Case Report

Pseudomyxoma Peritonei Originating from Gynecological Diseases: An Incidental Pathological Diagnosis

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Abstract

Psuedomyxoma peritonei is an uncommon disease characterized by the presence and accumulation of gelatinous material in the pelvis and abdominal cavity. It is an unusual manifestation of mucinous ascites of controversial origin, though often associated with a primary tumour of the appendix. Diagnosis of pseudomyxoma peritonei is often incidental during exploratory laparotomy because there are no specific diagnostic clinical symptoms. We report three females with gynaecological diseases with incidental finding of pseudomyxoma peritonei in the absence of appendiceal or gastrointestinal tumour. Tissue biopsies were fixed in 10% formalin and processed in paraffin wax. Representative sections were stained with haematoxylin & eosin, periodic acid Schiff, diastase and mucicarmine. Three females aged 25years, 42years and 50years presented respectively with a year, 5years and 12years history of abdominal swelling and abdominal mass. The older female also complained of vaginal protrusion with significant weight loss. Clinical diagnosis in the two younger females was advanced ovarian carcinoma, while the older female was diagnosed with third degree utero vaginal prolapse. All three had exploratory laparotomy which revealed unilateral multi lobulated left ovarian masses, with extensive mucinous peritoneal and omental deposits. There were no obvious gastrointestinal lesions and the appendices had no gross pathology. Two of the patients had oophorectomy with peritoneal and omental clearance of mucinous materials while the third patient had vaginal hysterectomy and peritoneal clearance of gelatinous deposits. The gelatinous materials showed papillary and multi locular cystic masses containing gelatinous fluid. Histopathological diagnosis of pseudomyxoma peritonei in a background of mucinous ovarian cystadenoma, and utero vaginal prolapse with epidermialization of the cervical lining epithelium was made. Pseudomyxoma peritonei occurs in the absence of a primary appendiceal disease and there are no specific diagnostic clinical symptoms. Definitive diagnosis requires histopathological analysis however, clinicians should entertain a high index of suspicion during laparotomy when mucin pools are present in the absence of gross pathology particularly in females with benign gynaecological diseases. Our cases were classed histologically into the benign disseminated peritoneal adenomucinosis type.

Keywords: Gynecological diseases, incidental diagnosis, pseudomyxoma peritonei

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Introduction

Pseudomyxoma peritonei is an uncommon disease characterized by the presence and accumulation of gelatinous material in the pelvis and abdominal cavity. It is an unusual manifestation of mucinous ascites often associated with a primary tumor of the appendix.[1,2] It is characterized by the accumulation of extracellular mucin deposits on peritoneal and epiploic surfaces by mucin-producing tumors of the appendix as demonstrated by molecular and immunohistochemical studies.[3] Other organ involvement such as the gastrointestinal system, breast, lung, urinary bladder, ovary, and uterus is thought to be a secondary seeding event.[4-6] The diagnosis of pseudomyxoma peritonei is often incidental during exploratory laparotomy for other medical conditions and the documented incidence is 2/10,000 laparotomies.[7] Pseudomyxoma peritonei as a disease entity

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was described separately by Rokitansky, Cruveilhie, and Werth, respectively, in 1842, 1848, and 1884.[8-10] It is predominantly seen in females in the fifth decade of life though some authors have documented disease in males with a male-to-female ratio of 1:3, while other studies have postulated a wide age range of occurrence.[7,11,12] We report three females with an incidental diagnosis of pseudomyxoma peritonei originating from gynecological diseases in the absence of an appendiceal tumor.

Cases Report

Three females with gynecological diseases are presented. The first was a 42-year-old, who presented with 5 years' history of abdominal swelling and abdominal mass with associated significant weight loss. She was not a known diabetic or hypertensive and had no other clinical complaints. She had no significant contributory medical history or associated remarkable symptoms. An abdominopelvic examination revealed a left mildly tender ovarian mass only. Abdominal and pelvic ultrasound revealed a left 20 cm × 10 cm × 8 cm multiloculated ovarian mass and pools of mucin, the right ovary and uterus were within normal limits. Radiological examination revealed no abnormalities in the abdominal organs, however, similar mucin pools were seen in the peritoneal cavity. Clinical and radiological diagnosis was that of an advanced ovarian carcinoma. She had exploratory laparotomy which revealed a unilateral multi-lobulated left ovarian mass, mucinous peritoneal and omental deposits. There were no gross gastrointestinal and appendiceal lesions. She had left oophorectomy with peritoneal and omental clearance of mucinous deposits. The second patient was a nulliparous 25-year-old, who presented to a peripheral hospital with 1-year history of abdominal mass and swelling. The detailed clinical history was not provided by the attending physicians who sent an oophorectomy specimen along with mucinous cystic deposits to the pathology laboratory for histopathological analysis with a clinical diagnosis of ovarian malignancy. The third female was a 50-year-old grand multiparous (P_{12+0} A5) who presented with 12 years' history of vaginal protrusion. The protrusion had no surface ulceration. There were no urinary symptoms of dysuria and frequency but she had poor urinary stream and incomplete bladder emptying. There was no vaginal bleeding or discharge. She was not hypertensive, diabetic, or asthmatic. Abdomino-pelvic examination revealed a third-degree uterovaginal prolapse, which was reducible but spontaneously protruded with Valsalva maneuver. There were no other palpable abdominal or adnexal masses. There was no abdominopelvic tenderness or stress incontinence. Systemic examination was unremarkable. Abdominopelvic ultrasound showed a 3.4 cm (AP) uterus, with preserved endometrial and myometrial echo plate complex, a bulky 14.4 cm cervix, and an extensive septate cystic lesion containing 650 ml of clear fluid arising from the left paracolic gutter and reaching the spleen. The liver, gall bladder, left kidney, loops of bowel, urinary bladder, pouch of Douglas, and adnexae were unremarkable while the right kidney showed moderate hydronephrosis. Based on the clinical diagnosis of procidentia with huge mesenteric cyst, she had vaginal hysterectomy with pelvic floor repair, exploratory laparotomy, and cyst excision.

Pathological specimens

All the tissue specimens received in the pathology laboratory were fixed in 10% formalin, processed in paraffin wax and stained with hematoxylin and eosin, periodic acid–Schiff and Mucicarmine. They comprised respectively of multiloculated

Figure 1: Gelatinous mass with granular and multi-cystic surfaces

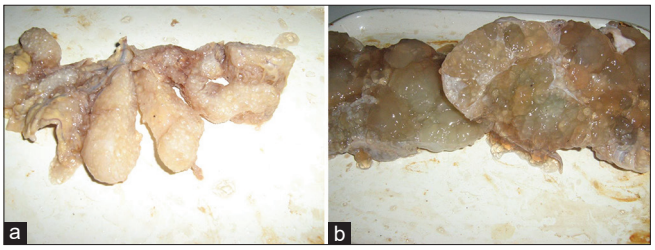

Figure 2: (a) Papillary multilocular gelatinous masses. (b) Multiloculated gelatinous masses with discreet nodules

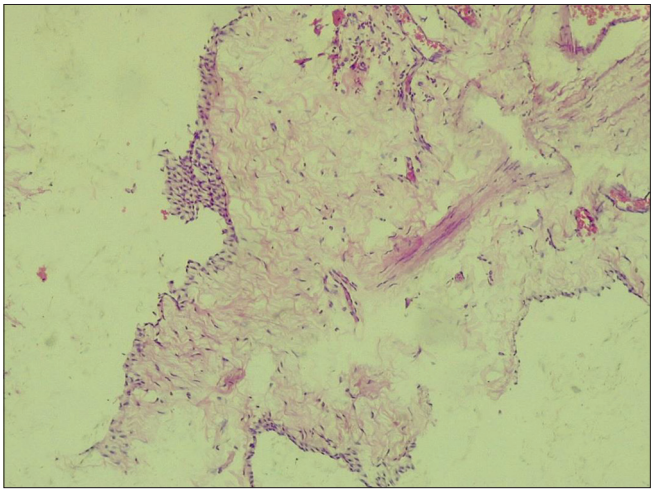

Figure 3: Ovarian cystadenoma showing focal multi-layered lining epithelium without atypia. H and E, ×100

gelatinous masses and an oophorectomy specimen. The multiloculated gelatinous masses weighed 1000 g [Figure 1]. The oophorectomy specimen weighed 425 g and measured 18 cm × 12 cm × 9 cm in dimension. Cut surfaces of the ovarian specimen were multicystic with thinned intervening septa. A microscopic examination of the ovary showed a strip of ovarian stroma containing numerous cysts lined by columnar epithelium. Within the cystic, cavities were extracellular mucin similar to the accompanying multiloculated gelatinous masses. A histopathological diagnosis of pseudomyxoma peritonei with mucinous ovarian cystadenoma was made. The second specimen was also a left ovarian mass that weighed 210 g and measured 13 cm × 10 cm × 8.5 cm in dimension. It was accompanied by a 520 g papillary multilocular cystic masses containing gelatinous fluid [Figure 2a and b]. Microscopy showed ovarian stroma and numerous cysts lined by columnar epithelium without atypia [Figure 3]. Pools of extracellular mucin were seen in the accompanying gelatinous material. A diagnosis of pseudomyxoma peritonei with mucinous ovarian cystadenoma was made. The third specimen received was a hysterectomy specimen that weighed 75 g and measured 18 cm × 5 cm × 4 cm. Cut surfaces showed a patent endometrial cavity and an unremarkable cervix. This was accompanied by a 20 g gelatinous cystic tissue with engorged veins on the surface. The cyst contained 80 ml of clear fluid. Microscopic sections from the hysterectomy specimen confirmed uterocervical prolapse with epidermalization of the cervical epithelium and chronic cervicitis. The accompanying cystic tissue was diagnosed as pseudomyxoma peritonei.

Discussion

The term "pseudomyxoma peritonei," a false mucinous tumor of the peritoneum due to the seeding of the peritoneal cavity by mucin-producing epithelium was introduced by Werth in 1884 though, this abnormal peritoneal mucinous accumulation had been previously described by Rokitansky and Cruveilhie.[8-10] It is often an incidental finding during laparotomy for various conditions and incidence is approximately 2/10,000 laparotomies with a mean age of 54–58 years at presentation and a female preponderance which may be three times higher when compared with males.[7,12] However, a few studies documented an equal sex distribution.[11] All the three patients in this report are females in the second to fourth decades of life.

Clinical symptoms of pseudomyxoma peritonei at hospital presentation are not specific and are often linked to the primary underlying mucinous disease. However, palpable abdominal mass with distension as seen in our patients has been documented in patients with pseudomyxoma peritonei.[1,13,14]

Immunohistochemical and molecular studies have documented the appendix as the most common organ of involvement and theories on its pathogenesis hinges on the rupture of underlying mucin-producing lesions such as mucocele, borderline mucinous tumors, cystadenoma, and cystadenocarcinoma with resultant spillage and accumulation of mucin within the peritoneal cavity.[3,15-17] Mucinous malignant tumors in particular have a higher propensity for rupture and spillage.[9] None of our three patients had appendiceal lesion or malignant disease. It is noteworthy that, mucinous tumors are not limited to the appendix, they also occur in the ovary and other gastrointestinal organs. Whereas, secondary ovarian involvement by pseudomyxoma peritonei occur as either a bilateral or right-sided disease with mucinous implants found on ovarian surfaces.[2,18-20] These implants may locally invade adjacent tissues such as the bowel, abdominal wall, and urinary bladder however, metastases to distant sites are infrequent. Interestingly, there is the involvement of the left ovary in two of our patients diagnosed of benign cystadenoma without atypia while the third patient had no ovarian pathology. In addition, ovarian mucinous neoplasms associated with pseudomyxoma peritonei often have gelatinous masses which distend the abdomen and may result in intestinal obstruction with or without peritonitis.[21] None of our patients had an intestinal obstruction. However, they had associated abdominal swelling with huge gelatinous masses weighing from 520 g to 1000 g. Other documented rare sites of involvement of pseudomyxoma peritonei include the fallopian tubes, breast, colon, stomach, pancreas, urinary bladder, lung, and uterus as seen in our third patient with uterine prolapse.[4-6,22]

The definitive diagnosis of pseudomyxoma peritonei can be achieved with histopathological analysis of tissue biopsy as in our cases or with cytology of peritoneal washings.[23] In spite of laparotomy findings of pools of mucin, the attending clinicians may not necessarily consider pseudomyxoma peritonei as a likely diagnosis but rather an occult malignant tumour as seen in the patients in this report. Radiological examinations such as computed tomography scan have been most useful in preoperative diagnosis but this requires expertise in interpretation because the mucinous material have a similar density to fat.[2,24]

Pseudomyxoma has been classified into two broad categories of disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis (PMC) and graded into grades 0, 1, 2, and 3 based on the degree of malignancy histologically.[4,11,25] The DPAM group is entirely benign and a grade 0 disease while the PMC group has varying histologic features of malignancy and is graded from 1 to 3. However, there may be some overlapping discordant features in each category. DPAM accounts for approximately 60% of cases and is characterized by abundant extracellular mucin containing simple or focal proliferation of mucinous epithelium with none or little atypia and little to no mitotic activity as seen in all three cases in this report. Whereas, PMC accounts for the remaining cases and histologically show abundant mucinous epithelium with cytological features of malignancy along with peritoneal lesions.

The treatment of choice is surgical debulking and clearance of mucinous deposits as done for all three patients in this report.[13,19] However, pseudomyxoma peritonei is fraught with

recurrence, though none of the patients in this report have represented following treatment. The role of chemotherapy and radiotherapy in the treatment of pseudomyxoma peritonei is limited and may be justified in the presence of underlying malignant disease.

Conclusion

Pseudomyxoma peritonei occurs in the absence of a primary appendiceal disease and there are no specific diagnostic clinical symptoms. Definitive diagnosis requires histopathological analysis, however, clinicians should entertain a high index of suspicion during laparotomy when mucin pools are present in the absence of gross pathology particularly in females with benign gynecological diseases.

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Conflicts of interest

There are no conflicts of interest.

References

1. Harshen R, Jyothirmayi R, Mithal N. Pseudomyxoma peritonei. Clin Oncol (R Coll Radiol) 2003;15:73-7.
2. Jacquemin G, Laloux P. Pseudomyxoma peritonei: Review on a cluster of peritoneal mucinous diseases. Acta Chir Belg 2005;105:127-33.
3. 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;154:1849-55.
4. Costa MJ. Pseudomyxoma peritonei. Histologic predictors of patient survival. Arch Pathol Lab Med 1994;118:1215-9.
5. Hawes D, Robinson R, Wira R. Pseudomyxoma peritonei from metastatic colloid carcinoma of the breast. Gastrointest Radiol 1991;16:80-2.
6. Kurita M, Komatsu H, Hata Y, Shiina S, Ota S, Terano A, *et al*. Pseudomyxoma peritonei due to adenocarcinoma of the lung: Case report. J Gastroenterol 1994;29:344-8.
7. Mann WJ Jr., Wagner J, Chumas J, Chalas E. The management of pseudomyxoma peritonei. Cancer 1990;66:1636-40.
8. Weaver CH. Mucocele of appendix with pseudomucinous degeneration. Am J Surg 1937;36:347-51.
9. Lanteri R, Santangelo M, Rapisarda C, Racalbuto A, Di Cataldo A, Licata A. Pseudomixoma peritonei: A case report. Cancer Ther 2009;7:31-4.
10. Werth R. Klinische und anatomische untersuchungen zur lechre von den bauchgeschwiilsten und der laparotomie. Arch Gynaecol Obstet 1884;24:100-18.
11. 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;19:1390-408.
12. Gough DB, Donohue JH, Schutt AJ, Gonchoroff N, Goellner JR, Wilson TO, *et al*. Pseudomyxoma peritonei. Long-term patient survival with an aggressive regional approach. Ann Surg 1994;219:112-9.
13. Hinson FL, Ambrose NS. Pseudomyxoma peritonei. Br J Surg 1998;85:1332-9.
14. Esquivel J, Sugarbaker PH. Clinical presentation of the Pseudomyxoma peritonei syndrome. Br J Surg 2000;87:1414-8.
15. Fernandez RN, Daly JM. Pseudomyxoma peritonei. Arch Surg 1980;115:409-14.
16. Cucinotta E, Palmeri R, Lorenzini C, Melita G, Melita P. Appendiceal mucocele. Chir Ital 2002;54:107-10.
17. Brueggen C, Baird G, Meisheid A. Pseudomyxoma peritonei syndrome of appendiceal origin: An overview. Clin J Oncol Nurs 2007;11:525-32.
18. Young RH, Gilks CB, Scully RE. Mucinous tumors of the appendix associated with mucinous tumors of the ovary and pseudomyxoma peritonei. A clinicopathological analysis of 22 cases supporting an origin in the appendix. Am J Surg Pathol 1991;15:415-29.
19. Prayson RA, Hart WR, Petras RE. Pseudomyxoma peritonei. A clinicopathologic study of 19 cases with emphasis on site of origin and nature of associated ovarian tumors. Am J Surg Pathol 1994;18:591-603.
20. Seidman JD, Elsayed AM, Sobin LH, Tavassoli FA. Association of mucinous tumors of the ovary and appendix. A clinicopathologic study of 25 cases. Am J Surg Pathol 1993;17:22-34.
21. Zoetmulder FA, Sugarbaker PH. Patterns of failure following treatment of pseudomyxoma peritonei of appendiceal origin. Eur J Cancer 1996;32A: 1727-33.
22. De Bree E, Witkamp A, Van De Vijver M, Zoetmulde F. Unusual origins of Pseudomyxoma peritonei. J Surg Oncol 2000;75:270-4.
23. Jackson SL, Fleming RA, Loggie BW, Geisinger KR. Gelatinous ascites: A cytohistologic study of pseudomyxoma peritonei in 67 patients. Mod Pathol 2001;14:664-71.
24. Walensky RP, Venbrux AC, Prescott CA, Osterman FA Jr., Pseudomyxoma peritonei. AJR Am J Roentgenol 1996;167:471-4.
25. Sugarbaker PH, Jablonski KA. Prognostic features of 51 colorectal and 130 appendiceal cancer patients with peritoneal carcinomatosis treated by cytoreductive surgery and intraperitoneal chemotherapy. Ann Surg 1995;221:124-32.