PSEUDOMYXOMA PERITONEI

by

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Pseudomyxoma peritonei has provoked considerable controversy. Werth (1884) established the ovarian origin of pseudomyxoma peritonei in most of his cases. He considered that typical myxomatous deposits adherent to the peritoneum only develop with a lapse of time after the spontaneous rupture of a cyst. Virchow (1884) considered the condition due to implantation metastasis. Still a third view offered by Peters (1899) was that different types of pseudomyxoma of ovarian origin resulted from implantation metastasis, peritoneal irritation or a mixture of both reactions. Willis 1952 considered that pseudomyxoma peritonei is not a single entity of uniform causation and prognosis. .Novak (1952) states that living strands of epithelium are cast off from the lining of the cyst and get attached to the peritoneum and continue to secrete mucus. Shaw (1952) was of the opinion that the jelly-like material escaping from the ovarian cyst causes irritation of the peritoneum and the mesothelium of the peritoneum is converted thereby into tall columnar epithelium which secretes mucin. Pseudomucinous cystadenomas are often encountered in gynaecological practice. A relatively rare complication of this tumour is pseudomyxoma peritonei, characterised by an intensive production of an alkaline, extremely hydrophilic pseudomucin which is insoluble in water and jelly-like in consistency. It is generally accepted that the main source of the jelly is the direct spill of the contents of a ruptured loculus into the peritoneal cavity and continuing extravasation from the mucus secreting epithelial lining of the cyst. The problem arises why some cysts rupture and others of equal or greater size do not? Trauma to the abdominal contents as a result of external violence can be excluded as a factor, since rupture is invariably silent. Malpas (1959) points out that silent rupture is characteristic of infiltration of the malignant cyst wall by its invasive lining epithelium. Wilson (1912) believed that the jelly-like consistency of the contents of the loculus oozing slowly into the peritoneal cavity explained the lack of dramatic signs which usually accompany perforation of a hollow organ. In contrast to insidious rupture, accidental spilling of the gelatinous content of a benign cyst, during removal or evacuation before removal, rarely leads to pseudomyxoma peritonei. Cariker and Dockerty (1954) reported that spilling occurred on 63 occasions in the course of 217 laparotomies for benign mucinous tumours, without the development of pseudomyxoma.

The incidence of pseudomyxoma peritonei as a complication of pseudomucinous cystadenomas is reported to vary from 3.5 per cent to 9.2 per cent (Wilson,

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1912 Von Nagy, 1933, Shanks, 1961 and Kanaka Durgamba *et al*, 1964).

Case 1

Mrs. M, 48 years old, P2 + O was admitted on January 24, 1967 with enlargement of the abdomen for 12 years and a mass in the abdomen for 2 years. She was diagnosed as a case of ascites 11 years back for which paracentesis abdominis was performed. A bucketful of clear fluid was drained. Since then she has been having repeated paracentesis at intervals of 15 days to 3 months.

Menstrual cycles 3-4/30 days regular, flow normal, no dysmenorrhoea. For the last 5 months she had been having intermenstrual spotting off and on.

Obstetric history:

She had 3 full term normal deliveries. Last childbirth was 10 years back.

Physical Examination:

She appeared thin, emaciated and dyspnoeic. Abdomen was protuberant. There were visible veins all over the abdomen and multiple scars of 2 to 4 mm in diameter scattered all over the lower abdomen. She had 2 large ventral hernias which could be easily reduced. There was a mass of 25 x 20 cms filling the entire abdomen. Its consistency was cystic, surface nodular, dull on percussion and non-tender. Fluid thrill and shifting dullness were present.

Vaginal examination revealed a moderate degree of rectocele and a large enterocele. Cervix was directed forward, uterus was retroverted, normal in size. The abdominal mass could be felt with difficulty through the anterior and lateral fornices.

On speculum examination cervix appeared healthy.

Laboratory Investigations:

Haemoglobin 12.0 gm%; PCV—40%; blood sugar, blood urea, E.C.G., X-ray chest and liver function tests were within normal limits.

As the patient had dyspnoea, paracentesis was done on 14th February, 1967 and 14 pints of straw-coloured mucinous fluid was withdrawn. At this stage a provisional diagnosis of ovarian cyst with pseudomyxoma peritonei was made. After one week patient was again restless and 20 pints of straw-coloured mucinous fluid was drained by abdominal paracentesis. Smear made from the ascitic fluid was negative for malignant cells. It contained histiocytes, mesothelial cells and some polymorphs. Laparotomy was done on 24th February. Fifteen pints of straw-coloured mucinous fluid was sucked from the peritoneal cavity. After separating the adhesions between the parietal peritoneum and the omentum the ovarian cyst could be visualized. It was a multilocular pseudomucinous cyst. There was an opening of about 1 cm in diameter over the middle of the posterior wall of the cyst from which the mucinous fluid was seen leaking into the peritoneal cavity.

Right ovariotomy was done. After removing the ovarian cyst the uterus along with the adenexa of the left side could be visualised which were normal. There were multiple gelatinous nodules 1 to 3 cm in diameter in the pouch of Douglas. Total hysterectomy with left sided salpingo-oophorectomy was done. Enterocele was repaired by Moschowitzs' technique. Omentum, appendix, bowel, spleen and liver were normal. Peritoneal cavity was irrigated with normal saline. Ventral hernia was repaired and abdomen was closed. Her post-operative period was uneventful.

Histopathological Report:

Gross examination—large ovarian cyst measuring 28 x 20 x 10 cm. The cyst had burst in one area. The external surface was nodular, with moderate degree of congestion. On cut section, it showed multilocular cysts. The cysts measured 1-6 cms in diameter. They had smooth, transparent walls and contained clear mucinous fluid. There were areas of haemorrhages and necrosis. The uterus with the fallopian tube and ovary of the left side were normal. The nodules removed from the pouch of Douglas were well circumscribed measuring 2-3 cms in diameter. Cut surface revealed yellow-brown necrotic tissue.

Microscopic examination—Mucinous cystadenoma of the ovary with necrosis and old haemorrhages. The nodules removed from the pouch of Douglas revealed pseu-

domyxoma peritonei. Hernial sac showed pseudomyxoma with marked sclerosis.

Patient remained well for 1½ years after which she developed similar symptoms. On 21st January, 1969 she was readmitted with recurrent enlargement of the abdomen. She was given 30 mgm of Thio tepa intraperitoneally. After 2 days patient left against medical advice and was lost for follow-up.

Case 2

Mrs. B. K., aged 35 years was admitted on 8th May 1970 with enlargement of the abdomen, sensation of pressure and dull pain for seven months. Her menstrual cycles were regular 3-4/28 days, flow moderate and had no dysmenorrhoea. Her last menstrual period was on 28th April, 1970.

Obstetric History: P4 + 1. She had four full term normal deliveries. Last childbirth was 12 years back.

Physical Examination: Her general condition was fair. There was a suprapubic cystic mass with well defined margins, size of 24 weeks' pregnancy, arising from the pelvis, mobile non-tender and dull on percussion. Fluid thrill and shifting dullness were absent. On vaginal examination there was a second degree healed perineal tear and cystocele of moderate size with descent of the cervix upto the introitus. Uterus was retroverted and of normal size. The lower pole of the mass felt per abdomen could be felt through the anterior and lateral fornices with difficulty. A provisional diagnosis of ovarian cyst with third degree uterovaginal prolapse and cystocele was made.

Laboratory investigations: Haemoglobin 11 gm%; P.C.V. 32.5%, blood sugar, blood urea, urine, liver function tests and X-ray chest were within normal limits.

On 30th May 1970, laparotomy was done. On opening the peritoneal cavity jelly-like substance was encountered. On removing the 'jelly' an ovarian cyst of about 20 x 10 cms arising from the left side came into view. There was an opening about 2 cms in diameter over the upper aspect of the posterior wall of the cyst from which the jelly' was seen leaking into the peritoneal cavity. Her appendix was distended in the lower half. Total hysterectomy with removal of both appendages, appendicectomy and peritoneal toilet were performed. 30 mgm. of Thio tepa was put into the peritoneal cavity. Her post operative period was uneventful. She had no evidence of recurrence till 20 th April, 1971.

Histopathology Report:

Gross examination: Ovarian cyst measured 26 x 14 x 10 cms. The cyst contained gelatinous material. Cyst was seen to be multilocular with smooth wall. The appendix measured 2.5 x 1.5 x 0.5 cm. Cut surface showed thickened walls. Gelatinous material removed for the peritoneal cavity weighed 2.5 kgms.

Microscopic examination: Mucinous cystadenoma of the left ovary. The left fallopian tube shows endometriosis. The uterus with the fallopian tube and ovary of the right side were normal, and there was a mucocele of the appendix.

Discussion

The interesting features of the two cases reported were that case 1 had paracentesis done repeatedly for 11 years. At laparotomy there was free flow of mucinous fluid, absence of intestinal adhesions and benign nature of the ovarian cyst. Jones (1965) reported a case in which 75 paracentesis abdomini were carried out. In case 2, pseudomucinous cyst of the ovary was associated with mucocele of the appendix.

Pseudomyxoma peritonei, which might better be termed myxomatous peritonitis or mucinous ascites, is a condition in which the peritoneal cavity is filled with gelatinous mucin, which is chemically an acid mucopolysaccharide. It is associated with and arises from mucinous neoplasms of the ovary and appendix. The diagnosis is not difficult in these cases. The majority of the patients are in the 6th decade but it may also occur in the younger age group. Usual symptoms are gradual painless abdominal enlargement, breathlessness, oedema of the feet, urinary frequency may be present, colicky abdomi-

nal pain, ventral or femoral hernia. Fluid thrill and shifting dullness may be elicited depending upon the viscosity and the quantity of the fluid. Diagnosis during life is usually made by a paracentesis or laparotomy. Rosenfeld (1949) stated that blood sugar may be very low and there may be a change in albumin and globulin ratio. Cytology of the peritoneal fluid may clinch the diagnosis. In cases of pseudomyoma peritonei three types of cellular elements are seen, mesothelial cells, fibrotic cells, and epithelial cells which are cone-shaped or cylindrical.

Treatment: Surgical treatment is necessary to establish the diagnosis, remove the primary cyst and relieve the pressure symptoms by scooping out as much jelly as possible. Since the majority of the patients are premenopausal or post menopausal, removal of the uterus and other ovary is logical. The frequent association of mucocele of the appendix makes appendicectomy an essential additional procedure. If omentum is involved it should be resected. Wilson (1912) advocated irrigation of the peritoneal cavity with normal saline. Strauss and Strauss (1952) suggested instillation of enzyme 'wydase.' It converts the jelly into watery fluid which is easy to tap. The effect of this enzyme lasts for a few weeks only and hence repeated instillations are necessary. The enzyme has no effect on the primary tumour itself. Ralph *et al* (1963) irrigated the peritoneal cavity with 10 mgms. of mechlorethamine in 200 c.c. of water. His patients remained well 52 months, 48 months, 19 months after definitive therapy. Most of the authorities consider the condition resistent to radiotherapy (Malpas, 1959). In the United States of America it is generally agreed that deep X-ray therapy should be given post operatively. Cariker and Dockerty (1954) are of the opinion that it does not cure pseudomyxoma peritonei but helps in delaying the spread.

Relief of pressure symptoms is the commonest indication for further surgical treatment and repeated laparotomies may be required to relieve mechanical intestinal obstruction due to adhesions.

Prognosis: The prognosis depends on the growth capacity of the responsible cells, degree of malignancy, extent of deposits and adhesions. The disease has a tendency to recur. Willis (1952) stated that it is good in appendical cases, doubtful in 'benign' ovarian cases and hopeless in carcinomatosis cases. It is difficult to evaluate the incidence of malignancy because of lack of uniformity in the interpretation of the criteria of malignancy. Shank (1961) took 5 years survival as an indication of the benign nature of the disease. In the Mayo Clinic the incidence of malignancy is 39 per cent whereas in Birmingham it is 19.2 per cent. Masson and Hamrick (1930) reported an operative mortality of 12.8 per cent. Patients usually die from sepsis, toxaemia, due to localised intraperitoneal or retroperitoneal abscess, intestinal obstruction, deep vein thrombosis and pulmonary embolism.

Summary

Two cases of pseudomyxoma peritonei associated with the pseudomucinous cyst of the ovary and mucocele of the appendix are reported. Incidence, etiology, management and prognosis are discussed.

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