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# Pseudomyxoma Peritonei : A Case Report

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**Abstract:**

Pseudomyxoma peritonei is a relatively rare and poorly understood condition in which mucus accumulates within the peritoneal cavity. The presence of cells in the mucin, either inflammatory or neoplastic, distinguishes it from simple acellular mucus ascites caused by mucinous spillage. There is widespread seeding of the peritoneal and omental surfaces with a heavy cancerous glaze. This is principally a complication of borderline or malignant neoplasm of the ovary and/or appendix. This paper describes a case of previously healthy women who presented with an acute abdomen, and was diagnosed postoperatively with pseudomyxoma peritonei.

**Keywords:** Pseudomyxoma peritonei.

## Introduction

Pseudomyxoma peritonei (PMP) first described by Werth [1] is an uncommon and poorly understood disease characterized by abundant extracellular mucin in the peritoneum. The "myxomatous" appearance is attributed to the associated fibroblastic and vascular proliferation that is probably incited by the mucin. This results in multifocal peritoneal, serosal and ommental implants admixed with copious amounts of mucin accumulation within the abdomen and pelvis resulting in the belly full of jelly – "the jelly belly" [2]. PMP is a broad descriptive term embracing a wide spectrum of biological behavior of neoplasms from the benign to the borderline to the frankly malignant lesion. Other terminologies to reflect this spectrum of biological behavior ranges from d isseminated p eritoneal a denom ucinosis (DPAM) –the benign variant to p eritoneal m ucinousc arcinoma tosis (PMCA) – the malignant variant [3]. A definitive diagnosis of PMP requires the presence of a) mucinous neoplastic cells/epithelium, and b) mucinous ascites – diffuse intraabdominal mucin. Some authors also require the presence of diffuse mucinous implants for this diagnosis. Viable epithelial glandular cells must be identified within the mucin pools by histological analysis to diagnose PMP. Cases without epithelium are regarded as mucinous ascites.

Pseudomyxoma peritonei (PMP) is more commonly seen in women [4] who usually present with increasing abdominal girth and this tends to be related to underlying ovarian lesions which are usually mucinous tumors that can be associated with a teratoma. Though uncommon in men, these cases are virtually all associated with a lesion in the appendix [5]. Other possible primary sites include colorectum, gallbladder, pancreas, urachus, urinary bladder, breast and lung [6], but these are uncommon. These primary sites are usually associated with the malignant variant-PMCA of PMP. PMP can occur years (range from 5–35 years) later after the initial presentation of an appendiceal event [7] and, therefore, accurate diagnosis prior to surgery is often delayed and inaccurate. The disease may be localized in the right lower quadrant

initially and then become more generalized with mucinous peritoneal, serosal and ommental implants. There is increasing recognition that the two variants DPAM and PMCA are different with the DPAM remaining localized to the abdomen without metastatic behavior and the PCMA behaving like a mucinous (colloid) carcinoma with metastatic and invasive potential. 10% of patients die of PMP within 5.5 years of their initial presentation while others experience recurrent and/or residual disease. Advanced abdominal disease leading to intestinal obstruction accounts for majority of the patients' morbidity and mortality.

**Case Report**

A 54 year old woman was admitted with a 24 hour history of severe right iliac fossa pain associated with nausea, sweating, and hot and cold flushes. Over a period of a few hours, the niggling pain had moved from the central abdominal region to the right iliac fossa. She had no other abdominal, gynaecological, or urinary symptoms. At presentation, she was in considerable pain, apyrexial with right iliac fossa tenderness, guarding, and rebound tenderness No masses were palpable. Laboratory investigations including full blood count, amylase, liver function tests, urea and electrolytes were all within normal limits.

Tumour markers were negative. On clinical grounds, a diagnosis of local peritonitis secondary to acute appendicitis was made. Ct scan showed mild to moderate ascitis with diffuse omental and mesenteric fat stranding and subtle omental nodularity along the anterior abdominal wall. A well defined cystic attenuated lesion with thin peripheral wall enhancement and eccentric wall calcification is seen in the right iliac fossa, abutting the caecal tip, in the expected location of the appendix which is not identified discretely from it. It was suggestive of an appendiceal mucocele [Figure 1]. Patient was posted for a diagnostic laparoscopy. Pneumoperitoneum was created with a verres, ports were put one in the lt iliac fossa and the other in the suprapubic area. Mucinous material was seen all over the abdomen with nodules over the peritoneum, broad based appendix with mucocele was noted [Figure 2]. Appendisectomy was done and the omental and the peritoneal biopsy was taken. The patient was discharged on the third postoperative day. The histopathological report was traced to be mucinous adenocarcinima of the appendix. The patient was later posted for a exploratory laparotomy with right hemicolectomy with omentectomy with parietectomy.The patient had an uneventful post operative course.

**Discussion**

Pseudomyxoma peritonei is an indolent disease and preferentially affects women with an average age of 53 years [4]. It is traditionally believed that most cases of PMP originate from ovarian tumors. This belief is challenged recently by increased usage of immunohistochemical stains and molecular genetic studies, which showed a large proportion of these tumors to be secondary to appendiceal tumors in both men and women [5, 8, 9, 10]. The fact remains that more women than men suffer from this condition according to the published literature [4].

As symptoms remain non-specific the disease presents a great diagnostic challenge to clinicians. Clinical presentation is late and patients usually experience a long course of health deterioration before an accurate diagnosis is made. Due to its indolent nature, advanced stages of the disease with generalized peritoneal tumor implants, fistula formation and adhesions are common. In this advanced stage, abdominal symptoms caused by partial or complete bowel obstruction are the main complaints.

A precise diagnosis is difficult due to the lack of specific symptoms in the early stage of the disease. Routine

laboratory studies are seldom helpful in making this diagnosis. An accurate preoperative diagnosis of pseudomyxoma peritonei can be aided by radiological imaging with computed tomography if the characteristic "scalloping effect" on the surface of the visceral organs resulting from compression by the viscous mucinous secretions and the organizing fibrosis [1].

The role of magnetic resonance imaging as a diagnostic tool is unclear. In the majority of cases, it is often an unexpected finding at explorative laparoscopy, which remains the main diagnostic tool with the final diagnosis being confirmed by histopathology.

Mucinous neoplasms of the appendix are uncommon entities associated with a variety of underlying pathological processes ranging from simple appendiceal mucoceles to mucinous hyperplasia (hyperplastic polyp), serrated adenoma, mucinous adenoma/cystadenoma, mucinous neoplasm of uncertain malignant potential, mucinous neoplasm of low malignant potential and mucinous cystadenocarcinoma.[11]

The exact relationship of these lesions to PMP is still not clear though many cases of PMP, predominantly the DPAM variant, are associated with mucinous neoplasms of the appendix with low malignant potential (M-LMP). The PMCA variant of PMP seems to have a higher association with mucinous adenocarcinomas of the appendix and other rarer gastrointestinal sites. The pathological features of a borderline or low malignant potential appendiceal mucinous neoplasm consists of glands lined by mucinous epithelium with small basally located nuclei and inconspicuous nucleoli in a mucinous background as in a mucinous adenoma. The key associated finding is the presence of epithelium outside the appendix in association with the mucin and the peritoneal implant. A cystic ovarian tumor should always be excluded in women with an appendiceal mucinous neoplasm. The correct diagnosis of appendiceal mucinous neoplasm of low malignant potential (M-LMP) is almost never made pre-operatively and is suspected intraoperatively in about a third of the cases. These cases often have a long protracted clinical course with death occurring decades after the initial diagnosis.

Death may also occur in the interim period due to unrelated causes. Death directly related to PMP in these cases is due to extensive peritoneal fibrosis with bowel obstruction and not as result of lymph node, liver or lung metastases. Appendiceal mucinous cystadenocarcinoma however, should demonstrate overt cytoarchitectural features of carcinoma including cell clusters with significant cytological atypia, stratification, papillary formation with invasion of the bowel wall. These cases are associated with an aggressive clinical course often with liver, lung and lymph node metastatic disease. [11].

PMP on the other hand can be associated with a mucinous neoplasm either in the ovary or in the appendix that demonstrate fairly bland well differentiated mucinous epithelium with minimal nuclear features of malignancy, which further confirms that the origins and nature of the parent tumors may be variable and that the cytological features are not always concordant with those of PMP further adding difficulty in determining its biological behavior and its subtype. Thus the natural history of this disease remains largely poorly defined.

Recent studies reveal that PMP is a neoplastic disease of MUC2-expressing goblet cells. Mucinous tumors of the appendix also express MUC2, which supports an appendiceal rather than an ovarian origin for PMP. This helps to distinguish PMP secondary involvement of the ovary by PMP from a primary appendiceal mucinous tumor with peritoneal implants [12]. The exact origin and mechanism of this abundant mucin production has been recently extensively studied. It is believed that PMP is

characterized by the production of MUC2, a gel forming mucin that forms strong bonds with the surrounding stroma and is also believed to have tumor suppressor activities [12].

The extracellular accumulation of mucin is attributed to an alteration in cell polarity resulting in the glycoproteins being secreted predominantly in the stroma facing surfaces of the tumor cells in contrast to adenocarcinomas wherein the glycoproteins are secreted either into the luminal surface or dispersed within the cytoplasm [13].

The evolution of treatment strategies of PMP still remains debated though the current mainstay of the treatment remains surgical extirpation of the lesion. Repeated cytoreductive surgical debulking procedures including resections of the tumor implants, omentum and obstructive bowel are common due to recurrence of the disease. Intra-peritoneal chemotherapy (5FU, Mitomycin C) has minimal benefit with reduction in the number of foci of atypical epithelium and or atrophy and degeneration of the atypical neoplastic epithelium. Based on the Sugarbaker peritonectomy procedure [14] a study by Draco *et al* showed that cytoreductive surgery with intraperitoneal hyperthermic perfusion permitted complete tumor removal, and this study confirmed the efficacy of this combined treatment in terms of improved long-term survival and better local control of the disease [15]. Indeed, the current strategy of treatment includes cytoreductive surgery combined with intraoperative hyperthermic intraperitoneal chemotherapy (mitomycin at 42 degrees C). The aim is to avoid entrapment of tumor cells at operative sites and to destroy small residual mucinous tumor nodules. This is a combined treatment with a high toxicity, particularly surgically related [16], but is associated with long term survival compared to the traditional historical treatment [17]. As endorsed by Sugarbaker, this new combined treatment should be regarded as the standard of care for epithelial appendiceal neoplasms and pseudomyxoma peritonei syndrome [18] which is best administered in an established peritoneal carcinomatosis treatment centre.

Prognosis in this disease is closely related to the bulk of the disease as evaluated by the tumor site, preoperative tumor volume and completeness of tumor removal by cytoreductive surgery and the microscopic degree of differentiation of the neoplastic epithelium as evaluated by the histopathological examination [17, 18, 19, 20]. In this context, PMP patients with pre-operative elevated tumor markers such as CEA (carcino-embryonic antigen) and CA 19-9 are at increased risk of developing recurrent disease despite aggressive therapy. Likewise, PMP patients with normal levels of these tumor markers have an overall improved prognosis [21, 22].

**Conclusion**

PMP is a condition characterized by mucinous ascites and peritoneal implants, now thought to be of appendiceal origin. Surgical debulking is the recommended treatment both for primary and recurrent tumours. The surgeons should be alert to treating mucinous neoplasm of the appendix, with special care being directed towards adequate excision and through debridement at the initial diagnosis. The periodic post operative abdominal sonography or CT scan and tumor marker laboratory test are used to monitor the disease for any tumor regrowth.Chemotherapy with 5-Flourouracil or mitomycin clearly improves survival rates.

**Figure 1: Showing the mucocele of the appendix.**

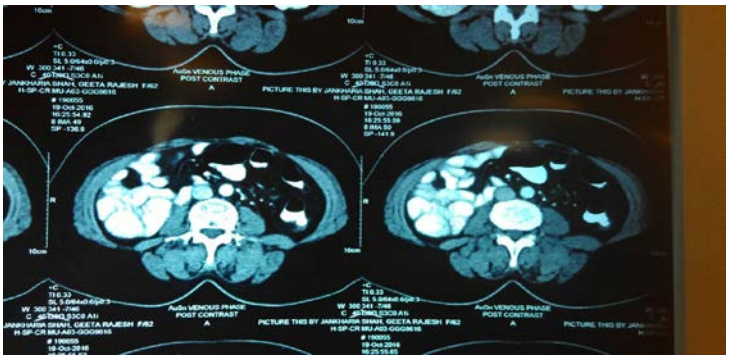
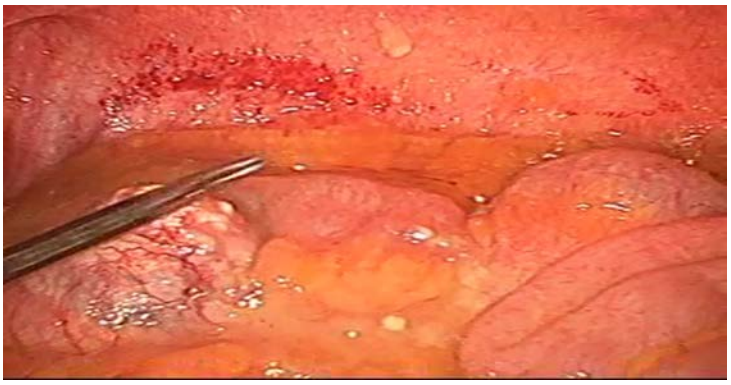


**Figure 2: Showing the jelly belly appearance.**



**References**


1. Werth R: Pseudomyxoma peritonei. Arch Gynaecol. 1884, 24: 100-118. 10.1007/BF01837425.View Article Google Scholar.
2. Fann JI, Vierra M, Fisher D, Oberhelman HA, Cobb L: Pseudomyxoma peritonei. Surg Gynecol Obstet. 1993, 177: 441-447.PubMed Google Scholar
3. Ronnett BM, Zahn CM, Kurman RJ, Kass ME, Sugarbaker PH, Shmookler BM: Disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis. A clinicopathologic analysis of 109 cases with emphasis on distinguishing pathologic features, site of origin, prognosis, and relationship to 'pseudomyxoma peritonei'. Am J Surg Pathol. 1995, 19: 1390-1408.View ArticlePubMed Google Scholar
4. Galani E, Marx GMF, Steer CB, Culora G, Harper PG: Pseudomyxoma peritonei: the 'controversial' disease. Int J Gynecol Cancer. 2003, 13: 413-418. 10.1046/j.1525-1438.2003.13318.x.View ArticlePubMed Google Scholar
5. Van Ruth S, Acherman YIZ, van de Vijver MJ, Hart AAM, Verwaal VJ, Zoetmulder FAN: Pseudomyxoma peritonei: a review of 62 cases. Eur J Surg Oncol. 2003, 29: 682-688. 10.1016/S0748-7983(03)00149-5.View ArticlePubMed Google Scholar
6. Sherer DM, Abulafia O, Fliakim R: Pseudomyxoma peritonei. A review of current literature. Gynecol Obstet Invest. 2001, 51: 73-80. 10.1159/000052897.View ArticlePubMed Google Scholar
7. Solkar MH, Akhtar NM, Khan Z, Parker MC: Pseudomyxoma extraperitonei occurring 35 years after appendectomy: a case report and review of literature. World J Surg Oncol. 2004, 2: 19-10.1186/1477-7819-2-19.PubMed CentralView ArticlePubMed Google Scholar
8. Ronnett BM, Shmookler BM, Diener-West M, Sugarbaker PH, Kurman RJ: Immunohistochemical evidence supporting the appendiceal origin of pseudomyxoma peritonei in women. Int J Gynecol Pathol. 1997, 16: 1-9.View ArticlePubMed Google Scholar
9. Szych C, Staebler A, Connolly DC, Wu R, Cho KR, Ronnett BM: Molecular genetic evidence supporting the clonality and appendiceal origin of pseudomyxoma peritonei in women. Am J Pathol. 1999, 154: 1849-1855.PubMed CentralView ArticlePubMed Google Scholar
10. Mukherjee A, Parvaiz A, Cecil TD, Moran BJ: Pseudomyxoma peritonei usually originates from the appendix: a review of the evidence. Eur J Gynaecol Oncol. 2004, 25: 411-414.PubMed Google Scholar
11. Pai RK, Longacre T: Appendiceal mucinous tumors and pseudomyxoma peritonei: Histologic features,

diagnostic problems and proposed classification. Advances in Anatomic Pathology. 2005, 12: 291-311. 10.1097/01.pap.0000194625.05137.51.View ArticlePubMed Google Scholar

12. O'Connell JT, Tomlinson JS, Roberts AA, McGonigle KF, Barsky SH: Pseudomyxoma peritonei is a disease of MUC2-expressing goblet cells. AmJ Pathol. 2002, 161: 551-564.View Article Google Scholar
13. Adsay NV, Merati K, Nassar H, Shia J, Sarkar F, Pierson CR, Cheng JD, Visscher DW, Hruban RH, Klimsta DS: Pathogenesis of colloid carcinoma of exocrine organs. Coupling of gel-forming mucin (MUC2) production with altered cell polarity and abnormal cell-stroma interaction may be the key factor in the morphogenesis and indolent behavior of colloid carcinoma in the breast and pancreas. Am J Surg Pathol. 2003, 27: 571-578. 10.1097/00000478-200305000-00002.View ArticlePubMed Google Scholar
14. Sugarbaker PH: Peritonectomy procedures. Ann Surg. 1995, 221: 29-42.PubMed CentralView ArticlePubMed Google Scholar
15. Deraco M, Baratti D, Inglese MG, Allaria B, Andreola S, Gavazzi C, Kusamura S: Peritonectomy and Intraperitoneal Hyperthermic Perfusion (IPHP): A strategy that has confirmed its efficacy in patients with pseudomyxoma peritonei. Ann Surgl Oncol. 2004, 11: 393-398. 10.1245/ASO.2004.07.002.View Article Google Scholar
16. Smeenk RM, Verwaal VJ, Zoetmulder FAN: Toxicity and mortality of cytoreduction and intraoperative hyperthermic intraperitoneal chemotherapy in pseudomyxoma peritonei-a report of 103 procedures. Eur J Surg Oncol. 2006, 32: 186-190. 10.1016/j.ejso.2005.08.009.View ArticlePubMedGoogle Scholar
17. Loungnarath R, Causeret S, Bossard N, Faheez M, Sayag-Neaujard AC, Brigand C, Gilly F, Glehen O: Cytoreductive surgery with intraperitoneal chemohyperthermia for the treatment of pseudomyxoma peritonei: A prospective study. Dis Colon Rectum. 2005, 48: 1372-1379. 10.1007/s10350-005-0045-5.View ArticlePubMed Google Scholar
18. Dhage-Ivatury S, Sugarbaker PH: Update on the surgical approach to mucocele of the appendix. J Am Coll Surg. 2006, 202: 680-684. 10.1016/j.jamcollsurg.2005.12.003.View ArticlePubMed Google Scholar
19. Sugarbaker PH, fernandez-Trigo V, Shamsa F: Clinical determinants of treatment failure in patients with pseudomyxoma peritonei. Cancer Treat Res. 1996, 81: 121-32.View ArticlePubMed Google Scholar
20. Miner TJ, Shia J, Jaques DP, Klimstra DS, Brennan MF, Coit DG: Long-term survival following treatment of pseudomyxoma peritonei: An analysis of surgical therapy. Ann Surg. 2005, 241: 300-308. 10.1097/01.sla.0000152015.76731.1f.PubMed CentralView ArticlePubMed Google Scholar
21. Sefre AF: Elevated tumor markers prior to complete tumor removal in patients with pseudomyxoma peritonei predict early recurrence. Colorectal Dis. 2005, 7: 382-386. 10.1111/j.1463-1318.2005.00773.x.View ArticleGoogle Scholar
22. 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 epithelial cancers of the appendix. J Surg Oncol. 2004, 87: 162-166. 10.1002/jso.20107.View ArticlePubMed Google Scholar.