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# A Case of Abdominal Cocoon

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***Abdominal cocoon is a rare disease of the peritoneum and almost invariably presents as an acute or subacute intestinal obstruction with or without a mass.***
***The etiology of this disease is largely unknown and abdominal cocoon of unknown etiology has been limited to the tropical and subtropical zones and primarily affects young adolescent females. In the temperate zone, only one case has been reported from the United Kingdom, but the patient was also born in Pakistan. No case of abdominal cocoon purely developed in the temperate zone has been reported.***
***Recently, we experienced a case of abdominal cocoon in a 34-year-old female patient(Korean) who had never been abroad. The diagnosis was made postoperatively by reviewing the literature. We herein report this rare condition developed in an unusual geographical location with a brief review of the literature.***

**Key Words** : *Abdominal cocoon, Temperate zone*

## INTRODUCTION

Abdominal cocoon is a rare disease of the peritoneum which refers to a condition where there is total or partial encasement of the small bowel by a dense fibrous membrane(Foo et al., 1978). This condition is usually presented as acute or subacute intestinal obstruction with or without a mass. The etiology is largely unknown. In some cases, however, association with the use of beta-blocker(Brown et al., 1974 ; Windsor et al., 1975 ; Eltringham et al., 1977 ; Harty, 1978 ; Ahmad, 1981) or liver cirrhosis with or without LeVeen shunt(Cambria and Shamberger, 1984 ; Greenlee et al., 1979) has been suggested. Abdominal cocoon of unknown etiology has primarily affected young females and shows tropical or subtropical distribution. In the temperate zone, only one case has been reported from the United Kingdom(Macklin et al., 1991). However, the reported case was also born and raised in Pakistan. To our knowledge, no case has been reported from the temperate zone.

The treatment of choice is surgical release of a cocoon encasing the bowel(Sieck et al., 1983 ; Macklin et al., 1991 ; Yip and Lee, 1992). Precise preoperative understanding of this condition can prevent unnecessary resection of the affected bowel.

Recently, we experienced a 34 year-old female patient(Korean) presenting with the symptoms and

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signs of acute intestinal obstruction. We did not appreciate this condition preoperatively and intraoperatively, and the affected bowel was resected unnecessarily. We believe that this is the first case of abdominal cocoon of unknown etiology purely developed in the temperate zone. We herein report this case with a review of the literature.

## CASE REPORT

A 34-year old multiparous housewife(Korean) was admitted to our hospital with a 1-day history of vomiting and left lower abdominal pain on November 3, 1993. She had had several similar episodes over the previous 10 years, but had had no specific treatment because of spontaneous symptomatic relief. Two months prior to this admission, she had been briefly admitted because of lower abdominal pain and diarrhea and diagnosed as having acute gastroenteritis. The symptoms were relieved with conservative management only. She was born in Korea and has never been abroad. She had no contributory past history, such as beta-adrenergic antagonist use, hepatic diseases, tuberculosis, or operations. Family history also was not remarkable.

On admission, she had an acutely ill-looking appearance. Blood pressure was 110/70 mmHg, pulse rate 90/min, body temperature 36.5℃, and respiratory rate 20/min. The conjunctivae were not pale, and the sclerae were not icteric. The examination of the lungs and heart was not remarkable. The abdomen was soft, but slightly distended. Bowel sound was increased in pitch and frequency. On palpation, there was direct tenderness on the epigastrium and left lower quadrant abdomen. A 4X4 cm sized, tender mass was palpated on the left lower quadrant abdomen. There were neither ascites nor hepatosplenomegaly.

On laboratory examinations, hemoglobin was 13.8 g/dl, hematocrit 41.7%, leukocyte 10,700/ $mm^3$ (neutrophil 91%, lymphocyte 7%, monocyte 2%), and platelet 289,000/$mm^3$. Blood chemistry tests showed calcium 9.9mg/dl, inorganic phosphorous 2.1mg/dl, total protein 7.5g/dl, albumin 4.5g/dl, aspartate transaminase 40.9 IU/L and alanine transaminase 30.0 IU/L. Urinalysis and other biochemical tests were all within normal ranges. Also chest X-ray and electrocardiogram were unremarkable.

A plain film of the abdomen showed an intermittently dilated small bowel. An abdominal ultrasonography showed a large echogenic mass associated with a small amount of ascites in the left lower abdomen(Fig. 1). Also a 4 cm-sized cystic lesion was found in the right adnexa. Computed tomography of the abdomen performed on the same day demonstrated the clustered gas-containing small bowel loop and dilated proximal small bowel with air-fluid levels due to intestinal obstruction. A small amount of ascites was seen in the left inguinal fossa(Fig. 2). A small bowel follow-through performed on the fourth day of admission revealed that the ileal loops were bunched and confined in the lower abdomen and pelvic cavity and giving rise to extrinsic mass effect on adjacent small bowel loops(Fig. 3).

Although the symptoms and signs of intestinal

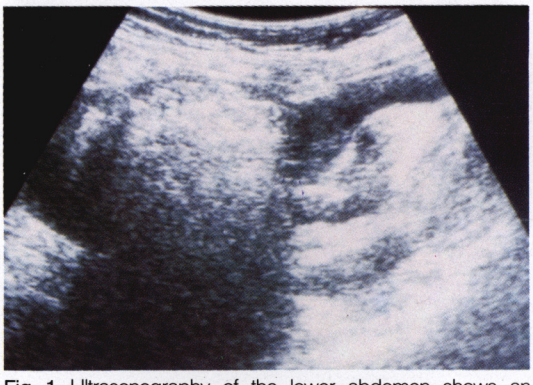

Fig. 1. Ultrasonography of the lower abdomen shows an echogenic mass with posterior shadowing.

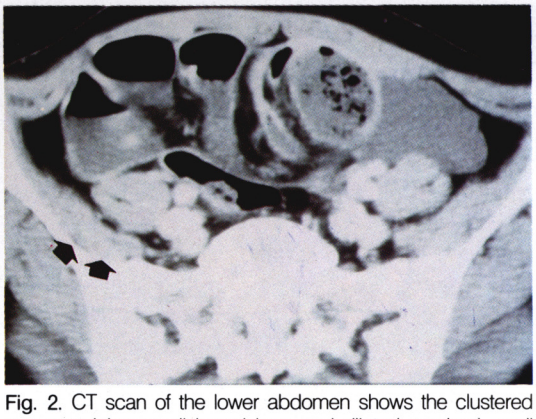

Fig. 2. CT scan of the lower abdomen shows the clustered gas-containing small bowel loop and dilated proximal small bowel with air-fluid levels due to intestinal obstruction. A small amount of ascites is seen in the left inguinal fossa(arrow).

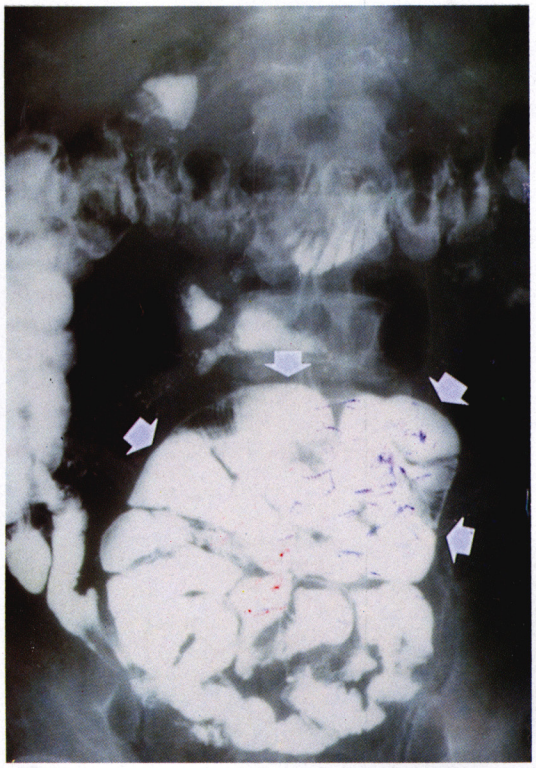

Fig. 3. Barium-follow-through shows that the ileal loops are bunched and confined in the lower abdomen and pelvic cavity and giving rise to extrinsic mass effect on the adjacent small bowel loop(arrow).

obstruction were abated with conservative treatment, an exploratory laparotomy was performed on the 7th hospital day to define and correct the underlying small bowel pathology. At operation, it was found that the considerable length of the distal small bowel, 30cm off the ileocecal valve, was encased in a whitish thickened membrane(Fig. 4). The sigmoid colon was displaced to the left. The greater omentum looked hypoplastic and was also encased in a fibrous tissue. The appendix adhered to the cecal wall without gross inflammation. A 5X4 cm cystic mass was found on the right ovary. Partial resection of the small intestine including the membrane and end-to-end anastomosis of the small bowel were performed. Also a right salpingo-oophorectomy and incidental appendectomy were performed. On incising the fibrous sac after resection, normal-looking small bowel loops were freed. Total length of the resected small bowel was about 1.2 meters. Some adhesions between the serosal layer and the fibrous sac were

Fig. 4. A photograph taken during the operation shows a white colored sac-like structure containing the part of the small bowel, adjacent to the ileocecal valve.

noted. The histology of the membrane revealed only fibrosis without inflammation. The histologic diagnosis of the resected ovary was follicular cyst. After the operation, she suffered from bowel frequency and loose stool for about 6 months, which were controllable with medication. Now, 15 months after the operation, she is in good health.

## DISCUSSION

Abdominal cocoon was first described and named by Foo et al.(1978). They reported 10 cases of adolescent females presenting with acute or subacute small bowel obstruction with or without a mass. Intestinal obstructions in these cases were caused by encasement of the small intestine with fibrous membrane, and they named it abdominal cocoon. Since their first report, 14 more cases have appeared in the English literature under the term of "abdominal cocoon".

Table 1 shows the summary of the age, sex, patients' race, authors' country, and attributable etiologies or associated diseases of the reported cases. Cases from No. 1 to No. 21 are abdominal cocoon of unknown etiology and cases from No. 22 to No. 24 are abdominal cocoon with attributable causes or associated diseases.

As shown, abdominal cocoon of unknown etiology primarily affected the female gender with one exception(Case No. 16) and the affected females were all adolescents except one case(Case No. 12). All the cases were born and/or resided in the tropical or

**Table 1.** Summary of age, sex, patients' race, authors' country and etiologies of the so-far reported cases.

| case | age | sex | reference | year | race | country | etiology |
|---|---|---|---|---|---|---|---|
| 1 | 14 | F | Foo et al. | 1978 | Chinese | Singapore | Unknown |
| 2 | 14 | F | 〃 | 〃 | Chinese | Singapore | 〃 |
| 3 | 15 | F | 〃 | 〃 | Chinese | Singapore | 〃 |
| 4 | 16 | F | 〃 | 〃 | Chinese | Singapore | 〃 |
| 5 | 17 | F | 〃 | 〃 | Chinese | Singapore | 〃 |
| 6 | 15 | F | 〃 | 〃 | Chinese | Singapore | 〃 |
| 7 | 16 | F | 〃 | 〃 | Chinese | Singapore | 〃 |
| 8 | 14 | F | 〃 | 〃 | Chinese | Singapore | 〃 |
| 9 | 13 | F | 〃 | 〃 | Malaysian | Singapore | 〃 |
| 10 | 18 | F | 〃 | 〃 | Malaysian | Singapore | 〃 |
| 11 | 12 | F | Sayfan et al. | 1979 | Caucacian | Israel | 〃 |
| 12 | 4 | F | Rao et al. | 1979 | Indian | India | 〃 |
| 13 | 17 | F | Marinho and Adelusi | 1980 | Neigerian | Neigeria | 〃 |
| 14 | 14 | F | Sieck et al. | 1983 | Arab | Saudi-Arabia | 〃 |
| 15 | 15 | F | Macklin et al. | 1991 | Pakistani | U.K | 〃 |
| 16 | 43 | M | Yip and Lee | 1992 | Chinese | Malaysia | 〃 |
| 17 | 13 | F | 〃 | 〃 | Chinese | Malaysia | 〃 |
| 18 | 14 | F | 〃 | 〃 | Chinese | Malaysia | 〃 |
| 19 | 15 | F | 〃 | 〃 | Indian | Malaysia | 〃 |
| 20 | 15 | F | 〃 | 〃 | Indian | Malaysia | 〃 |
| 21 | 15 | F | McFarlane | 1993 | African | Kenya | 〃 |
| 22 | 60 | M | Cambria and Shamberger | 1984 | Unknown | U.S.A | Liver cirrhosis LeVeen shunt |
| 23 | 43 | F | Seng et al. | 1993 | Malaysian | Malaysia | Beta-antagonist Constrictive pericarditis |
| 24 | 17 | M | 〃 | 〃 | Malaysian | Malaysia | Liver cirrhosis |

subtropical areas. Our present case was also a female patient, but could be distinguished because this case was rather older than the previously reported cases and was born and resided in Korea. Foo et al.(1978) hypothesized that the pathogenesis of abdominal cocoon might be due to retrograde menstration with superimposed subclinical primary peritonitis in that their 10 cases were all adolescent females usually within 2 years of their menarche. However, this hypothesis was refuted because of the development of abdominal cocoons in two premenarchal females(Rao et al., 1979 ; Sieck et al. 1983) and one male(Yip and Lee, 1992). Because of the peculiar geographical distribution of abdominal cocoon of unknown etiology, peritonitis by endemic microorganisms or by as yet unidentified environmental factors was proposed to be implicated in the pathogenesis of abdominal cocoon(Yip and Lee, 1992). Our case presented herein, however, might broaden the spectrum of microorganisms or environmenatal factors, which should not be limited to tropical or subtropical areas. The possibility of a congenital origin(Marinho and Adelusi, 1980) should be taken into account in that this condition can develop in a broad age range, in both sexes, and without geographical zone limitation. Also occasional association with the absence of the greater omentum(Macklin et al., 1991 ; Yip and Lee, 1992) or hypoplastic greater omentum as in our case might support the congenital origin of abdominal cocoon.

The fibrosis in abdominal cocoon resembles that seen in sclerosing peritonitis due to long-term administration of beta-adrenergic blockers such as practolol or propranolol(Brown et al., 1974 ; Windsor et al., 1975 ; Eltringham et al., 1977 ; Harty, 1978 ; Ahmad, 1981). Seng et al.(1993) reported an abdominal cocoon in a 43-year-old woman who had a history of taking propranolol and constrictive pericarditis(Case No. 23). It must be settled as to whether the term "sclerosing peritonitis" through the use of beta-adrenergic blockers is exchangeable with abdominal cocoon or not. If the original concept of abdominal cocoon advocated by Foo et al.(1978) is respected, it may be reasonable that sclerosing peritonitis through

the use of beta-adrenergic blockers is a distinct clinical entity. The mechanism by which beta-adrenergic blockers can induce fibrosis is well described elsewhere(Seng et al., 1993). Cambria and Shamberger(1984) described a case of abdominal cocoon possibly associated with the LeVeen shunt. Also 5 similar cases have been reported in abstract form(Greenlee et al., 1979). These cases support the strong association between the LeVeen shunt for cirrhotic ascites and the abdominal cocoon-like syndrome. Seng et al.(1993) also reported a case of abdominal cocoon in a 17-year-old male with liver cirrhosis(Case No. 24). Although there is evidence that the visceral peritoneum in patients with decompensated cirrhosis with ascites is abnormally thickened(Cambria and Shamberger, 1984), this patient had neither ascites nor a history of LeVeen shunt. Therefore this case seems to be a mere coincidence of abdominal cocoon and liver cirrhosis. However, it is yet to be seen whether liver cirrhosis itself is associated with abdominal cocoon or not.

Preoperative diagnosis of abdominal cocoon is difficult. Yip and Lee(1992) listed 4 main clinical features that help to suspect abdominal cocoon preoperatively. Those are as follows ; 1) a relatively young girl without an obvious cause of intestinal obstruction, 2) a past history of similar episodes which resolved spontaneously, 3) presenting with abdominal pain and vomiting but rarely the four cardinal symptoms of intestinal obstruction, and 4) the presence of a non-tender soft mass on abdominal palpation. The present case almost fits all the above 4 clinical features except for the patient's age.

The radiological findings do not seem to give specific clues to the diagnosis of abdominal cocoon preoperatively. As in our present case, however, small bowel follow-through occasionally brought about the characteristic serpentine configuration of the small bowel loop within the cocoon(Sieck et al., 1983 ; Yip and Lee, 1992).

Definitive diagnosis is made at laparotomy. Characteristically, the small bowel is found totally or partially coiled up in a concertina-like fashion encased in a dense white membrane(Sieck et al., 1983). In occasional cases, this membrane has extended to the surrounding organs especially the large bowel. When the membrane is incised, the dilated loops of bowel are easily freed and the membrane can be removed. Therefore, lysis of the membrane and adhesions is the treatment of choice for abdominal cocoon. However, if surgeons do not appreciate this condition well, unnecessary bowel resection is likely to be performed as in our case. Also two reported cases underwent some form of bowel resection(Foo et al., 1978 ; Sayfan et al., 1979 ; McFarlane, 1993). The case of Sayfan et al.(1979) was erroneously reported as having peritoneal encapsulation even after the operation. Thus abdominal cocoon should be differentiated with peritoneal encapsulation. Peritoneal encapsulation is thought to be a congenital condition resulting from an abnormal embryological development(Thorlakson et al., 1953) and is characterized by an accessory peritoneal sac derived from the peritoneum of the yolk sac as it is withdrawn rapidly into the abdominal cavity during the 12th week of gestation. It has two openings, one where the small bowel enters at the duodenojejunal junction and the other where the terminal ileum leaves. Contrary to abdominal cocoon, peritoneal encapsulation is largely asymptomatic and the diagnosis is made incidentally and late in life(Lewin and McCarthy, 1970 ; Sayfan et al., 1979 ; Sieck et al., 1983).

In conclusion, the addition of our present case to the so-far reported cases should indicate that abdominal cocoon of unknown etiology is not limited to tropical or subtropical zones. This rare condition, therefore, should be considered one of the possible causes of intestinal obstruction in areas other than tropical or subtropical zones.

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