International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Pseudomyxoma Peritonei: A Rare Presentation

Dr. B. Ananda Rama Rao[1], Dr. P. Saikumar[2], Dr. J. Srikanth[2]

[1]Professor of Surgery, [2]Resident of Surgery,
SVS Medical College, Mahabubnagar, Telangana, India

Corresponding Author: Dr. B. Ananda Rama Rao

ABSTRACT

Pseudomyxoma peritonei (PMP) is an uncommon borderline neoplastic disease generally originating from a primary perforated appendiceal mucinous tumour with distinctive peritoneal spread. The presence of cells in the mucin, either inflammatory or neoplastic, distinguishes it from simple acellular mucus ascites caused by mucinous spillage. Clinically PMP presents with variety of unspecific signs and symptoms like abdominal pain, distension, ascites and even bowel obstruction. Here we discuss a case of 60 years old male with mass per abdomen since one year and pain abdomen since six months, diagnosed as pseudomyxoma peritonei with the help of imaging studies and needle aspirations. Patient was managed surgically and intraperitoneal chemotherapy was given.

Keywords: Pseudomyxoma peritonei; Neoplastic; Perforated Appendix; Mucinous tumour.

INTRODUCTION

In 1884 the term Pseudomyxoma peritonei (PMP) was coined by Dr. Werth, describing it in association with a mucinous tumour of the ovary. [1] In 1901, Frankel [2] described a case of PMP associated with a cyst of the appendix. Since these early reports there has been ongoing debate as to the primary origin of PMP, particularly in women. Pseudomyxoma peritonei is an uncommon clinical entity with an incidence of one to two in million, yearly. [3] Classically it is characterized by diffuse intra-abdominal gelatinous collections (jelly belly) with implants of mucinous material on peritoneal surfaces and the omentum. [4] Many cases are diagnosed accidentally when investigating or operating for other reasons. PMP is generally considered as benign; however it should be considered as borderline malignancy due to its progression over time, massive abdominal distension and nutritional compromise the long term survival is poor in most patients with reported 5 and 10 year survival rates of 50% and 10%-30%, respectively. [5] Here we discuss our experience in managing a patient, diagnosed to have pseudomyxoma peritonei.

CASE REPORT

A 60 years old male presented to the surgery OPD with complaints of fullness of the abdomen sine 0ne year, pain abdomen since six months and anorexia. Vitals were normal, biochemical values were within normal limits. On examination a 20x15 cms non tender non mobile mass present in the centre of the abdomen probably intra peritoneal with no signs of ascites. USG abdomen was suggestive of Hydatid cyst. On suspicion of malignancy CT scan was done which showed three liters of mucinous material with deposits in subdiaphragmatic, Morison's and pelvis and no visualization of appendix suggestive of pseudomyxoma. Mucoid material obtained on needle aspiration was sent for cytology which

suggested Adenocarcinoma. Patient was posted for surgery and Evacuation of mucinous material + greater omentectomy with debulking + right hemicolectomy was done. Irrigation with 5% dextrose intraoperatively, followed by intraoperative chemotherapy with 5-Fluorouracil (20mg/kg for five days) and Mytomycin (15mg/kg on day one only). Irrigation with Sodium bicarbonate + 2000u of heparin + potassium chloride post operatively till effluent is clear. Resected specimen i.e. omental mass, colon and portion of small bowel & mucinous material sent for histopathology. HPE showed mucin secreting cells with atypia and mucin pools in pericolic area suggestive of pseudomyxoma peritonei.

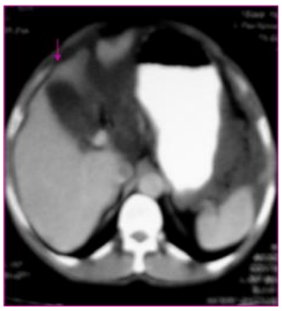
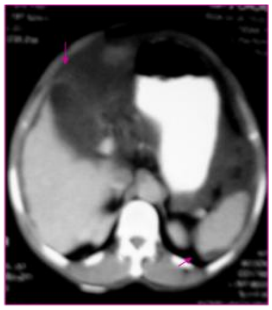
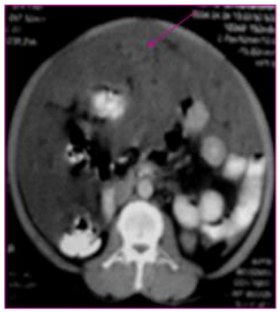


Fig-1: CT showing scalloping





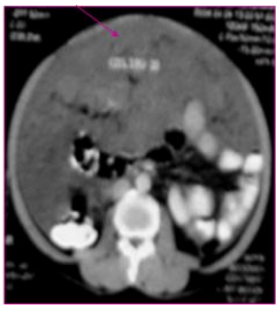


Fig-2: CT showing mucin deposits

Fig-3: mucin aspirated from peritoneum

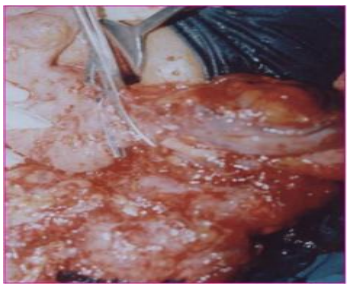


Fig-4: Omental cake

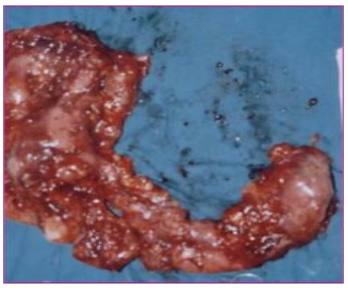


Fig-5: Resected specimen

Fig-6 Pathalogical gross specimen

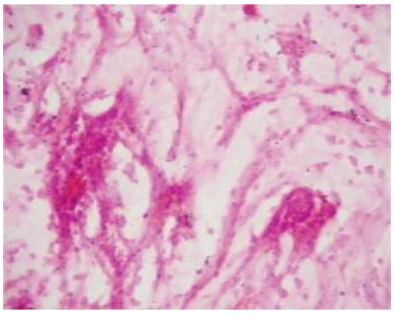


Fig-7 HPE showing mucin secreting cells

DISCUSSION

Most of the times PMP predominantly originates in the appendix in men and increasing evidence suggest a similar site of origin in females. [6,7] In recent studies MUC 2 over-expression has been suggested as a molecular marker for PMP. [8] The pathophysiology of PMP is thought to be due to progressive appendiceal adenoma growth to occlude the lumen which causes distension of the appendix by mucus and mucinous tumour cells. [3] Eventually appendix ruptures and subsequent slow leak of mucus containing epithelial cells from the adenoma occurs. The epithelial cells within the peritoneal cavity proliferate and produce large amounts of mucus. The tumour cell surfaces lack adhesion molecules, this prevents tumour cells adherence to peritoneum. The distinctive feature of PMP is its characteristic "redistribution" within the

peritoneal cavity, [9] in contrast to most carcinoma cells which implant near the site of perforation. The open lymphatic lacunae on the under surface of the right hemidiaphragm and the lymphoid aggregates in the omentum, absorb fluid, leading to bulky accumulations resulting in "scalloping" of the liver and an "omentalcake". A pathognomonic feature of PMP is the complete absence of tumour masses on the intestinal surfaces, especially the small bowel. In contrast the parts of the gastro-intestinal tract fixed to the retroperitoneum, are often heavily diseased and commonly require resection to remove macroscopic tumour involving the bowel. [10] Ronnett and colleagues classified low-grade tumours as Disseminated Peritoneal Adenomucinosis (DPAM) and high-grade tumours as Peritoneal Mucinous Carcinomatosis (PMCA), with an intermediate group (IG) demonstrating a mixture of DPAM and PMCA. Survival was significantly higher in the low-grade (DPAM) as compared with the high-grade tumours (IG and PMCA). These pathological classifications are important as they give some indication of prognosis following cytoreductive surgery (CRS) and HIPEC. Patients with low grade tumours (DPAM, MCP low grade *etc*.) appear to obtain maximum survival benefit from aggressive locoregional treatments while those with PMCA behave more like peritoneal carcinomatosis of colorectal origin. [11,12] The clinical presentation of PMP has been poorly defined due to few reports with large patient populations. The majority of patients are diagnosed during, or after, a laparotomy or laparoscopy, for suspected appendicitis, peritonitis or gynaecological cancer. Currently the optimal imaging modality for the diagnosis and staging of PMP is CT scan. [13] CT-scan findings may be pathognomonic for PMP, Typically CT appearances include areas of low attenuation, with islands of higher attenuation due to solid elements within mucinous material. Classically "scalloping" of visceral surfaces, particularly of the liver and spleen distinguishes mucinous from fluid ascites. [14] Once the peritoneal cavity is completely filled with PMP, CT-scan findings become less specific. The striking feature in most of the cases is the relative sparing of the small bowel and its mesentery or "compartmentalization" in the central abdomen by a large omental cake and massive mucinous ascites. [14] The role of MRI in staging PMP is under investigation. In summary, preoperative diagnosis could therefore be made with careful physical examination in conjunction with ultrasound and computed tomography. However, explorative laparotomy still remains the main diagnostic tool of choice. A positive finding is indicated by the presence of litres of yellowish-grey mucoid material involving both the omental and peritoneal surfaces. [15,16] The prognostic value of tumour markers in patients undergoing cryoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC) has been evaluated. Baratti et.al reported that normal preoperative CA125 correlated to the likelihood of achieving adequate CRS. Studies prove that elevated CA19.9 after surgery, or rising levels during follow-up, related to disease recurrence. [17] Reports suggest a significantly reduced recurrence-free interval for patients who had an elevated baseline CEA prior to complete cytoreduction. [18] When a patient presents with increasing abdominal girth as a result of presumed malignant ascites, laparoscopy should be performed through the midline. Ideally no lateral puncture or port sites should be used as this may result in abdominal wall tumour seeding, reducing the probability of disease eradication. [19] The indolent behavior of PMP led some to follow no active treatment, [20] however most patients with PMP untreated, will progress to terminal starvation through intestinal obstruction by mucinous ascites. [21] Prompt and aggressive treatment, including drainage of the mucus, surgical debulking of the primary and secondary tumour implants, and resection of the omentum should be instituted in each and

every patient. Commonly, a right hemicolectomy is performed on laprotomy. In order to prevent recurrence, resection of both ovaries and the appendix must be carried out in all female patients where the primary site is not found. Instillation of intraperitoneal mucolytics such as dextran sulphate, in concentrations of up to 5%, and plasminogen activators such as urokinase might be useful in preventing and treating recurrences [22] Postoperative intraperitoneal chemotherapy with 5-Fluorouracil is reasonably effective, particularly for ovarian carcinomas. [23] Intraperitoneal cisplatin and other chemotherapeutic agents have been used but with only minimal benefit. Radiotherapy of the abdomen with pelvic boost can be given in cases unresponsive to chemotherapy. Peritonectomy, omentectomy, and combination intraperitoneal chemotherapy with mitomycin C and 5-fluorouracil has been reported to achieve 10 year survival rates of up to 80%. [21]

CONCLUSION

Despite its morbid, debilitating nature with severe impact on life quality, PMP remains enigmatic. Mucin is the major contributor to the pathophysiology of PMP. As the predominant, gel-forming mucin secretor in PMP, MUC2 is responsible for the high degree of gelation and the characteristic feature of the clinical syndrome. Despite of the current standards of treatment, PMP frequently recurs, with limited treatment options. On this basis, in-depth investigations are warranted to illuminate unknown aspects of the disease and to seek novel therapeutic approaches for an enhanced treatment. Considering the substantial role of mucin in the pathogenesis of PMP, development of strategies for targeting mucin and its biology seems to be of particular significance.

REFERRENCES

1. Werth R. Klinische and Anastomische Untersuchungen Zur Lehre von der Bauchgeswullsten und der laparotomy. Arch Gynecol Obstet. 1884;84:100–118.
2. Frankel E. Uher das sogenaute pseudomyxoma peritonei. Med Wochenschr. 1901;48:965–970.
3. Smeenk RM, van Velthuysen ML, Verwaal VJ, Zoetmulder FA. Appendiceal neoplasms and pseudomyxoma peritonei: a population based study. Eur J Surg Oncol. 2008;34:196–201.
4. Moran BJ, Cecil TD. The etiology, clinical presentation, and management of pseudomyxoma peritonei. Surg Oncol Clin N Am. 2003;12:585–603.
5. Hinson FL, Ambrose NS. Pseudomyxoma peritonei. Br J Surg. 1998;85:1332–1339.
6. 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.
7. Sherer DM, Abulafia O, Eliakim R. Pseudomyxoma peritonei: a review of current literature. Gynecol Obstet Invest. 2001;51:73–80.
8. Ferreira CR, Carvalho JP, Soares FA, Siqueira SA, Carvalho FM. Mucinous ovarian tumors associated with pseudomyxoma peritonei of adenomucinosis type: immunohistochemical evidence that they are secondary tumors. Int J Gynecol Cancer. 2008;18:59–65.
9. Sugarbaker PH. Pseudomyxoma peritonei. A cancer whose biology is characterized by a redistribution phenomenon. Ann Surg. 1994;219:109–111.
10. Carmignani CP, Sugarbaker TA, Bromley CM, Sugarbaker PH. Intraperitoneal cancer dissemination: mechanisms of the patterns of spread. Cancer Metastasis Rev. 2003;22:465–472.
11. Smeenk RM, Verwaal VJ, Antonini N, Zoetmulder FA. Survival analysis of pseudomyxoma peritonei patients treated by cytoreductive surgery and hyperthermic intraperitoneal chemotherapy. Ann Surg. 2007;245:104–109.
12. Ronnett BM, Yan H, Kurman RJ, Shmookler BM, Wu L, Sugarbaker PH. Patients with pseudomyxoma peritonei associated with disseminated peritoneal adenomucinosis have a significantly more favorable prognosis than patients with peritoneal mucinous carcinomatosis. Cancer. 2001;92:85–91.
13. Jacquet P, Jelinek JS, Chang D, Koslowe P, Sugarbaker PH. Abdominal computed

tomographic scan in the selection of patients with mucinous peritoneal carcinomatosis for cytoreductive surgery. J Am Coll Surg. 1995;181:530–53.
14. Sulkin TV, O'Neill H, Amin AI, Moran B. CT in pseudomyxoma peritonei: a review of 17 cases. Clin Radiol. 2002;57:608–613.
15. Ronnett BM, Shmookler BM, Diener-West M, *et al*. Immunohistochemical evidence supporting the appendiceal origin of pseudomyxoma peritonei in women. *Int J Gynecol Pathol*1997;16:1–9
16. Buy JN**,** Malbec L, Ghossain MA, *et al*. Magnetic resonance imaging of pseudomyxoma peritonei. *Eur J Radiol*1989;9:115–8
17. Baratti D, Kusamura S, Martinetti A, Seregni E, Laterza B, Oliva DG, Deraco M. Prognostic value of circulating tumor markers in patients with pseudomyxoma peritonei treated with cytoreductive surgery and hyperthermic intraperitoneal chemotherapy. Ann Surg Oncol. 2007; 14:2300–2308.
18. Alexander-Sefre F, Chandrakumaran K, Banerjee S, Sexton R, Thomas JM, Moran B. Elevated tumour markers prior to complete tumour removal in patients with pseudomyxoma peritonei predict early recurrence. Colorectal Dis. 2005;7:382–386.
19. Sugarbaker PH. New standard of care for appendiceal epithelial neoplasms and pseudomyxoma peritonei syndrome? Lancet Oncol. 2006;7:69–76.
20. Friedland JS, Allardice JT, Wyatt AP. Pseudomyxoma peritonei. J R Soc Med. 1986;79:480–482.
21. Sugarbaker PH. Pseudomyxoma peritonei. Cancer Treat Res. 1996;81:105–119.
22. Beller FK**,** Zimmerman RE, Nienhaus H. Biochemical identification of mucus of pseudomyxoma peritonei as the basis of mucolytic treatment. *Am J Obstet Gynecol*1986;155:970–3
23. Hsieh SY**,** Chiu CT, Sheen IS, *et al*. A clinical study on pseudomyxoma peritonei. *J Gastroenterol Hepatol*1995;10:86–91.

How to cite this article: Rao BAR, Saikumar P, Srikanth J. Pseudomyxoma peritonei a rare presentation. Int J Health Sci Res. 2018; 8(2):304-308.

****