a significant role for collagen in the pathogenesis. [25] In addition, keloid formation in the abdominal wall scars of

four out of the ten cases reported by Foo et al. and the identical twins also suggest collagen abnormality in this condition. It is hypothesized that abdominal cocoon is a developmental malformation with genetic predisposition in which, collagen tissue disorder plays important role. [11] This condition has not been reported in neonates and infants. Therefore, this hypothesis may be a less likely explanation of the aetiology. Further work is still required to identify the cause of this rare entity.

Pathology

This condition is characterized by the presence of a thick, fibrous, cocoon-like membrane within which the small bowel is partially or totally encased. The parts of the bowel within the membrane are matted together by soft adhesions and may be dilated proximal to point of obstruction (Figures 1&2). The encasement, in some cases, involves the colon, [4] liver [20] or stomach. [5] The membrane consists of proliferating fibro-connective tissue and chronic non-specific inflammatory cells. In some cases, it is formed by mature fibrous tissue without evidence of inflammation. The encasing membrane and adhesions between bowel loops are usually easily separable from serosa of involved intestines.

It is important to differentiate abdominal cocoon from the following conditions with similar cocoon formation:

1. Encapsulating Peritoneal Sclerosis (EPS)

This condition has known underlying aetiology. It is characterized by a thick greyish white fibrous membrane covering the small intestine and causes intestinal obstruction. It is a rare complication of chronic ambulatory peritoneal dialysis. The frequency of this condition among peritoneal dialysis patients is from 0.54% to 4.4%. [26] Other reported causes of SEP include Practolol which is a beta blocker, peritoneo-venous shunting, sarcoidosis, systemic lupus erythematosis, peritoneal and intra-abdominal malignancy, endometriosis, tuberculosis and gastrointestinal diseases. [26] It is a more serious condition with high mortality and usually treated by non-surgical methods. The underlying causes are treated medically and patients who are on peritoneal dialysis are switched to haemodialysis. [27] Varieties of terms that have been used indiscriminately to describe this condition are peritoneal fibrosis, peritoneal sclerosis, sclerotic

thickening of the peritoneal membrane, sclerotic obstructive peritonitis, calcific peritonitis, abdominal cocoon and in recent years, sclerosing encapsulating peritonitis (SEP). It is believed that a more accurate description would be "encapsulating peritoneal sclerosis'' which is more descriptive of the morphological changes. [26] Some authors refer to abdominal cocoon as "Idiopathic Sclerosing Encapsulating Peritonitis". Use of this terminology for abdominal cocoon should be avoided because SEP is a distinct entity. The membrane in this condition is firmly attached to bowel serosa and does not separate easily; differentiating it from the membrane in abdominal cocoon. [26]

2. Peritoneal Encapsulation (PE)

This is a very rare condition characterized by the presence of an accessory peritoneal sac which covers the small intestine. However, the serosa of the small intestines is not covered by the fibro-collagenous membrane seen in abdominal cocoon. It was first described by Cleland 1868 [28] and less than 20 cases have been reported in the English literature. [29] It is a congenital condition resulting from abnormal return of the mid-gut loop to the abdominal cavity during early development. The accessory sac is derived from the original dorsal mesentery which normally forms the transverse mesocolon. [30] Patients with this condition are usually asymptomatic but few may present with intestinal obstruction. [29] Surgical excision of the accessory sac results into recovery. This entity can be confused with abdominal cocoon. A case reported by Devay et al. as abdominal cocoon was found to be peritoneal encapsulation after review of clinical picture. [31]

Clinical Features and Investigations

Patients with abdominal cocoon usually present with clinical features of acute, sub-acute or chronic intestinal obstruction. Recurrent attacks of colicky abdominal pain, vomiting, abdominal distension and constipation are the main presenting symptoms. Weight loss, nausea, anorexia and a palpable abdominal mass may be present in this condition. Duration of symptoms in the cases reported so far ranged from few hours to ten years. An unusual presentation of abdominal cocoon with jaundice and increasing abdominal distension was reported in a 25 year old Chinese woman. [32] In a review by Devay et al. [33] abdominal pain, with or without other symptoms of obstruction was present in 38(97%) out of 39 reported cases. Abdominal mass was also present

in 17(44%) patients. Diagnosis of abdominal cocoon is rarely made pre-operatively because of the non-specific nature of the clinical and imaging findings. Plain x-ray of the abdomen may show dilated bowel loops and multiple air-fluid levels indicating bowel obstruction. Abdominal sonography may also reveal evidence of bowel dilatation and intra-abdominal mass. Rokade et al. [34] made a pre-operative diagnosis of abdominal cocoon in a 26 year old female patient through ultrasonography. The sonographic picture showed a clustered pattern of matted small bowel loops and the covering membrane. Upper gastrointestinal contrast studies can aid pre-operative diagnosis. A useful finding is the presence of "Cauliflower sign" which describes a fixed cluster of dilated small-bowel loops lying in a concertina-like fashion. [6] In addition, delayed transit time may be observed in small bowel contrast study.

Computed tomography (CT) is one of the reliable tools in the investigation of intestinal obstruction. Characteristic feature of this condition on CT of the abdomen is a concentration of multiple small bowel loops in the centre of the abdomen surrounded by thick enhancing saclike structure. Gupta et al. reported a case of a patient who presented with mild intestinal obstruction and who had typical pre-operative CT findings of an abdominal cocoon, which was confirmed at surgery. [12]

MRI can display the typical features of abdominal cocoon seen in both barium studies and CT. This further enhances pre-operative diagnosis of this rare condition.[15] Capsule endoscopy is a novel diagnostic tool used in the diagnosis of gastro-intestinal disorders. Although, it is not commonly employed in intestinal obstruction, pre-operative diagnosis of a case of abdominal cocoon by this technique has been reported. [35] Laparoscopy has been used as well in the diagnosis of some cases of abdominal cocoon.[13]

Treatment

Abdominal cocoon requires emergency surgical treatment. Nearly all reported cases were diagnosed during exploratory laparotomy for intestinal obstruction. Unlike post-surgical adhesive bowel obstruction which may resolve with expectant treatment, cocoon adhesions need adhesiolysis before recovery can be achieved. Adequate pre-operative preparation is essential in all patients. A midline laparotomy incision is usually employed to gain access into the peritoneal cavity. The usual finding is the thick, whitish, cocoon-like membrane encasing the dilated small intestines partially or completely. The membrane, in some cases, covers other intra-abdominal

viscera including stomach, liver and large bowel. Lysis of adhesion is carried out by carefully incising the covering membrane and separating it from the serosa of bowel using a combination of blunt and sharp dissections. Intra-loop adhesions are also similarly separated. Although the process of separation is usually easy, care is taken to avoid bowel injury. Bowel resection is not required unless non viable intestine is encountered. Resection was necessary in 5(14%) patients among 36 reported cases reviewed by Devay et al. [33] Successful excision of cocoon membrane and freeing of entrapped bowel during diagnostic laparoscopic procedure has been reported. [13] Incident appendectomy is recommended. It is believed that the appendix may be difficult to locate if patient subsequently develops acute appendicitis and requires surgery. [36] However, prevention of another abdominal surgery for appendicitis in these patients who could have developed post-operative bowel adhesions appears to be more convincing.

Complete long-term recovery is expected after excision of the membrane.

Although complications are rare following the procedure, early post-operative intestinal obstruction, [37] intra-abdominal infections, entero-cutaneous fistula and perforated bowel were reported. [5] Recurrent abdominal cocoon is rarely seen. A 26 year old woman who had laparoscopic adhesiolysis for abdominal cocoon presented 6-7 months later with a recurrence for which she had laparotomy. [34]

Conclusions

Abdominal cocoon remains a rare cause of intestinal obstruction and should be differentiated from Sclerosing Encapsulating Peritonitis and Peritoneal Encapsulation. Pre-operative diagnosis is still difficult and a high index of suspicion is required. Recurrent symptoms of bowel obstruction are characteristic and surgical excision of the encasing membrane with adhesiolysis results in recovery. Awareness of this condition by surgeons especially those practicing in the tropical and subtropical regions will facilitate appropriate management of the patients.

References

1. **Foe KT, Ng KC, Rauff A, Foong WC, Sinniah R.** Unusual small intestinal obstruction in adolescent girls; the abdominal cocoon. *Br J Surg* 1978; 65(6):427-430.
2. **Deeb LS, Mourad FH, El-Zein YR, Uthman SM.** Abdominal cocoon in a man: preoperative diagnosis and literature review. *J Clin Gastroenterol* 1998; 26:148–150.
3. **Rajagopal AS, Rajagopal R.** Conundrum of the cocoon: report of a case and review of the literature. *Dis Colon Rectum.* 2003; 46:1141–1143.
4. **Sahoo SP, Gangopadhyay AN, Gupta DK, Gopal SC, Sharma SP, Dash RN.** Abdominal cocoon in children: a report of four cases. *J Pediatr Surg* 1996; 31:987–988.
5. **Xu P, Chen LH, Li YM.** Idiopathic Sclerosing Encapsulating Peritonitis (or abdominal cocoon): A report of 5 cases. *World J Gastroenterol.* 2007; 13(26):3649-3651.
6. **Hur J, Kim KW, Park M, Yu J.** Abdominal Cocoon: Pre-operative Diagnostic Clues from Radiologic Imaging with pathologic correlation. *AJR* 2004; 182: 639-641.
7. **Masuda C, Fujii Y, Kamiya T, et al.** Idiopathic sclerosing peritonitis in a man. *Intern Med.* 1993; 32:552-555.
8. **Yip FWK, Lee SH.** Abdominal cocoon. *Austr N Z J Surg.* 1992; 62:638-642.
9. **Seng LK, Mahadaven M, Musa A.** Abdominal cocoon: a report of two cases. Br J. Surg 1993;80(9):1144.
10. **Le C, Pitcher ME, Jones T.** The Abdominal Cocoon. *Aust. NZ J. Surg.* 1996; 66:777.
11. **Srivastava P, Sahu M.** Abdominal cocoon in identical twins: Is it congenital collagen tissue disorder? Report of two cases with review of literature. *Indian J Surg.* 2007; 69(1):25-27.
12. **Gupta S, Shirahatti RG, Anand J.** CT Findings of an Abdominal Cocoon. *AJR.* 2004; 183:1658-1660.
13. **Makam R, Chamany T, Ramesh S, Potluri VK, Varadaraju PJ, Kasabe P.** Laparoscopic management of abdominal cocoon. *J Min Access Surg* 2008;4:15-7
14. **Altinli E, Sumer A, Celic A.** Abdominal Cocoon: A rare cause of intestinal obstruction. *Israeli Journal of Emergncy Medicine.* 2007; 7(3):42-44.

15. **Manassa-Moussa L, Bleibel L, Sader-Ghorra C, Smayra T, Aoun NJ.** MRI findings in Intestinal Cocoon. *AJR* 2006; 186:905-906.
16. **Al-Ebrahim E, Khalifah A, Al-Hajry F.** Idiopathic Sclerosing Encapsulated Peritonitis(abdominal cocoon) A Case Report and Literature Review. *Basrah Journal of Surgery*. September, 13, 2007.
17. **Githeko AK, Mc Farlene GA.** Abdominal cocoon: report of 5 cases. *East Afr. Med.* J. 2000; 77:286-288.
18. **Marinho A, Adelusi B.** The abdominal cocoon. Case report. *Br J. Obstet Gynaecol.* 1980; 87; 249-250.
19. **Irabor DO, Atalabi O.** Case report-abdominal cocoon. *West Afr J Med.* 2001; 20(4):265-267.
20. **Ibrahim NA, Oludara MA.** Abdominal cocoon in an adolescent male patient. *Tropical Doctor.* 2009; 39:254-256.
21. **Cleffken B, Sie G, Reidl R, Heineman E.** Idiopathic sclerosing encapsulating peritonitis in a young female – diagnosis of abdominal cocoon. *J Pediatr Surg* 2008; 43(2):e27-30.
22. **Macklin J, Hall C, Feldman A.** Unusual cause of small bowel obstruction in adolescent girls: the abdominal cocoon. *J R Coll Surg Edinb.* 1991; 36:50-52.
23. **Stassen WN, McCollugh AJ, Hilton PK.** Spontaneous bacterial peritonitis caused by Niesseria Gonorrhoea. Evidence for a trans-fallopian route of infection. *Gatroenterology.* 1985; 88: 804-807.
24. **Wilcox CM, Dismukes WD.** Spontaneous bacterial peritonitis. A review of pathogenesis, diagnosis and treatment. *Medicine.* 1987; 66:447-456.
25. **Kobayashi TK, Ueda M, Nishino T, Nishida S, Yoneyama C, Watanabe S.** Appearance of “collagen balls” in ascitic fluid cytology with abdominal cocoon (encapsulating peritonitis). *Diagn cytopathol* 1997; 16:469-470.
26. **Kawaguchi Y, Kawanishi H, Mujais S, Topley N, Oreopoulos DG.** Encapsulating peritoneal sclerosis; Definition, Etiology, Diagnosis and treatment. *Peritoneal Dialysis International* 2000; 20(suppl 4):543-555.
27. **Herrero JC, Monila A, Lentisco C, et al.** Sclerosing encapsulating peritonitis: a latent threat. A change in theapproach to surgical treatment. *NEFROLOGIA* 2007; 27(6):729-736.

28. **Cleland J.** On an abnormal arrangement of the peritoneum with remarks on the developments of the mesocolon. *J Anat Physiol.* 1868; 2:201-6.
29. **Sherigar JM, McFall B, Wali J.** Peritoneal encapsulation: presenting as small bowel obstruction in an elderly woman. *Ulster Med J.* 2007;76(1):42-44.
30. **Chew MH, Sophian HI, Chan G, Ong HS, Wong WK.** A problem encapsulated: the rare peritoneal encapsulating syndrome. *Singapore Med J* 2006; 47(9):808-810.
31. **Devay AO, Gomceli I, Korukluoglu B, Kusdemir A.** Correction: An unusual and difficult diagnosis of intestinal obstruction: The abdominal cocoon. Case report and review of the literature. *World J Emerg Surg* 2008;3:36
32. **Ranganathan S, Abdullah BJJ, Sivanesaratnam V.** Abdominal Cocoon Syndrome. *J HK Coll Radiol* 2003;6:201-203
33. **Devay AO, Gomceli I, Korukluoglu B, Kusdemir A.** An unusual and difficult diagnosis of intestinal obstruction: The abdominal cocoon. Case report and review of the literature. *World J Emerg Surg* 2006;1:8
34. **Rokade ML, Ruparel M, Agarwal JB.** Abdominal cocoon. *J Clin Ultrasound.* 2007;35(4):204-205.
35. **Yang XY, Chen CX, Zhang BL, et al.** Diagnostic effect of capsule endoscopy in 31 cases of subacute small bowel obstruction. *World J Gastroenterol.* 2009; 15(19): 2401–2405.
36. **Attar ZB, Nazir Z, Moazam F.** The abdominal cocoon- an unusual cause of intestinal obstruction in adolescents. *Ann Trop Paediatr.* 1993; 13:99–102.
37. **Zheng YB, Zhang PF, Ma S, et al.** Abdominal cocoon complicated with early postoperative small bowel obstruction. *Ann Saudi Med.* 2008; 28(4):294-296.