ORIGINAL ARTICLE

PSEUDOMYXOMA PERITONIE IS A RARE CLINICALLY CHALLENGING ENTITY: REVIEW OF THREE CASES

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ABSTRACT

Background: Pseudomyxoma Peritonei is a rare disease. It is identified operatively by gelatinous ascites accompanied by mucinous material in the peritoneum, usually arising from appendix and ovary. Other sites from where it may arise are colon, gallbladder, pancreas or stomach. The clinical presentation is inconsistent and preoperative diagnosis is often difficult. The focus of this study is to consider the clinical, histopathological aspects and survival of pseudomyxoma-peritonie in a tertiary care hospital of Karachi.

Methods: A single center Ziauddin University and hospital, histopathology database from 2006-2016 was looked up retrospectively for the incidence of pseudomyxoma-peritonie on biopsy and reviewed patient's record. All cases operated on the basis of clinical presentation and radiological findings. Diagnosis made incidentally on histopathology, originated from three different sites.

Results: Total 1388 appendectomies were performed, 749(53.96%) were male, and 639(46.03%) female. Carcinoid tumor was present in 11(0.79%) and 3(0.21%) were of pseudomyxoma-peritonie variety. In the later 3, two were males and one was a female with ages in 42, 85, 52 years respectively. All three-patients came to the emergency with variable presentation. The preoperative diagnosis was of a perforated appendix, large mesenteric cyst and in the third-case a large cystic-cum-solid abdominal mass. Histopathology confirmed pseudomyxoma-peritonie. The primary sites diagnosed were appendix, pancreas and ovary.

Conclusion: Pseudomyxoma-peritonie is a challenging task to diagnose clinically. On the basis of case series, this intra-peritoneal tumor has varied presentation. Patients usually present in late stage of disease with mucinous tumor cells implanted in the peritoneal mesothelium and in clinically apparent ascites.

KEY WORDS: Pseudomyxoma-peritonie, Mucinous tumor, Appendiceal tumor, Peritoneal carcinomatosis, Rare entity

INTRODUCTION

Pseudomyxoma-peritonei is an infrequent[1] misdiagnosed disease. Globally incidence is 1-2 million/year with varied and slightly perplexing presentations which includes abdominal distention and sometime acute appendicitis[2,3]. Pseudomyxoma-peritonei is distinguished by the collection of profuse mucinous jelly like material in the peritoneal cavity accompanied by a related mucinous tumor of ovarian or gastrointestinal tract origin [4]. Herein, we report a collective case series of pseudomyxoma-peritonie, in one of the above mentioned organs.

A 42-years-old male presented to emergency department with two-day history of severe abdominal pain, slightly increased abdominal girth, vomiting and fever. No significant past medical and surgical history. On physical examination he had a distended abdomen and mild tenderness in lower abdomen. On laboratory findings he had highly raised white cell count and low hemoglobin. Ultrasound showed mild ascites. X-ray abdomen revealed multiple air fluid levels. CT scan with contrast showed specks of air close to appendix, raising the possibility of appendiceal rupture or abscess, Bilateral pleural effusion with consolidation and collapse of right lung was also seen. Exploratory laparotomy, omentectomy and appendectomy were performed. Intra-operatively jelly like material was found filling the peritoneal cavity, Other finding showed adherence of small and large bowel to the

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omentum and abdominal wall with mild ascitic fluid and an inflamed appendix. Recoverey was uneventfull and patient was discharged. Histopathology was consistent with well-differentiated mucinous colloid adenocarcinoma of appendiceal origin with intra-peritoneal metastatic pseudomyxoma-peritonie. The patient remained asymptomatic for approximately three-months after surgery, than he had recurrent abdominal pain. Re-investigation revealed recurrence of tumor. Chemotherapy was commenced with Foxfol-4 (Flurouracil, Leocovorin calcium, Oxaliplatin) but patient had a partial response with slightly decrease in the bulk of the tumor in six-months after commencing chemotherapy. After this the patient refused for further treatment. He is alive with recurrent disease but not well as he was assessed on follow up after his first surgery. Second patient is an 85-year old male who presented to the emergency department with six-day history of lower abdominal pain radiating to lower back with generalized weakness and three-day history of relative constipation. His past medical history included hypertension. On physical examination he had a distended abdomen, and a hard non tender mass palpable in lower abdomen extending to the epigastrium. Shifting dullness was positive. Mass was also confirmed on per rectal examination. Laboratory findings only showed low hemoglobin. CT scan (abdomen and pelvis) with contrast showed cystic mass in pelvis extending in epigastrium, displacing adjacent bowel loops, anteriorly abutting abdominal wall, posteriorly abutting aorta, coeliac vessels, inferiorly urinary bladder , no infiltration in any viscera and vessels, there was also a left inguinal hernia and moderate ascitic fluid. Patient was operated for mesenteric cyst evacuation and excision. Intra-operative huge cyst contained jelly like material with multiple peritoneal deposits of mucin. The obstruction appeared as a result of disseminated mucinous tumor cells and ascites. Following surgery, the patient had a normal clinical course until the fourth-postoperative day when he had myocardial infarction and also a chest infection. He was shifted to ICU but unfortunately passed away on sixth-postoperative day because of severe chest infection, myocardial infarction, malignancy and advanced age. Histopathological examination and immunohistochemistry raised a strong possibility of gastrointestinal origin pseudomyxoma-peritonie, with peritoneal metastasis. Patient had intestinal obstruction and underwent emergency surgery. The urgent nature of surgery in elderly patient was the reason for poor outcome.

The third-case is a 52-years-old post menopausal woman who presented to the emergency department with one-week history of crampy lower abdominal pain, incessant vomiting, increased abdominal girth, peripheral edema, and four-day history of absolute constipation. She had history of hypertension and past surgery of appendectomy and bilateral multiple ovarion cyst four-years prior to presentation. Patient had borderline atypical proliferative mucinous-cystadenocarcinoma according to last histopathology report. Chemotherapy not administered because of financial reasons. Physical examination showed a distended abdomen with generalized tenderness, shifting dullness or fluid thrill was present. No lymphadenopathy was present. Mass was confirmed on pelvic and per rectal examinations. Laboratory data showed a highly increased white blood cell count with normal hemoglobin and hematocrit values. Complete liver and renal function tests were in normal limits. The patient was dehydrated and malnourished with low sodium, increase serum urea nitrogen. Radiologic studies suggested intestinal obstruction. CT scan without contrast showed medium multiple tubular, fluid-filled masses in the pelvis, calcification along wall of the mass and air bubbles within mass. Patient optimized and planned for emergency exploratory laparotomy. Intraoperative finding showed peritoneal cavity filled with mucinous material, massive ascites, giant solid-cum-cystic mass adhered to right adnexa, omentum, abdominal wall and sigmoid colon. After vigilant dissection, necrotic small bowel was identified. Small bowel originated mass identified, resection of mass with bowel, ileostomy and mucus fistula was made, right oophorectomy, omentectomy, and debulking laparotomy performed. Patient remained unwell with unsatisfactory condition on operative-day. On first-postoperative day she passed away. Her cause of death was considered to be circulatory failure due to significant ascites and a large tumor. Patient had a borderline atypical proliferative mucinous-cystadenocarcinoma on histopathology. It was further augmented with immunohistochemical studies revealed previous ovarian cyst to be a borderline mucinous tumor. The present biopsy was an invasive malignant mucinous-carcinoma presumably primary in the ovary. There was a probability of omental metastasis emerged in the gastrointestinal tract. Synchronous tumor described in the previous studies as two unrelated main sites like ovary and gastrointestinal tract tumor.

METHODS

We described three-cases from a single tertiary care center in the department of General Surgery Ziauddin University and Hospitals. Analysis started from the standpoints of histopathology.

Database searched retrospectively for the incidence of pseudomyxoma-peritonie in biopsy and reviewed patient's record. A ten-year period of 2006-2016 was considered and found only three-cases.

RESULTS

Total 1388 appendectomies performed, 749(53.96%) were male, 639(46.03%) were female.

Carcinoid tumor was present in 11(0.79%) and 3(0.21%) were diagnosed as a pseudomyxoma-peritonie malignancy (Table 1). In pseudomyxoma-peritonie two were males, one was female with ages in 42, 85, 52 years respectively (Tale 2). All three-patients came in the emergency with variable presentation, operated for perforated appendix, mesenteric huge cyst and large cystic plus solid abdominal mass respectively. All three-cases of pseudomyxoma-peritonie diagnosed incidentally on histopathology, originated from three-different sites (Table 2). One-patient recovered postoperatively and remained asymptomatic for three-months after surgery but he had recurrence, while two-patients expired on their postoperative course. All three-patients presented in a very advance stage disease.

Table 1: Gender distribution with percentage

	Male = n(%)	Female = n(%)	Total = n(%)
Appendecotomics	749(53.96%)	639(46.03%)	1388
Carcinoids	5(45.45%)	6(54.54%)	11(0.79%)
Pseudomyxoma peritonie	2(66.66%)	1(33.33%)	3(0.21%)

Table 2: Primary site of origin of tumor with age and gender

Pseudomyxoma peritonie	Age (years)	Gender	Primary site of origin
	42	Male	Appendix
	52	Female	Ovary
	85	Male	Gastrointestinal tract/ Pancreas

DISCUSSION

In previous studies there were several single case reports instead of case series therefore it is hard to distinguish a specific presentation of the disease. In recent times there has been a lot of debate over the exact origin of the tumor. Limited research has been done in this area which suggested ovaries as the main site in females. In contrast some supports the peritoneum other go for gastrointestinal tract. However these tumors can be malignant form their initial manifestation and can present as atypical cells and epithelial proliferation[5-7]. Pseudomyxoma-peritonie is distinguished by intraperitoneal collection of mucinous fluid or ascites produced by malignant cells [8]. Most often it found unexpectedly in advanced stage and with a possible devastating complications. Studies showed 75% female patients [9,10]. But in our collective series we had male dominancy. The appendix, gastrointestinal tract and ovary are frequent primary sites of pseudomyxoma-peritonie [6,9-15]. Additional sites are lungs, common bile duct, stomach, pancreas, urachus, uterus and fallopian-tubes [16-21]. In females 44% showed ovarian pseudomyxoma-peritonie. Gastrointestinal origin mostly come from appendiceal mucinous-adenoma [2,6,10,21] The abdominal distension is a common sign especially in advanced stage disease due to ascites, along with ovarian or appendicular mass [2 ,10, 22, 23]. Few studies suggested that presentation and appearance of this disease had obvious histopathological findings [6, 24-26]. Debulking laparotomy in symptomatic patients is done which is not therapeutic. The only purpose to reduce bulk of disease is to reduce pressure effects due to mucus accumulation. Recurrent disease needs redo or more aggressive surgery.

CONCLUSION

On the basis of case series, this disease is a challenging task to diagnose preoperatively. Pseudomyxoma-peritonie presentation is varied and non specific. Patients usually present in late stage of disease from mucinification of the peritoneal mesothelium and mucinous tumor cells remains in the ascites and increase the chances of recurrences.

Conflict of Interest: None

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