PSEUDOMYXOMA PERITONEI

(A Report of three cases)

by

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The characteristics of typical pseudomyxoma peritonei are still not well established. Its usual relationship to pseudomucinous cystadenoma, rarely to mucocele of the appendix, the widespread distribution of the gelatinous material throughout the peritoneal cavity, the tendency to recurrence, the unpredictable course to a fatal termination—a course sometimes rapid, sometimes very slow and, even in frankly malignant cases, the long restriction of the lesions to the peritoneal cavity is still not clearly understood. Certain aspects of its aetiology and behaviour still call for discussion, in particular whether it is at all benign pseudomucinous cyst of the ovary, and finally the importance of the massive peritoneal and parietal reaction so often present.

Malpas (1959) has suggested that pseudomyxoma peritonei of ovarian origin is always a malignant condition and that the common benign pseudomucinous cyst is not causally related. Shanks' (1961) experience of the disease has not been in accordance with the observations of Malpas. These diametrically opposite views and opinions have prompted us to present 3 cases of pseudomyxoma peritonei recently treated in our hospital.

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CASE REPORT:

Case 1. Mrs. A. M., aged 19 years, Para 2 + 0 was admitted at Eden Hospital on 10.10.77 with the complaints of gradual swelling of abdomen and pain since her last child birth (i.e. 2 months back); anorexia, loss of weight and irregular fever—same duration.

Menstrual History: Menarche—12 years. Cycles regular—28 ± 2 days duration of flow—3 to 5 days. L.M.P.—no menstruation since last child birth.

Obstetric History: Two term normal deliveries at home; both boys; alive and well. She did not attend any Antenatal Clinic during her last pregnancy.

There was no significant past history.

Evaluation on Admission: Cachectic with low general condition. There was moderate anaemia. No cervical lymphadenopathy. Blood pressure—100/70 mm of Hg. no oedema.

Cardio-vascular system showed no abnormality.

Abdominal Examination: A huge mass occupied right iliac, umbilical hypogastric and right hypochondriac regions. Margins were well defined with slight irregularity except the lower one. It was not tender partly solid and partly cystic with mobiliy from side to side. There was moderate amount of free fluid in the peritoneal cavity.

Vaginal Examination: The vulva and vagina were normal. The uterus was retroverted,

normal in size and mobile. The lower margin of the mass felt per abdomen, was felt high up in the pelvis through right fornix. The left fornix and pouch of Douglas were clear.

The provisional diagnosis was malignant ovarian tumour with ascites.

Investigations:

Haemoglobin: 9.5 gm.%. Total W.B.C. count —7,500 cells per c.c. Poly—74%, lympho—24%, mono—2%, eosino—0% E.S.R.—28 mm/st hour. Fasting blood sugar—78 mg.%. Urea—31 mgm%. Stool—no abnormality detected.

Straight X-ray of abdomen and chest: no abnormality detected.

Laparotomy was done on 19.11.77 by right paramedian incision (7 inches long). A large thick walled cyst arising from right ovary apparently non-malignant occupied almost whole of abdomen. Yellowish white gelatinous material filled the peritoneal cavity due to spontaneous leakage from the cyst. There was a ping-pong ball sized cyst in the left ovary. Right ovariotomy and left sided ovarian resection was done. While toileting, many small cysts were found adherent with the omentum, in the liver bed, behind the stomach and also in pouch of Douglas. These were removed as much as possible. The appendix was normal. The amount of gelatinous material was 3 litres. The patient received 2 bottles of blood after operation. The postoperative period was uneventful. She was discharged on 28.11.77.

The histological picture of the cysts from both ovaries showed the picture of pseudomucinous cyst adenoma without any malignant change.

The patient turned up for follow-up on 2.1.78 and her condition was satisfactory.

Case 2: Mr. R. P.; aged 46 years; para 2 + 0, was admitted on 6.7.77 at Eden Hospital with complaints of swelling of abdomen for 1 year and oliguria, insomnia, anorexia etc. for 1 month.

Past History: She had panhysterectomy for bilateral ovarian cyst in 1973 and laparotomy for pseudomyxoma peritoneal in 1976 with extensive adhesions followed by course of Telecobabt therapy.

Menstrual History: Surgical menopause since 1973.

Obstetric History: Two full term normal deliveries—one boy and one girl, alive and well. Last childbirth about 10 years ago.

Evaluation on Admission: The patient was moderately anaemic and cachectic. Blood pressure—110/75 mm. of Hg. No abnormality was detected in cardio-vascular or respiratory system.

Abdominal Examination: There was severe type ascites.

Vaginal Examination: There was pressure—prolapse of vault with moderate cystocele and mild rectocele. No definite mass was felt on vaginal examination.

Investigation:

On 13.7.77: Haemoglobin—8 gm%. Total W.B.C. count—9,000 cells per c.c. Poly—70%; lympho—25% mono—2%, eosino—3%. Fasting blood sugar—85 mg%. Blood urea—35 mgm.%.

Straight X'ray of the chest showed pleural thickening on left side.

Examination of peritoneal fluid did not show malignant cells.

Sugar—20 mg.%; Cloride—660 mg.% Protein 2.4 gm%.

The patient was treated with intravenous Endoxan therapy 200 mg. daily for 7 days and repeated paracentesis. About 500 ml. of haemorrhagic pseudomucinous material was drained on 9-7-77 and 11-7-77; Symptomatic treatments for bed sore, oliguria, constipation, dyspnoea etc. were done; Radiotherapy was not advocated. Ultimately the patient expired on 23-7-77.

Case 3: Mrs. K.D., aged 40 years, was admitted in the medical ward in June, 1967 with abdominal swelling, irregular fever and amenorrhoea for a few months (Mazumdar and Mondal, 1967). On examination, the feel of the abdomen was cystic with presence of fluid thrill. A lump was also palpable in the lower quadrant of the abdomen. There was no other abnormality either in the systemic or laboratory examination except a moderately high E.S.R.

The patient was put on antitubercular therapy. On diagnostic puncture with a thick bore needle, gelatinous material came out from abdomen which suggested some other pathology rather than tuberculosis. Laparotomy was performed by which 10 kg. of pseudomucinous materil was drained out. Right ovary showed polycystic tumour and signs of old rupture. Total hysterectomy with salpingo-oophorectomy was carried out. The material collected was pseudomucin as it did not precipitate on addition of Acetic acid.

Discussion

Aetiology: Originally pseudomyxoma peritonei was believed by Virchow (1884) to be a primary peritoneal lesion. He described 1 case as a myxomatous degeneration of the peritoneum or a chronic myxomatous peritonitis. Spencer Wells (1865) in England and Pean (1880) in France, failed to make distinction between pseudomyxoma and colloid carconomatosis of the peritoneum. Schroder (1874) described it as "infectiousness" for the peritoneum following perforation of "cystoma myxomatosa" and coined the nomenclature "Peritonitis myxomatosa". Wilson (1912) subsequently established pseudomyxoma peritonei as a disease entirely associated with ruptured pseudomucinous cystadenoma. He did not find a single malignant case of pseudomyxoma peritonei in a personal series of 331 ovarian tumours. Cariker and Dockerty (1954) on the other hand recorded pseudomyxoma peritonei as a complication of 44 tumours all of which were cystadenocarcinomas. Shanks' (1961) experience has been quite the reverse, with pseudomyxoma presenting on 2 occasions only as a complication of pseudomucinous cystodenocarcinoma, an incidence of malignancy of only 17 per cent. Malpas (1959), however, has strongly proposed that most, and perhaps all, cases of pseudomyxoma peritonei associated with ovarian tumours are malignant in the long run.

Pseudomyxoma Peritonei of Appendicular Origin: Pseudomyxoma peritonei of appendicular origin is a rare disease. Eliot (1957) reported only 2 cases of pseudomyxoma peritonei out of 30 mucocele of the appendix developing in 13,000 appendicectomies—an incidence of 0.2 per cent. There is close relationship between mucinous cystic epithelium with the endodermic intestinal epithelium, as emphasised by those who accept the teratomatous etiology of mucinous cyst. Whatever the reason there is little doubt that the appendicular form of the disease has a better prognosis than the ovarian (Masson and Hamrick, 1930).

Clinical Behaviour: Big pseudomucinous cyst may rupture spontaneously or their contents may escape into the peritoneal cavity as a result of tapping, or from a traumatic rupture or from an accidental spill at operation, may determine a foreign-body type of peritoneal reaction resulting in intra-abdominal transformation of the peritoneal mesothelium to a mucin secreting epithelium. This altered peritoneal mesothelium continues its mucus secretion, with the gradual accumulation in the peritoneal cavity of large amount of gelatious material, the so called pseudomyxoma peritonei. Evacuation of this material at operation is almost invariably followed by reaccumulation because of the impossibility of altering the secretion of the mucinous mesothelium.

Among 13 cases of 'pseudomyxoma', Shanks (1961) found 10 cases originating from benign and 2 in malignant ovarian tumours and one arising from an appendicular mucocele. Of 7 survivors, 2 had recurrences. From this study Shanks (1961) concluded that pseudomyxoma peritonei was neither invariably associated with malignant disease of the ovary, nor is it always fatal. In Mayo clinic series Masson and Hamrick (1930) found only 43 per cent incidence of malignancy in the 30 cases recorded upto that time. Malpas (1959) on the other hand is of the opinion that most and perhaps all cases of pseudomyxoma peritonei prove to be malignant in the long run. But the question still remains—if pseudomyxoma peritonei is always a malignant

condition, how so many cases survive for prolonged period even when primary lesion appears to be quite active?

In Case 1 the pathology was of ovarian origin where spontaneous leakage might possibly have taken place during the stress and strain of preceding labour. The treatment undertaken with conservation of a portion of the left ovary along with the uterus was justifiably decided in this case considering the age of the patient and apparent non-malignant nature of the cysts. The benign nature of the cysts was further substantiated by histopathological examinations which did not indicate any further treatment as well. It is controversial whether the mere presence of pseudomucinous material in the peritoneal cavity in this type of case require further management at the present state. However, the importance of close follow-up cannot be ignored, which is also undertaken here to detect the early signs, if any, of the possible recurrence. The patient did turn up on 2-1-78 when her condition was found to be satisfactory.

The second case was an interesting case of recurrence and posed a therapeutic problem where originally—pelvic clearance was done considering the age of the patient and to minimise the chance of future malignancy. However, the case presented again with pseudomyxoma peritonei after 2 years when surgery followed by anti-cancer chemotherapy was instituted, keeping in view of possible malignancy in current condition. Unfortunately, this regime could not prevent the further recurrence. So the patient was admitted again for the third time with malignant cachexia which led her fatal termination within a short period.

Case 3 was initially diagnosed and treated as a case of encysted tubercular peritonitis because of abdominal swelling, irregular fever, amenorrhoea and moderately high E.S.R. ultimately the correct diagnosis was clenched by diagnostic puncture with a thick bore needle, followed subsequently by laparotomy.

Treatment

The common dictum in the surgical treatment of pseudomucinous cyst is that it must be removed entire without tapping however big it may be in size. This is to avoid development of pseudomyxoma peritonei in future from the spilled gelatenous material coming in the peritoneal cavity following tapping of the cyst. But there is very little justification in such conjecture. Very often it leads to an unnecessarily long abdominal incision. In practice it has been found that even if there is accidental spill, often unavoidable, rarely if ever, it leads to any complications, certainly not to pseudomyxomatous changes. Even it has taken place, once the cyst has been removed, no pseudomyxomatous deposits will reform if the primary tumour was truly benign.

If on laparotomy pseudomyxoma peritonei is found together with psudomucinous cyst, the primary cyst should be removed and jelly like material from the peritoneal cavity should be removed as much as possible to relieve pressure symptoms. If the patients are in perimenopausal age, as is the usual feature, panhysterectomy is justified. If there is associated mucocele of the appendix, appendicectomy may be done as an additional procedure. When the omentum is extensively involved Shanks (1961) advocates removal of the omentum. Repeated laparotomies may be necessary to remove a vast quantity of jelly. But blind tapping of the abdominal cavity to

relieve pressure is not only unjustified but definitely harmful (Shanks, 1961). Intraperitoneal administration of Radioactive Gold advocated by Strauss and Strauss, (1952) may keep the condition in check for sometime.

Although deep X-ray therapy is advocated in the postoperative period by majority in the United States, Cariker and Dockerty (1954) from their experience in Mayo Clinic state that it would not cure pseudomyxoma, but helps in delaying spread. Malpas (1959) however states that pseudomyxoma peritonei is radio-resistant.

Prognosis

Willis (1952) has nicely summarized the problem of prognosis of pseudomyxoma peritonei by stating that "it is good in appendical cases, doubtful in 'benign' ovarian cases and hopeless in carcinomatous cases". Possible factors that influence the prognosis are—(1) extent of the disease, (2) primary ovarian cyst—benign or malignant, (3) implants in the peritoneal cavity completely removable or not, (4) panhysterectomy should be performed or not and (5) postoperative radiotherapy administered or not. Extensive involvement of the peritoneal cavity and histological demonstration of implanted cells from the pseudomucinous cyst is associated with a recurrence of the disease and, in some cases, may prove to be fatal.

Summary

1. Clinical and pathological studies have been made of three cases of pseudomyxoma peritonei.

2. A review of current literature has been made regarding its aetiology, clinical behaviour, prognostic criteria and problems in treatment.

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