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Case Report

Pseudomyxoma Peritonei of Unknown Origin: A Case Report

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Abstract

Pseudomyxoma peritonei (PMP) is a rare condition characterized by disseminated intra-abdominal collection with mucinous implants on peritoneum and omentum. Although appendix has usually been implicated as the primary site some reports found no cause. This case also describes a PMP with no identifiable primary site. A 48 year old male presented with right hypochondrial pain, fever and abdominal distention underwent laparotomy, found to have intra peritoneal mucoid materials with septated ascites. Imaging techniques also failed to identify the primary site. Biopsy confirmed PMP. Because of advanced intra peritoneal spread and dense adhesion no other surgical interventions attempted. His past history was remarkable for choledocholithiasis and repeated surgical intervention.

Keywords: *Pseudomyxoma Peritonei; Surgery; Ascites*

Introduction

Pseudomyxoma peritonei (PMP) is a rare, diagnostically challenging and poorly understood disease. The incidence of PMP was reported as one to two per million per year [1]. It was first described by Werth in 1884. He observed massive intra peritoneal accumulation of gelatinous pseudomucin from ovarian cystadenoma [2]. But recent studies confirmed generally it arise from a perforated epithelial tumour of appendix [3]. In the past, there has been much discussion regarding the definition and pathology of PMP. It is characterized by disseminated intra-abdominal collection with mucinous implants on the peritoneum and omentum [3-5]. PMP was regarded as wide spectrum of neoplasm from benign and borderline to malignant lesion [6]. However, its behaviour shows that it should be regarded as borderline malignancy [3]. Recent studies define PMP as an intestinal grade I mucinous adenocarcinoma that arise from primary adenoma [7].

Symptoms of PMP are non-specific which makes late clinical presentation and diagnostic challenge. Routine laboratory studies are seldom helpful in making the diagnosis. This makes the patient to get long term health deterioration before actual diagnosis is made [4,5]. Therefore, incidental diagnosis during laparotomy are common [4,7]. Treatment consists of complete tumour excision through radical debulking surgery but optimal management remains controversial [4].

This report presents a case of patient with PMP presented with abdominal distention and review of the current literature.

Case Report

A 48 year old gentleman presented with right hypochondriacal (RHC) pain and fever for 3 days duration. Abdominal distention was present in last 2 weeks. Patient has history of vomiting and stool not passed for last two days. There were no other symptoms. He had a history of significant surgical intervention in the past. He underwent open cholecystectomy and common bile duct (CBD) exploration for multiple gall bladder and CBD calculi 10 years age. Repeated Endoscopic Retrograde Cholangio-Pancreatography (ERCP) was done to

extract residual CBD calculi. In addition, open CBD exploration and stone extraction was also done. The patient denied significant alcohol consumption. On presentation patient was looks unwell, icteric, pulse rate was 110/ min and blood pressure was 110/70. Abdominal examination revealed tenderness at RHC region and abdominal distention consistent with ascites. Bowel sound was present and rectum loaded with soft faeces. No signs of chronic liver cell disease.

Regarding the blood investigations, we observed elevated level of total bilirubin (10.42 mg/dl) with high indirect bilirubin (7.02 mg/dl). Alkaline phosphatase level was 142 u/l (53 - 128 u/l). Elevated Carcino Embryonic Antigen (CEA) level (76.1 mg/ml) was also noted. Complete blood count and the other routine biochemical analysis were in normal range. Abdominal Ultrasonographic findings of the patient were reported as large septated cystic and solid areas seen in abdomen suggestive of septated ascites. Computed Tomography (CT) revealed ascites and soft tissue density mass seen in omentum and mesentery compatible with PMP. Prominent CBD with mild intra hepatic duct dilatation was noted. The appendix was normal.

During exploratory laparotomy, peritoneal cavity was filled with semi liquid gelatinous mucoid materials which made as a mass lesion in the RHC region with difficult access (Figure 1). Multiple adhesions were noted between intra-abdominal and intra pelvic structures. Biopsy was taken from implants on greater omentum and peritoneal thickening. Appendix looks normal in the ileo caecal region. Peritoneal cavity was irrigated and closed. Patient recovered uneventfully and was discharged on seventh post-operative day.

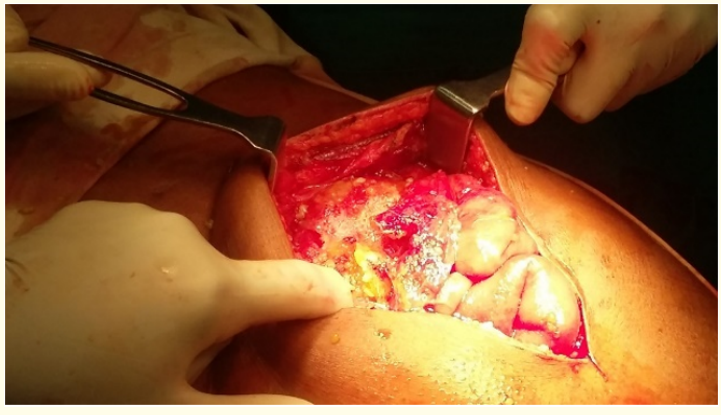

Figure 1: *Peritoneal cavity was filled with gelatinous mucoid materials which made as a mass lesion in the RHC region.*

Histological examination revealed abundant mucin arranged within fibrotic nodules. Inflammation and micro calcification were present. Associated mucinous carcinoma was not evident. The findings were compatible with PMP - low grade. Oncology team decided to start on systemic chemotherapy.

Discussion

PMP is a rare, chronic and relapsing disease [4]. It is most prevalent in women aged 50 - 70 years [7]. PMP should be considered as a spectrum of disease, ranges from mucinous ascites to frank mucinous adenocarcinoma. In addition, intestinal mucinous tumour, particularly colorectal cancer or any mucinous neoplasm may present with clinical, radiological and pathological features resembling PMP [7]. In the past, there has been much discussion regarding the definition, pathology, site of origin and prognosis of PMP [8]. It was believed that most cases of PMP originate from ovarian tumour. This believe has been challenged by recent immunohistochemical and molecular genetic studies which showed these tumours are secondary to appendiceal tumour in both men and women [9,10].

Sugarbaker., *et al.* defines PMP as a grade I mucinous adenocarcinoma that arise from a primary appendiceal adenoma [11]. In most cases appendix has been identified as the primary site of origin [12]. The literature survey showed many other sites of origin including ovary, fallopian tube, colorectum, small bowel, stomach, gall bladder, lung and pancreas [2,4,5,7,12]. Primary site of origin was unknown in some cases [12].

PMP is a slowly progressive disease. The initial growth of an appendiceal adenoma occludes the appendiceal lumen. Distension of the appendix by mucus and mucinous tumour cells occurs. The appendix eventually ruptures because of intra luminal pressure, which leads to leakage of mucus containing neoplastic cells and spread throughout the abdominal cavity. In most cases appendicular perforation is an occult event. The epithelial cells within the peritoneal cavity continue to proliferate producing large quantities of mucus. Because of lack of cell adhesion molecules and surrounding mucus the tumour cells move with the normal flow of peritoneal fluid. The distinctive feature of PMP is its characteristic "redistribution" within the peritoneal cavity. The intra peritoneal distribution of PMP is determined by movement and absorption of peritoneal fluid and gravity. 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 as the mucus is absorbed and epithelial cells "filtered out" and concentrated which result "scalloping" of the liver and an "omental cake". Gravity also plays a role as mucus and cells concentrate in dependent portions of the abdomen and pelvis such as the recto-vesical pouch, the right retro-hepatic space and the paracolic gutters. As the disease progresses and becomes generalized redistribution extends to the left hemi diaphragm, engulfs the spleen and stomach. Finally, it spreads throughout the peritoneal cavity. The resultant gastrointestinal tract compression eventually leads to mechanical and functional gastrointestinal obstruction [3].

A combination of lack of specific symptoms, slow and occult clinical course of tumour leads to delay in diagnosis. Acute presentation during advanced disease are common and along with nonspecific symptoms [7,12]. Reported symptoms are abdominal distention, abdominal pain, palpable mass, weight loss, intermittent bowel obstruction, haematuria and urinary retention [5,12]. In addition, studies show 27% presented with suspected appendicitis, 23% with increasing abdominal distention and 14% with new onset hernia [3]. Abdominal distension is caused by multifocal peritoneal, serosal and omental implants associated with copious amount of intraperitoneal mucin [5]. Inflammatory changes due to tumour implants can lead to fistula formation or adhesion which favours intermittent or chronic partial bowel obstruction [5].

Physical examination and laboratory tests are non-discriminative [12]. Some degree of anaemia maybe presents [2]. Elevated level of Carcino Embryonic Antigen (CEA) and Carbohydrate Antigen 19-9 (CA 19-9) were present in 42% of patients with PMP. But both were non disease specific [12]. Elevated level of CEA and CA 19-9 preoperatively was observed to be associated with increased risk of recurrent disease even after aggressive surgical intervention [5]. In addition, it can be used for follow up evaluation of patient after intervention [12]. Recent studies show the usefulness of imaging techniques in diagnosis of PMP. Plain film of abdomen shows that central displacement of the bowel with obliteration of the psoas border when abdomen is distended with mucin. Widely disseminated calcific lesions can also be seen occasionally. Features of bowel obstruction present at advanced disease condition [7]. Ultrasound scan abdomen also valuable which shows echogenic masses with ascites, multiple septation and scalloping of liver [13-16]. CT is the optimal imaging modality for diagnosis and staging of PMP. Characteristic features are posterior displacement of the intestine with numerous low density masses and calcification, diffuse peritoneal infiltration appearing similar to ascetics with septated fluid pockets filling the peritoneal cavity and scalloping of visceral surfaces particularly the liver and spleen [3,7].

WHO published a classification system for PMP into low and high grade. This categorization criteria correlate with prognosis [17]. In addition, it also described in three diagnostic criteria. Low grade tumour was described as Disseminated Peritoneal Adenomyosis (DPAM) including histologically benign peritoneal lesion, High grade tumour as Peritoneal Mucinious Carcinomatosis (PMCA) with pathological features of adenocarcinoma and condition with intermediate features between DPAM and PMCA. There was a statistically significant difference in survival between patient and DPAM and PMCA with intermediate group [3,8].

Optimal treatment for PMP is still controversial [12]. Untreated patient with PMP will develop terminal starvation through intestinal obstruction by mucinous ascites [3]. Most studies recommend a complete cytoreduction with radical peritonectomy and resection of involved organ followed by perioperative intra peritoneal chemotherapy. This procedure showed prolonged survival at the cost of morbidity. Most of the time repeated intervention is necessary for management. But each intervention is associated with morbidity

and mortality, which further debilitate the patient condition [12]. Adhesions following laparotomy are surrounded by recurrent tumour, which facilitates infiltration of inflammatory cells and neovascularization leads to dense adhesion. This makes surgical procedure more difficult [2]. In the case of advanced intra peritoneal spread, aggressive surgical procedure does not produce complete cytoreduction. Instead, it increases the morbidity. During this instance management plan should focus on symptomatic treatment and preservation of function. Intra peritoneal chemotherapy is not recommended for early stage of PMP with low grade malignant histology [12]. Response to systemic chemotherapy was transient and primarily recommended for patient with extensive peritoneal disease and high grade cyst-adenocarcinoma [5]. Sugarbaker., *et al.* introduced and popularized the approach combining Cytoreduction Surgery (CRS) with Heated Intraperitoneal Chemotherapy (HIPEC). CRS is to clear the macroscopic tumours and HIPEC for residual microscopic disease [3]. Studies show that median survival of patient was approximately 2 years who underwent repeated surgical procedures [2]. Major prognostic factors are stage of local invasion throughout the peritoneal cavity, histological grade of causative lesion and completeness of cytoreduction [5,12]. Death is usually secondary to intestinal or biliary obstruction, fistula formation, peritonitis or pulmonary embolism but not due to visceral invasion or metastatic disease [2].

Lack of specific symptoms and signs make the presentation and diagnosis late. Imaging techniques are helpful in diagnosis. Therefore, PMP must be considered as a differential diagnosis in patient presenting with abdominal distention and ascites.

Conclusion

Pseudo myxoma peritonei is a rare condition and usually present as an acute abdomen. This condition is diagnostically challenging one in surgical practice. The management or optimal treatment for pseudo myxoma peritonei is still controversial. The complete cytoreduction with radical peritonectomy' omentectomy and resection of involved organ and followed by chemotherapy prolonged survival rate with morbidity.

Bibliography

1. Smeenk RM., *et al*. "Appendiceal neoplasms and pseudomyxoma peritonei: a population based study". *European Journal of Surgical Oncology* 34.2 (2008): 196-201.

2. T Yilmazlar., *et al*. "Pseudomyxoma peritonei: a case report and current concepts in the literature". *Techniques in Coloproctology* 3.3 (1999): 157-160.

3. Bevan KE., *et al*. "Pseudomyxoma peritonei". *World Journal of Gastrointestinal Oncology* 2.1 (2010): 44-50.

4. Maria Homeag., *et al*. "Pseudomyxoma Peritonei". *Current Health Sciences Journal* 35.3 (2009): 193-196.

5. Hamid AG., *et al*. "Treatment of Pseudomyxoma Peritonei: A Case Report". *Journal of Cancer Prevention and Current Research* 2.4 (2015): 00044.

6. Ioannidis O., *et al*. "Pseudomyxoma retroperitonei: report of 2 cases and review of the literature". *Revista Española de Enfermedades Digestivas* 104.5 (2012): 268-275.

7. S Jivan and V Bahal. "Pseudomyxoma peritonei". *Postgraduate Medical Journal* 78.917 (2002): 170-172.

8. Ronnett BM., *et al*. "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"". *American Journal of Surgical Pathology* 19.12 (1995): 1390-1408.

9. Sherer DM., *et al*. "Pseudomyxoma peritonei. A review of current literature". *Gynecologic and Obstetric Investigation* 51.2 (2001): 73-80.

10. Ronnett BM., *et al*. "Immunohistochemical evidence supporting the appendiceal origin of pseudomyxoma peritonei in women". *International Journal of Gynecological Pathology* 16.1 (1997): 1-9.

11. Sugarbaker PH. "Pseudomyxoma peritonei". *Cancer Treatment and Research* 81 (1996): 105-119.

12. AK Agrawal., P *et al*. "Pseudomyxoma peritonei originating from urachus-case report and review of the literature". *Current Oncology* 21.1 (2014): e155-165.

13. Lawate PS., *et al*. "Computed tomography and sonographic features of pseudomyxoma peritonei". *American Journal of Roentgenology* 152.2 (1989): 429.

14. Seshul MB and Coulan CM. "Pseudomyxoma peritonei: computed tomography and sonography". *American Journal of Roentgenology* 136.4 (1981): 803-806.

15. Foster DR. "Ultrasound findings in pseudomyxoma peritonei". *Australasian Radiology* 29 (1985): 39-41.

16. Yeh HC., *et al*. "Ultrasonography and computed tomography in pseudomyxoma peritonei". *Radiology* 153.2 (1984): 507-510.

17. Carr NJ., *et al*. "Pathology and prognosis in pseudomyxoma peritonei: a review of 274 cases". *Journal of Clinical Pathology* 65.10 (2012): 919-923.

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