CASE REPORT

PSEUDOMYXOMA PERITONEI: A RARE CLINICAL ENTITY

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ABSTRACT

Pseudomyxoma peritonei is a rare clinical condition, arising from mucinous tumour of the appendix and occasionally from the ovary. The clinical presentation is caused by accumulation of secretion of MUC-2 secreting cells in the peritoneal cavity with no place of drainage. Plain films of the abdomen and ultrasound have role in diagnosis but CT scan is the modality of choice. Cytoreductive surgery and perioperative chemotherapy have promising results. We are presenting a case of incisional hernia, turned out as rare clinical entity of pseudomyxoma peritonei with failure to recognize the primary site of origin due to lack of previous record of surgery.

Key words Pseudomyxoma peritonei, Mucinous adenocarcinoma.

INTRODUCTION:

Pseudomyxoma peritonei has a reported incidence of approximately one per million per year and clinically present at laparotomy with 'jelly belly'.[1] Commonly it arises from mucinous tumour of the appendix and occasionally from the ovary, colon, rectum, stomach, gall bladder, bile duct, small intestine, urinary bladder, lung, breast, pancreas, fallopian tube.[2] Sometimes this condition arise from retroperitoneal tissues as well, which is known as pseudomyxoma extraperitonei.[3] We are presenting a case of pseudomyxoma peritonei as an incidental finding in a patient, who presented as a case of incisional hernia.

CASE REPORT:

A 45 years old female presented in surgical outpatient department with complaint of swelling in left iliac region for the last one and half year. The swelling first noticed after three months of third surgery. It was initially smaller in size, which has gradually increased. It was reducible and cough impulse was positive. There was no associated history of fever, haemoptysis, haemetemesis, melena, altered bowel habit, jaundice and cough. There was a significant history of generalized body weakness and lethargy. The past surgical history of the patient revealed that she has been operated three times with no proper record of surgery. According to her attendant the first surgery was hysterectomy, second was laparotomy with colostomy and third one was closure of colostomy. The first two surgeries were done three years back, while the third surgery was done eighteen months back. It was the colostomy closure site where an incisional hernia developed.

On examination she was well built, orientated and cooperative. Her vitals were normal. She had pallor but jaundice and cyanosis were absent. Lymph nodes were not palpable. Examination of the abdomen revealed slight distension with previous surgical scar marks in the lower abdomen. On coughing a swelling appeared in the left iliac region which was reducible. This defect in the anterior abdominal wall was admitting the tip of three fingers. Abdominal examination revealed no mass, fluid thrill and shifting dullness. Examination of the respiratory, cardiovascular and nervous system was also normal. Laboratory investigations showed that the Hb% of 10.6 gm/dl, TLC 8000/ mm^3 , blood sugar 7.5 m.mol/l, urea 2.3 m. mol/l , creatinine 74 m.mol/l, while HBSAg and Anti-HCV were non reactive. Urinary analysis and chest radiograph were normal. Ultrasonography was reported as normal. Finally a diagnosis of incisional hernia was made.

Abdomen was opened via previous colostomy closure scar mark. Hernial sac was identified and opened. On opening peritoneal cavity jelly like fluid admixed with blood protruded out of abdomen. Approximately one large kidney tray of fluid

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removed. Incision was increased and it was seen that peritoneum was studded with thick nodules; some of them were full of jelly like material and so was greater omentum. Small intestine was spared but pelvic cavity was involved. Appendix and ovaries were not visualized due to frozen structures. Peritoneal haemostasis secured and wound closed. The entire resected specimen was sent for histopathological examination. Postoperative recovery of the patient was uneventful. The histological report revealed that it was an adenocarcinoma- mucus secreting type. Finally diagnosis of pseudomyxoma peritonei was established. Patient was sent to oncology department for further management.

DISCUSSION:

The term pseudomyxoma peritonei has been used in reference to any condition, benign or malignant, in which the peritoneal cavity is filled with gelatinous substance.[4] Sugarbaker and colleagues defined the pseudomyxoma peritonei as a pathologically and prognostically homogenous group of cases characterized histologically by benign peritoneal tumour that are frequently associated with an appendiceal mucinous adenoma. Cases of peritoneal carcinomatosis, regardless of the presence of abundant extra cellular mucus production, are excluded from this precise definition of pseudomyxoma peritonei.[5]

Ronnet and colleagues categorized the abdominal mucinous tumour presenting as pseudomyxoma peritonei into three broad categories 1) adenomucinosis 2) a hybrid group 3) mucinous adenocarcinoma. The prognosis with regard to recurrence and survival was dependent on the histopathological type. Patients with adenomucinosis have an approximately 80%, 10 years survival.[6] In our patient the histopathological findings showed that it was adenocarcinoma-mucus secreting type. There was an abundant accumulation of mucus in the peritoneal cavity. All these findings suggestive of pseudomyxoma peritonei.

Recent research indicates that in the pseudomyxoma peritonei clinical symptoms may be caused by an over whelming increase in Muc-2 secreting cells as well as the fact that the excessively produced mucin has no place to drain.[7] The origin of pseudomyxoma peritonei is controversial. Clinical morbidity and mortality result from the fact that copious amount of extracellular and peritoneal mucin results in distortion and loss of function of visceral organs.[8]

Currently cytoreductive surgery combined with perioperative intraperitoneal chemotherapy is the standard treatment for the patient with peritoneal spread of primary appendiceal tumour.[9] In our patient pseudomyxoma peritonei was an incidental finding with obscure primary due to the fact that patient had no previous record so we did debulking surgery and sent the patient to oncology department for further management..

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