JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH

How to cite this article:
FERNANDES H , D'SOUZA CLEMENT R S , NAIK RAMESH C N , MARLA J N , PAILOOR K . PSEUDOMYXOMA PERITONEI –A RARE CLINICAL ENTITY.Journal of Clinical and Diagnostic Research [serial online] 2010 April [cited: 2010 April 5]; 4:2286-2290.

Available from
http://www.jcdr.net/back_issues.asp?issn=0973-709x&year=2010 &month= April &volume=4&issue=2&page=2286-2290 &id=563

CASE REPORT

Pseudomyxoma Peritonei –A Rare Clinical Entity

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ABSTRACT

Pseudomyxoma Peritonei (PMP) is a rare, chronic, poorly understood disease that is characterized by disseminated mucinous ascitis and peritoneal implants. In order to diagnose PMP, viable epithelial glandular cells must be identified within the mucinous pools by histological analysis. We report here, two male patients and one female patient who presented to our hospital within a span of 4 months. Histopathology records of the last 10years do not reveal any case of PMP. In case1, the diagnosis of PMP was made after laparotomy, as the radiological findings suggested hollow viscus perforation. The case 2 patient had second debulking surgery after 11 years. The case 3 patient developed PMP 3 years after the first surgery. In this patient, preoperative diagnosis was made due to the typical CT findings and appendicial origin was confirmed by CK 7 negativity. All three patients underwent debulking surgery and were referred to the Oncology department for further treament. Histopathology reports of case 1 and case 3 revealed benign epithelium, whereas case 2 had foci of borderline epithelium, indicating disease progression. All three were symptom free after a short follow up of one, two and three months in case 1, 3 and 2, respectively. In all the three cases, PMP was secondary to tumours of the appendix.

Key Words: pseudomyxoma peritonei, mucinous cystadenoma appendix, DAPM, PMC

Key Message: Awareness of this indolent and rare condition is an important prerequisite for early diagnosis and appropriate management.

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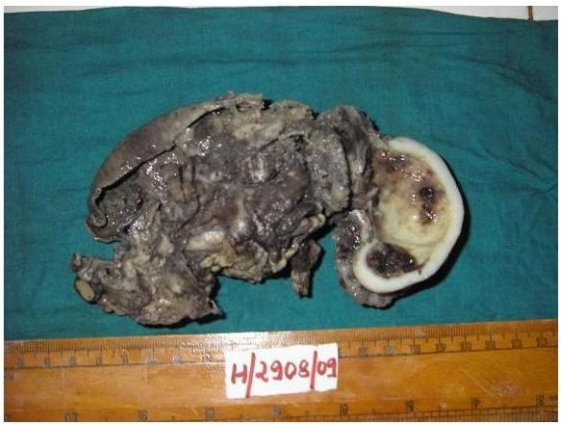
Introduction

Pseudomyxoma Peritonei (PMP) is a clinical term used to describe the finding of abundant mucoid or gelatinous material in the pelvis and abdominal cavity, which is surrounded by fibrous tissue [1]. Viable epithelial glandular cells must be identified within the mucinous pools by histological analysis in order to diagnose PMP. Pseudomyxoma Peritonei is an extremely rare tumour that is generally discovered intraoperatively, with an incidence of 2 in every 10,000 laparotomies [2]. Though PMP is more commonly seen in women, we report here, 3 cases of PMP, out of which two are men, who presented to our hospital within a span of 4 months (April–July 2009).

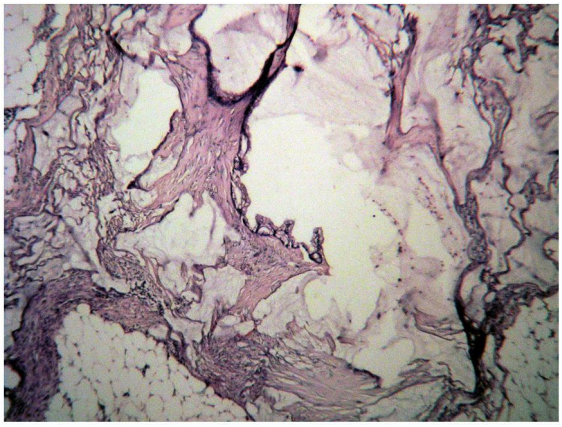
Case 1

A 69 year old male presented with pain in the abdomen of sudden onset. He also complained of fever of a duration of 3 days. On examination, tenderness was found to be present all over the abdomen. He was a diabetic with no previous history of surgery. Ultrasound and Computed Tomography (CT) scan revealed ill defined septate collections in the pelvis, right ileac fossa, paracolic gutters and morrison's pouch. Apart from this, it also showed free intraperitoneal air, suggestive of hollow viscus perforation with peritonitis and paralytic ileus. Extension of collection into bilateral inguinal

canals was also seen. Laparotomy revealed multiple peritoneal, omental mucinous deposits and a dilated appendix. Purulent fluid which had collected in the pelvis was drained out. Resection of ileocaecal segment with anastomoses and loop ileostomy was done. Resected specimen consisted of an ileocaecal segment with multiple fibromucinous masses. The appendix was dilated [Table/Fig 1] (Fig 1a), with lumen showing gelatinous material. A perforation was noted at the base, with the adjacent areas showing fibromucinous nodules. Histology revealed a single layer of bland epithelium at places, forming stratification and papillary infoldings in pools of extracellular mucin [Table/Fig 1] (Fig 1b). Surrounding stroma showed fibrosis and inflammatory infiltrate. Sections from the dilated appendix also revealed similar epithelial lining and pools of mucin. A diagnosis of mucinous cystadenoma of appendix with PMP was made.



(Table/Fig 1) Fig 1a: Specimen Showing Dilated Appendix

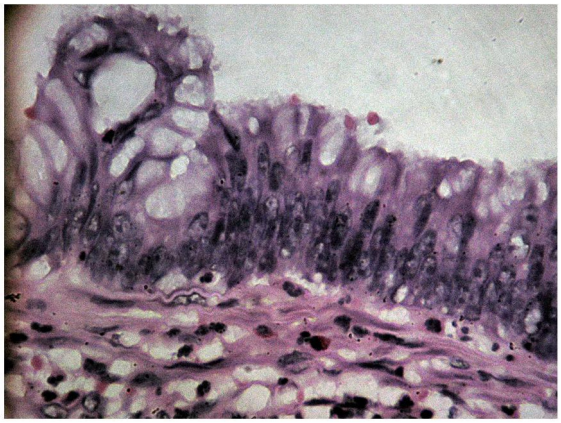


(Table/Fig 1) Fig 1b: Section Showing Bland Mucin Secreting Epithelium In Extracellular Pools Of Mucin.

Case: 2

A 48 year old male with a history of surgery for mucinous cystadenoma of the appendix with PMP 11 years ago, presented with complaints of abdominal distention. Abdominal examination revealed multiple firm masses in the epigastric, right ileac and lower abdomen regions. CT scan detected a cystic mass with septations involving the mesentery, omentum, lesser sac and perigastrium indenting the gastric wall, scalloping the body and the tail of the pancreas. He underwent exploratory laparotomy for debulking of PMP. The specimen consisted of right hemicolectomy and mucoid material amounting to about 2 litres. The hemicolectomy specimen showed multiloculated mucoid cystic lesions in the ileocaecal junction and the hepatic flexure [Table/Fig 2](Fig 2a). Microscopy revealed bland epithelium in pools of mucin with focal areas showing papillary tufting, stratification and prominent nucleoli [Table/Fig 2] (Fig 2b).

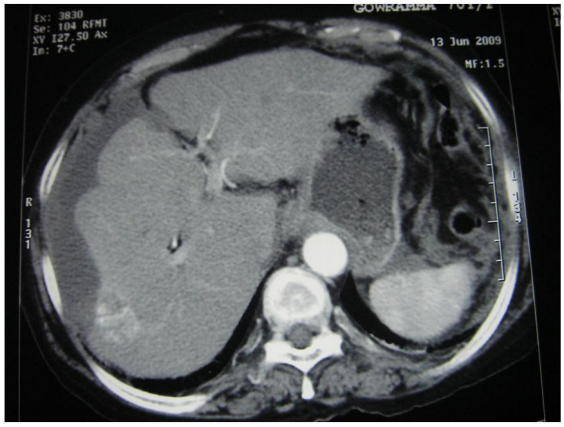
(Table/Fig 2) Fig 2a: Specimen Showing Multiloculated Mucoid Cystic Lesion in Ileocaecal Junction.



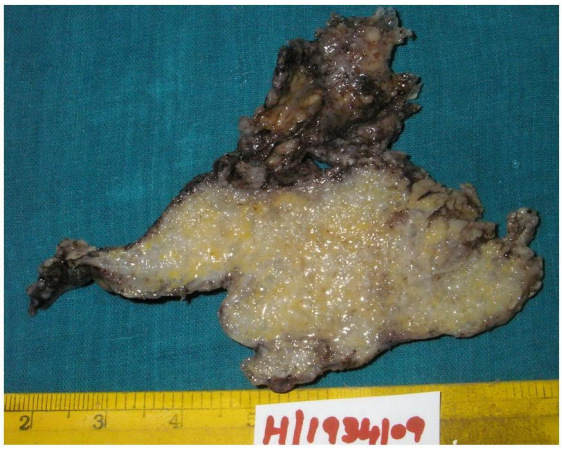
(Table/Fig 2) Fig 2b: Section Showing Papillary Tufting, Stratification and Prominent Nucleoli.

Case 3

A 68 year old female was admitted for pain and distension of the abdomen, with a similar history 3 years ago, for which she underwent surgery (Details of which are not available). Her abdominal examination revealed ascitis. CT scan detected gross ascitis with fluid collection, causing scalloping of the diaphragmatic surface of the liver and omental surface thickening and peritoneal calcification [Table/Fig 3] (Fig 3a). Debulking surgery showed mucinous deposits on the surface of the liver, spleen, omentum and the posterior abdominal wall. Evidence of surgical removal of the uterus, bilateral ovaries and appendix was noted. The specimen consisted of an omental cake [Table/Fig 3] (Fig 3b) and multiple mucoid nodules. The histolopathological findings were similar to that of case 1. Appendicial origin was confirmed by CK7 negativity.



(Table/Fig 3) Fig 3a: CT Scan Showing Scalloping Of Diaphragmatic Surface Of The Liver.



(Table/Fig 3) Fig 3b: Specimen of Omental Cake with Mucinous Areas.

Discussion

Pseudomyxoma peritonei, first described by Werth (1884), is an indolent disease, preferentially affecting women of an average age of 53 years. Most often, the patient presents with progressively increasing abdominal girth. Males commonly present with inguinal hernia, while females present with an ovarian mass. Many cases are incidentally detected at laparatomy done for acute abdomen such as acute appendicitis orpartial or complete bowel obstruction, by finding a belly full of jelly - "The jelly belly".

An accurate preoperative diagnosis of PMP can be aided by radiological imaging with CT if the characteristic "scalloping effect" on the surface of the visceral organs resulting from

compression by viscus mucinous secretions and the organizing fibrosis is seen [3]. Characteristic distribution of mucin deposits in the peritoneal cavity follow the principle of redistribution, based on the intraperitoneal fluid dynamics favouring deposits at all sites of fluid reabsorption [4].

The site of origin of PMP is a matter of debate. Current evidence suggests that PMP results from the rupture of a mucinous cystadenoma or mucinous cystadenocarcinoma of the ovaries or appendix, with appendecial involvement being more common [1]. Other sites include the pancreas, bile duct, colon, gall bladder, urinary bladder, breast, lung urachus and the renal pelvis [5]. In women, it may be associated with mucinous tumours of the ovary. In women without any detectable cystic ovarian lesion and in men, the primary source is a low-grade mucinous tumour of the Gastrointestinal Tract, usually the appendix. PMP can occur years (5 - 35 years) later, after the initial presentation of an appendicial event [3]. The removal and thorough histological examination of the appendix is indicated in cases of PMP with a mucinous cystic ovarian tumour. In cases where an appendiceal mucinous neoplasm is found, it should be considered as the primary site and the ovaries should be considered as secondary [1]. In equivocal cases or when the details of the previous surgery are not available, as in case 3, cytokeratin 7 negativity strongly suggests appendicial origin [6].

Numerous researchers have attempted to classify PMP from the time of its first description by Werth (1884). Most recently, these tumours have been classified into diffuse peritoneal adenomucinoses (DPAM) and peritoneal mucinous carcinomatosis (PMC) [1]. PMP with benign and borderline appearance have been termed as DPAM and are usually secondary to lowgrade mucinous tumours of the appendix [4]. Patients with this finding have a benign or protracted clinical course. In malignant epithelium (PMC), the clinical course is usually fatal and is generally secondary to an adenocarcinoma of the appendix or the ovary. Recent studies reveal that PMP is a neoplastic disease of MUC-2, expressing the goblet cells [7]. Mucinous tumours of the appendix also express MUC-2, which supports an appendiceal rather than ovarian origin for PMP. Clinical symptoms may be caused by an overwhelming increase in MUC-2 secreting cells, as well as the fact that the excessively produced mucin has no place to drain. Despite it's tendency to spread and it's sometimes malignant nature, solid organ invasion and metastasis are rare. Death is frequently caused due to intestinal obstruction or an unrelated cause.

Currently, debulking surgery with or without intraoperative hyperthermic peritoneal chemotherapy is the standard treatment [8]. Combined treatment which is usually given for PMC, has high toxicity (particularly surgically related). Prognosis in this disease is closely related to the bulk of the disease, as evaluated by the tumour site, preoperative tumour volume and the completeness of tumour removal by cytoreductive surgery and by the microscopic degree of differentiation of the neoplastic epithelium, as evaluated by histopathological examination [9]. PMP with preoperative elevated tumour markers such as CEA (carcinoembryonic antigen) and CA19-9 are at an increased risk of developing recurrent disease despite aggressive therapy [10].

Conclusion

Pseudomyxoma peritonei almost always originates from a perforated appendiceal tumour. It is important to accurately classify the histological findings, owing to its correlation with clinical behaviour. These patients have a prolonged clinical course and often require repeated surgery to remove mucinous material. Awareness of this indolent and rare condition is an important prerequisite for early diagnosis and appropriate management.

References

[1] Tavassoli FA, Devilee P. Pathology and genetics of tumors of breast and female genital organs. World Health Organisation classification of tumors .IARC Press, Lyon , 2003; 123-24.
[2] Mann WJ Jr,Wagner J,Chumas J, Charles E. The management of Pseudomyxoma Peritonei. Cancer 1990; 66: 1636-40.
[3] Li C, Kanthan R, Kanthan SC. Pseudomyxoma Peritonei a revisit: Report of 2 cases and

literature review. World J. Surg Oncol 2006; 4:60
[4] Sugarbeker PH. Pseudomyxoma Peritonei. A cancer whose biology is characterized by redistribution phenomenon. Ann Surg 1994; 219:109-11
[5] Shah V. B, Amonkar GP, Deshpande JR, Bhaleka Mucinous adenocarcinoma of the renal pelvis with pseudomyxoma peritonei. Indian J. Pathol. Microbiol-2008; 51:536-37.
[6] Guerrieri C, Franlund B, Fristedt S, Gillooley JF, Boeryd B. Mucinous tumors of the vermiform appendix and ovary and pseudomyxoma peritonei. Histological implications of cytokeratin K expression. Human Pathol. 1997; 28:1039-45.
[7] O'connell JT, Tomlinson JS, Robert AA, McGonigle KF, Barsky SH: Pseudomyxoma Peritonei is a disease of MUC-2 expressing goblet cells. Am J. Pathol 2002; 161:551-64.
[8] Pestieau SR, Yan Hui, Shmookler M, Sugarbaker PH. Histopathologic Analysis in 46 patients with Pseudomyxoma Peritonei Syndrome: Failure versus Success with a second look operation Mod Pathol 2001; 14:164-71.
[9] Miner TJ,Shia J,Jaques DP,Klimstra DS,Brennan MF,Coit DG. Longterm survival following treatment of pseudomyxoma peritonei.An analysis of surgical therapy. Ann Surg 2005; 241:300-08.
[10] Carmignani CP,Hampton R,Sugarbaker CE,Chang D,Sugarbaker PH. Utility of CEA and CA19-9 tumor markers in diagnosis and prognostic assessment of mucinous tumors of the appendix. J Surg Oncol 2004; 87:162-66.