PSEUDOMYXOMA PERITONEI: A REVIEW OF 17 CASES

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ABSTRACT
Objective: To evaluate the various presentations, causes and management of Pseudomyxoma Peritonei (PMP), and to create awareness among young surgeons regarding its importance.
Design & Duration: Retrospective study from Jan. 1999 to Dec. 2005.
Setting: Surgical Unit of Khyber Teaching Hospital, Peshawar and Gynaecology & Obstetrics Unit, Hayatabad Medical Complex, Peshawar.
Patients: All patients who had Pseudomyxoma Peritonei during the study period.
Methodology: The clinical records of all patients undergoing laparotomy for various causes were reviewed and cases of PMP separated. Their biodata, clinical presentation, clinical diagnosis, investigation results, operative findings, histopathology report and outcome were recorded.
Results: Out of 23,573 cases that underwent laparotomy 17 cases of PMP were on record. All were diagnosed incidentally per-operatively. Most (47.05%) cases were seen in the age group of 30-40 years and 58.82% patients were females. Abdominal pain, mass, abdominal distension and intestinal obstruction were the common presenting features. Ultrasound and CT scan reported three cases as ovarian cysts, three as multiple encysted fluid collections with thin cyst walls, two as ascites, two as ascites with peritoneal thickening, but none as PMP. Surgery was the mainstay of treatment. Appendicectomy with clearing of mucin in 11(64.70%), oophorechtomy and appendicectomy in 3(17.64%), bilateral oophorechtomy and hysterectomy in 2(11.76%), right hemi-colectomy in 1(5.88%) and second look surgery for complications were done in 2(11.76%) cases. Mortality was 11.76% and due to complication including one due to intestinal obstruction and another due to septicemia following second look surgery. Histopathological tissue diagnosis was available in only 13 cases and included mucinous adenoma appendix in five, mucinous cystadenoma ovary in three, mucinous epithelial cells of unknown origin in four and mucinous cystadenoma of borderline malignancy in one case.
Conclusion: PMP is under reported in our setup. Pre-operative diagnosis is difficult and incidental findings usually go unnoticed due to lack of awareness and standard management protocol. There is a need to create awareness among surgeons, radiologists, pathologist and oncologist regarding this condition for the better outcome.

KEY WORDS: Pseudomyxoma Peritonei, Gelatinous Ascites, Ovarian Cyst, Mucocoele Appendix

INTRODUCTION

Pseudomyxoma Peritonei (PMP) is a rare condition that is characterized by gelatinous ascites, associated with gelatinous implants on the peritoneal surfaces and the omentum[1,2]. It is a pathologic diagnostic term applied to both benign and malignant mucinous neoplasms that produce abundant extracellular mucin resulting in a variable and poorly predictable prognosis[3]. It is found more frequently in females and usually occurs following pseudomucinous cyst of ovary or rupture of a mucocele of appendix[4]. Its incidence is 2/10,000 laparotomies[5]. The diagnosis is often delayed due to vague symptoms, though progressive abdominal distension and discomfort, pain, appendiceal or pelvic mass with no impairment of general health are common presentations, or it may be an incidental finding at laparotomy.

The aetiology and treatment of this condition is currently controversial[6]. Recently cytoreductive surgery with

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intraperitoneal hyperthermic chemotherapy has emerged as a promising treatment for this debilitating condition[7].

Lack of proper pre-operative diagnosis, confirmatory laboratory investigations and little knowledge about the condition results in under reporting of the condition. Surgeons and gynecologists often come across a few cases of PMP in their clinical practice.

This study aimed to describe the various presentations, aetiology and the treatment of pseudomyxoma peritonei in our clinical setup, and also to create awareness among young surgeons and gynecologists regarding the importance of the condition.

PATIENTS & METHODS

This retrospective study was carried out by the Surgical and Gynecology Units of Khyber Teaching Hospital and Hayatabad Medical Complex, Peshawar from Jan. 1999 to Dec. 2005. The clinical records of all the patients undergoing laparotomy during the study period were reviewed. Patients characteristics including age and sex, clinical presentation, investigations performed, provisional diagnosis, laparotomy findings, surgical procedure carried out and any adjuvant treatment provided, as well as the histopathological diagnosis were recorded on a proforma. The follow-up record was not well maintained, but was recorded where available. The data was compiled and the results tabulated.

RESULTS

Out of the total 23,573 patients that underwent laparotomy for various causes during the study period, 17 were diagnosed as Pseudomyxoma peritonei. All were incidentally diagnosed during laparotomy, when jelly like mucinous material was found all over the peritoneal cavity.

Table II. Clinical Features (n=17)

Presentation	No.	%
Symptoms		
Pain in the abdomen	8	47.05
Intestinal obstruction	2	11.76
Abdominal distension	3	17.64
Mass	4	23.52
Signs		
Tender rt. iliac fossa	7	41.17
Palpable mass	4	23.52
Ascites and distended abdomen	4	23.52
Tender abdomen	2	11.76

Pts. Characteristics	No.	%
Age		
< 20 years	2	11.76
21-30 years	3	17.64
31-40 years	8	47.05
> 40 years	4	23.52
Sex		
Males	7	41.17
Females	10	58.82

Table I. Demography (n=17)

Table I shows the age and sex distribution of patients, 10 (58.82%) were female and 7 (41.18%) male. The commonest symptom was abdominal pain and the tenderness in right iliac fossa the commonest sign (Table II). Results of the different investigations are shown in Table III. Tumour markers were done in six cases, mostly after surgery; CA 125 levels were found raised only in three patients.

Table III. Investigations & Findings (n=17)

Investigation & Findings	Number
Ultrasound (16)	
Cystic mass	5
Ascites	4
Distended loops	4
Encysted fluid collection	3
CT Scan (7)	
Ovarian cyst	3
Focal collection	2
Ascites with septae	2
X-Ray Abdomen (9)	
Fluid levels	4
Unremarkable	5

Procedure	No.	%
Appendicectomy with clearing of mucin	11	64.70
Oopherectomy and Appendicectomy	3	17.64
Bilat. Oopherectomy and Hysterectomy	2	11.76
Right Hemicolectomy	1	5.88

Table IV. Surgical procedures (n=17)

All patients underwent surgery; Appendicectomy was the commonest procedure (82.34%) done (Table IV). Tissue was sent for histopathology in all the cases but 13 cases of PMP were confirmed on tissue diagnosis (Table V). Two cases underwent second look surgery for abdominal distension and intestinal obstruction; palliative ileostomy was done. Two (11.76%) patients died in this series; both had intestinal obstruction. One died during surgery due to wide spread disease and debilitating condition and the other after second look surgery due to septicemia. Follow-up was not well maintained, but five patients are still coming, two have vague abdominal pain on and off and another obstructive symptoms, the rest two are symptom free. Adjuvant chemotherapy with cisplatinum, cyclophosphamide and adriamycin was given to one patient with mucinous cystadenoma of border line malignancy on the advice of oncologist. However, none of the remaining patients received intraperitoneal or adjuvant chemotherapy.

DISCUSSION

PMP was first described by Karl in 1842 as gelatinous ascites with multifocal peritoneal epithelial implants secreting copious mucin[8]. Recent studies have redefined it as a specific clinicopathologic syndrome in which the mucinous epithelium is intimately associated with pools of extracellular mucin and fibrosis. It is diagnosed pathologically as disseminated peritoneal adenomucinosis (DPAM)[9].

It is more common in females, nine out of 17 diagnosed cases, with a mean age of 53 years[6,10]. In another series 38 out of 64 patients were females[11]. Similarly in our study 58.82% were females. It was common in adults with 11 patients in the 3rd and 4th decades of life; only three cases were above 40 years of age.

Clinical presentation is variable and pre-operative diagnosis difficult[10,12]. The condition is usually discovered as an incidental finding at laparotomy[13], as was the case in our study. Most (47.05%) patients presented with pain; abdominal mass and/or distension, and intestinal obstruction were the other presentations. An abdominal mass was felt in four (23.52%) cases clinically and tenderness in right iliac fossa in seven (41.17%) patients. In a study comprising of 217 cases, 27% presented as suspected acute appendicitis and 23% with increasing abdominal distension[14].

Histopathology	No.	%
Mucinous Adenoma Appendix	5	29.41
Mucinous Cystadenoma Ovary	3	17.64
Mucinous epithelial cells of unknown origin	4	23.52
Mucinous Cystadenoma of borderline malignancy	1	5.58

Table V. Histopathology (n=13)

Imaging techniques are helpful in some cases. Ultrasonography demonstrates minute quantities of free fluid in the peritoneal cavity and provides quantitative and qualitative information about ascites, ovarian cysts, encysted fluid collections, and particularly appendiceal mucocele which appears as an elongated echo-poor mass, with less distinct cyst walls than expected[15,16]. Computed tomography (CT) is more helpful; PMP is characterized by low quantity mucinous ascites, septae and calcification in wide spread disease[10]. Visceral scalloping is classically attributed to PMP[17]. In our review ultrasound showed cysts in five cases, ascites in four and loculated fluid collection with thin walls in three cases. CT was done only in seven cases but the findings were not interpreted as PMP.

The origin of PMP is controversial but recent evidence supports an appendiceal rather than ovarian origin for virtually all cases of PMP, ovary may be involved in some females[9,11,13]. Appendiceal cause was common in our study as well and ovarian mucinous tumour was diagnosed in four cases. It has also been reported a few years after appendicectomy and also after urachal adenocarcinoma[4,18].

No effective treatment is known, modern treatments include radical surgical excision with appendicectomy (and oopherectomy in women), and adjuvant hyperthermic intraperitoneal and systemic chemotherapy[19]. Appendicectomy with clearing the mucin collection

was done in 11 of our patients, appendicetomy and oopherectomy in two cases, bilateral oopherectomy with hysterectomy and clearance of mucin from peritoneal cavity in three cases and right hemicolectomy in one patient as the base of the appendix was indurated. In a study on 45 patients of PMP, six peritonectomy procedures including total gastrectomy were required for complete cytoreduction[20]. Extensive or optimal surgical cytoreduction combined with intraoperative heated intraperitoneal chemotherapy (HIPEC) in 46 patients improved survival rate by 81% at 3 years[21]. HIPEC with mitomycin-C heated to 42 to 44 degrees to treat microscopic residual disease as hyperthermia enhances penetration of cytostatic drugs into tumour tissue, an overall 5 year survival upto 86% was achieved[22]. Bowel obstruction, biliary obstruction, bone marrow suppression, abdominal pain, seizures, neutropenia and thrombocytopenia were common causes of morbidity and mortality with this treatment[23]. Chemotherapy for PMP was not used in any of our patients because of retrospective diagnosis and lack of standard management protocol. The oncologist did not advise it except in one case with mucinous cystadenoma ovary of borderline malignancy who was given a cisplatinum containing regime.

CONCLUSION

PMP is difficult to diagnose clinically, and is usually diagnosed per-operatively. Many surgeons are exposed to this condition but due to inexperience fail to identify it. Variable presentation along with lack of proper planning and standard protocol for management leads to the under reporting of this condition.

Proper pre-operative workup for diagnosis, appropriate surgical procedure, adjuvant chemotherapy and increased awareness regarding PMP among clinicians is a must for proper management and better outcome. Characteristic CT scan appearances needs to be highlighted. The importance of follow-up should be emphasized, though the patients mostly disappear after operation and return only when they develop complications.

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