Malaysian Journal of Medical Sciences, Vol. 11, No. 1, January 2004 (81-85)

CASE REPORT

ABDOMINAL COCOON IN ASSOCIATION WITH ADENOMYOSIS AND LEIOMYOMATA OF THE UTERUS AND ENDOMETRIOTIC CYST : UNUSUAL PRESENTATION

Nor Haznita Mohd. Noor, Nik Mohamed Zaki, Gurjeet Kaur*, Venkatesh R. Naik*, Ahmad Zahari Zakaria**

Department of Obstetric and Gynaecology, Department of Pathology*, Department of Surgery**, School of Medical Sciences, Universiti Sains Malaysia, Health Campus 16150 Kubang Kerian, Kelantan, Malaysia**

Abdominal cocoon or sclerosing encapsulating peritonitis is a rare condition. A 46 year old Malay woman with adenomyosis and leiomyomata of the uterus and ovarian endometriotic cyst in association with abdominal cocoon is reported.

Key words *: suppression, reaction formation, persecution, delusion*

Submitted-26.12.2002, Accepted-31.12.2003

Introduction

The abdominal cocoon, or sclerosing encapsulating peritonitis (SEP) was first described and named by Foo et al in 1978 (7). It refers to a rare condition in which there is a total or partial encasement of the small bowel by a fibrinocollagenous cocoon-like sac. The aetiology of the disease remains obscure (21). It can be categorized into primary or secondary (4). In primary or idiopathic SEP, those who were affected were mainly young women in perimenarchal age from tropical and subtropical regions particularly from Asia and Africa (15). It rarely occurs in male (4,7,16). Several conditions are known to be associated with secondary SEP as shown in Table 1.

We report a rare finding of a woman who presented clinically as a malignant ovarian tumour but was later confirmed to have endometriosis, adenomyosis and leiomyomata in association with abdominal cocoon.

Table 1 *: Secondary causes of SEP*

Treatment with protocol
Chronic ambulatory peritoneal dialysis
Ventriculoperitoneal shunting
Sarcoidosis
Mycobacterial infection
Systemic Lupus Erythematosus
Luteinized thecoma of ovary

Case History

A 46 year old unmarried Malay woman presented with an abdominal mass of 5 to 6 years duration which had been increasing in size over the past 3 months. She did not complain of abdominal pain or change in her bowel or urinary habits. She experienced shortness of breath especially on lying flat. During that period she also had loss of appetite and weight. There was no leg swelling or pain.

Menstrual history

She attained menarche at the age of 15 years and had regular 28 to 30 days cycle with 6 to7 days flow. It was associated with mild dysmenorrhoea. There was no history of menorrhagia or intermenstrual bleed. She was not sexually active and never had a Pap smear done before.

She had no significant past gynaecological, surgical or medical history. There was no family history of breast, ovarian, endometrial or bowel cancer.

Examination revealed a pale but comfortable woman. Her blood pressure was noted to be high in the ward and she was started on Atenolol. Cardiorespiratory systems were unremarkable. Further examination revealed a soft, non tender grossly distended abdomen. Afirm mobile mass was

felt at the lower abdomen, floating in ascites. Margin and surface of the mass was not well appreciated. It was possible to get below the mass. Shifting dullness and fluid thrill were present.

Chest X-ray was normal. Trans abdominal ultrasound revealed a huge multiloculated cystic mass (18 x 16cm) with solid area. There was no papillary projection. Gross ascites was present. The liver and both kidneys were normal.

Serum for tumour markers showed only raised Ca 125 level (349.6 iu/ml) while other blood tests were within normal limits.

At laparotomy, 10 litres of yellow coloured ascitic fluid was drained. The small bowel appeared to be wrapped up to the upper part of the abdomen with a thick peritoneal covering (abdominal cocoon)(Figure 1). A huge cystic tumour mass (16 X 18 cm) was found with the capsule intact which ruptured during mobilization. It contained chocolate material. This tumour mass was stuck together with the encapsulated sac over the whole pelvis. Both tubes and normal ovarian tissues were not identifiable as they were enclosed within the tumour mass. The mass was adherent to the rectum, uterus, bladder and posterior abdominal wall. A band of omentum was also stuck to the mass. When the encapsulated fibrocartilagenous mass was opened, the uterus was found to be enlarged and the bladder was adhered to anterior surface of uterus. Both ureters were identified and preserved. Cytoreductive procedure was performed.

Post operative period was uneventful. She was discharged well and came back to the clinic for follow up four weeks later looking very healthy and the serum CA 125 was normal (<35 iu/ml).

Pathological Examination:

Gross examination:

Specimen consisted of uterus without cervix which was partially cut open. It measured 130 x 120 x 70 mm, and irregular cystic structure, probably ovary, was attached to it. Cut section through the uterus showed multiple intramural and subserosal fibroids measuring 20 to 50 mm in diameter. They showed typical whorled appearance and areas of trabeculation and spots of haemorrhage. The ovarian cyst wall was greyish brown measuring 7 mm in thickness. The inner surface was smooth and covered with altered blood. The cyst was filled with altered blood. No solid structure were seen.

Microscopic examination:

Section of the endometrium showed compact stroma and proliferative type of glands. Deep in the myometrium were numerous foci of endometrial glands and stroma. The fibroid showed broad interlacing bundles of smooth muscle fibres with extensive hyalinisation, no mitotic figures were seen. Section from the ovarian cyst wall showed a dense fibrocollagenous tissue (Figure 2). The inner surface was lined by granulation tissue, haemorrhage and numerous hemosiderin laden macrophages. In one foci ossification was also seen. No endometrial glands were present in the wall of the cyst.

The peritoneal fluid cytology revealed no malignant cells and a diagnosis of adeno mycosis and leiomyomata of the uterus with endometriotic cyst was made.

Figure 1 *: Intraoperative finding of abdominal cocoon*

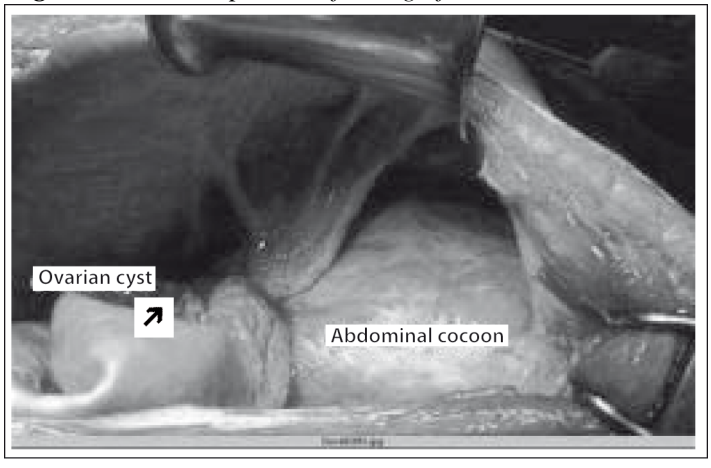
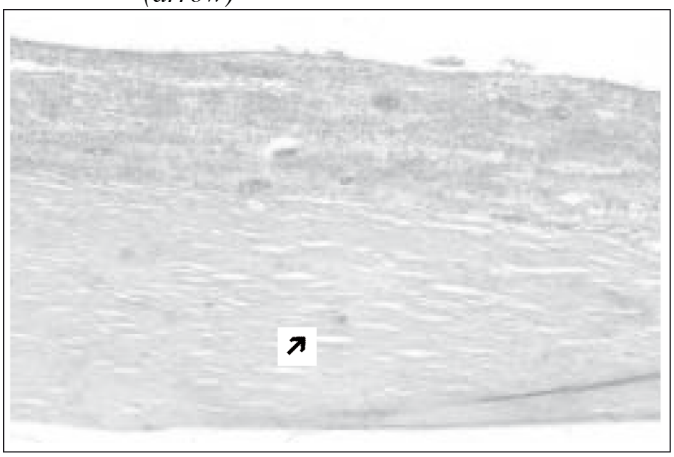


Figure 2 : *Cyst wall composed of fibrocollagenous tissue (arrow)*



Discussion

Sclerosing encapsulating peritonitis or abdominal cocoon is a rare entity and its aetiology remains unknown. The main pathogenesis in this condition appears to be peritonitis leading to sclerosis with membrane formation (22). In primary or idiopathic SEP the stimulus for this inflammatory reaction is open to speculation. Some suggest that it could be due to subclinical viral peritonitis which ascends vaginally with transfallopian migration or through haematogenous spread (16,24). Others postulate that retrograde menstruation may be the initial inflammatory stimulus(16). The argument against this is that the condition also afflicts premenarchal girls and males. In some patients, congenital absence of greater omentum is postulated to be the predisposing factor (12,24).

In secondary SEP reported in association with practolol intake, reduced adenosine 3',5'- cyclic monophosphate : guanosine 3'5'- cyclic monophosphate ratio has been shown to initiate excessive cellular proliferation (1). In others, presence of infective organisms, foreign body or autoimmune reaction may explain the origin of inflammatory reaction (7-9,11,18).

Clinically, it is rare cause of acute or subacute intestinal obstruction and was reported to be present in 81% of reported cases of abdominal cocoon (10,21). While palpable bowel mass may be present in 63% of them, more often than not the correct diagnosis is not made pre operatively (2). Laparotomies were often performed with other diagnosis in mind such as mesenteric cyst, ovarian cyst, appendicitis, intestinal polyp, intussusception or bowel obstruction of unknown cause (2,6,7,12-14,17,19,20-24). In our patient, the predominant presenting features were of malignant ovarian tumour, thus abdominal cocoon which was a rare association with gynaecologic condition was not even considered in the clinical work up pre operatively.

The membrane encasing the bowel loops consist predominantly of a proliferation of fibroblasts resulting in diffuse fibrous thickening without any or only mild inflammation (24). Small intestine is predominantly involved but the length may be variable. In our patient the whole length of small bowel, most of large bowel, liver, gall bladder, stomach and spleen was encased within the cocoon and all wrapped up to the upper part of the abdomen.

In 1994, P.B. Clement et al (5) reported six cases of sclerosing peritonitis associated with luteinized thecoma. There seems to be a resemblance of the clinical feature in four of their cases with our patient where by the dominating feature is the presence of the ovarian tumour. Furthermore, all the cases in their series had ascites and in some of them the ascitic fluid was voluminous as in our patient. It seems that the only pathology in gynaecology that has been associated with abdominal cocoon is luteinized thecoma of ovary (5). However, only the finding of sclerosing peritonitis and not abdominal cocoon with encapsulating features was specifically described in their cases.

In contrast, the pathological diagnosis in our patient was adenomyosis and leiomyomata of the uterus and ovarian endometriotic cyst. So far there is no reported association between abdominal cocoon with these conditions. Both endometriosis and primary SEP share similar postulated pathogenesis through retrograde menstruation. However, it cannot be the sole pathogenic mechanism for abdominal cocoon formation as

many others who suffer from varying degree of endometriosis do not have this complication. Voluminous ascites in association with extensive peritoneal endometriosis is extremely rare (25). In 70% of cases it affects young nulliparous black women. However, the presence of abdominal cocoon was not mentioned in any of the cases.

It was possible that this patient may be having several unrelated pathology at one time, which were primary SEP, adenomyosis and leiomyomata of the uterus and endometriotic cyst. However it was unlikely as the initial clinical manifestation of primary SEP is usually of small bowel obstruction. Furthermore, ascites was not reported in any case of primary SEP. Although adenomyosis and leiomyomata of the uterus and endometriotic cyst may be asymptomatic, it was surprising that she remained without symptom with primary SEP as many present perimenarchal.

Treatment of abdominal cocoon reported in the literature consisted mainly of operative removal of thickened peritoneum and adhesiolysis. Bowel resection is deemed necessary if nonviable and it is associated with increased morbidity and mortality. As our patient did not have any bowel symptoms, only cytoreductive surgery for ovarian tumour was performed without operating on the abdominal cocoon. Follow up data suggests that sclerosing peritonitis associated with lutienized thecoma is self limiting (5). Danazol has been shown to be effective in patients with ascites and peritoneal endometriosis (25). However the duration of treatment and potential of fertility are presently undetermined.

Management of such a rare case present a dilemma as it is impossible to decide on the best therapeutic approach on table as most gynaecologists would never see such a case.

What intrigued the authors, including the pathologist involved, was the presence of two rare entities in the same patient, which are the voluminous ascitic fluid associated endometriosis, a benign gynaecological disease and the abdominal cocoon.

Correspondence :

Assoc. Prof. Dr. Nik Mohamad Zaki, MD (UKM), Mmed O & G (USM), Fellow Gynae. Oncology
New Castle University
Department of Obstetric & Gynaecology,
School of Medical Sciences,
Universiti Sains Malaysia, Health Campus,
16150 Kubang Kerian, Kelantan, Malaysia

References

1. Ahmad S. Sclerosing peritonitis and propanolol. *Chest* 1981; **79**:361-2.
2. Attar ZB, Nazir Z, Moazam F. The abdominal cocoon: an unusual cause of intestinal obstruction in adolescents. *Ann Trop Paediatr* 1993; **13**:99-102.
3. Brown P, Baddeley H, Read AE, Davis JD, McGarry J. Sclerosing peritonitis, an unusual reaction to a beta-adrenergic blocking drug (practolol). *Lancet* 1974; **21**:1447-81.
4. Burstein M, Galun E, Ben-Chetrit E. Idiopathic sclerosing peritonitis in a man. *J Clin Gastroenterol* 1990; **12**:698-701.
5. Clement PB, Young RH, Hanna W, Sculley RE. Sclerosing peritonitis associated with luteinized thecomas of the ovary: a clinicopathological analysis of 6 cases. *Am J Surg Pathol* 1994; **18**:1-13.
6. De Wind CM. Unusual cause of small bowel obstruction in adolescent girls: the abdominal cocoon. *J R Coll Surg Edinb* 1992; **37**:60.
7. Foo TK, Ng KC, Rauff A, Foong WC, Sinniah R. Unusual small intestinal obstruction in adolescent girls: the abdominal cocoon. *Br J Surg* 1979; **65**:427-30.
8. Holland P. Sclerosing encapsulating peritonitis in chronic ambulatory peritoneal dialysis. *Clin Radiol* 1990; **41**:19-23.
9. Kaklamanis P, Vayopoulos G. Chronic lupus peritonitis with ascites. *Ann Rheum Dis* 1991; **50**:176-7.
10. Kobayashi TK, Ueda M, Nishino T, Nishida S, Yeneyama C, Watanabe S. Appearance of 'collagen balls' in ascitic fluid cytology with abdominal cocoon (encapsulating peritonitis) [letter]. *Diagn Cytopathol* 1997; **16**:469-70.
11. LaFerla G, McColl KEL, Crean GP. CSF induced sclerosing peritonitis: a new entity? *Br J Surg* 1986; **73**:7.
12. Macklin J, Hall C, Feldman MA. Unusual cause of small bowel obstruction in adolescent girls: the abdominal cocoon. *J R Coll Surg Edinb* 1991; **36**:50-2.
13. Marinho A, Adelusi B. The abdominal cocoon : case report. *Br J Obstet Gynaecol* 1980; **87**:249-50.
14. McFariane GA. The abdominal cocoon. *Trop Doct* 1993; **23**:134-5.
15. McLaughlin K, Butt G, Madi A, Mc Millan M, Mactier R. Sclerosing peritonitis occurring in a haemodialysis patient. *Am J Kidney Dis* 1996; **27**:729-32.
16. Narayanan R, Kabra SG, Bhargava BN, Sangal BC. Idiopathic sclerosing encapsulating peritonitis. *Lancet* 1989; **2**:127-9.
17. Navani S, Shah P, Pandya S, Doctor N. Abdominal cocoon – The cauliflower sign on barium small bowel series. *Indian J Gastroenterol* 1995; **14**:19.

18. Ngo Y, Messing B, Marteau P, Nouel O, Pasquiou A, Lavergne A, et al. Peritoneal sarcoidosis: an unrecognised cause of sclerosing peritonitis. *Dig Dis Sci* 1992; **37**:1776-80.

19. Rao PLNG, Mitra SK, Pathak IC. Abdominal cocoon – a cause of intestinal obstruction in a 4 year old girl. *Indian Pediatr* 1979; **16**:1047-8.

20. Sayfran J, Adam YG, Relf R. Peritoneal encapsulation in childhood. *Am J Surg* 1979; **138**:725-8.

21. Seng LK, Mahadaven M, Musa A. Abdominal cocoon: a report of two cases. *Br J Surg* 1993; **80**:1149

22. Sieck JO, Cowgill R, Larkworthy W. Peritoneal encapsulation and abdominal cocoon. Case reports and a review of the literature. *Gastroenterology* 1983; **84**:1579-601.

23. Tsunoda T, Mochinaga N, Eto T, Furul J, Tomioka T, Takahara H. Sclerosing encapsulating peritonitis combined with peritoneal encapsulation. *Arch Surg* 1993; **128**:353-5.

24. Yip FWK, Lee SH. The abdominal cocoon. *Aust NZ J Surg* 1992; **62**:638-42.

25. Halme J, Chafe W, Currie JL. Endometriosis with massive ascites. *Obstet Gynecol*