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Pseudomyxoma Peritonei: A Rare Case Report with Review of Literature

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Abstract:

Pseudomyxoma peritonei is rare, chronic, relapsing, diagnostically challenging disease which is characterized by disseminated mucinous ascites. The presence of inflammatory or neoplastic cells in mucin distinguishes it from simple ascites. It is mainly due to complication of borderline or malignant neoplastic lesion of appendix and/or ovary. The clinical outcomes vary in different histological types and corresponding treatment modalities. Most of the clinicians think that surgical debulking with appendectomy is the line of treatment but this is still controversial as the role of chemotherapy is still being evaluated. We report a case of 50 years female and review unique clinical features and the misconceptions surrounding pseudomyxoma peritonei.

Key words: Appendectomy, Ascites, Neoplasm Recurrence, Peritoneal Neoplasms, Pseudomyxoma Peritonei.

Introduction

The syndrome pseudomyxoma peritonei is very rare and frequently misdiagnosed condition with a reportable incidence of 2/10000 of all laparotomies [1]. It has variable clinical presentations like abdominal distension, ovarian tumors or appendicitis like syndrome [2]. It was first described by Werth in 1884. He described that the accumulation of gelatinous pseudomucin in the peritoneum is due to perforation of ovarian cystomas [3]. But later it was proved to be arising more commonly from appendiceal tumor [4]. Diagnosis always requires clinical suspicion followed by commuted tomography and histopathological findings [3].

Case Report

A 50 years female presented with abdominal pain and increased abdominal girth since one year, loss of appetite since two months and diarrhea since last one month. These symptoms were not accompanied by fever. Physical examination revealed abdominal distension and abdominal tenderness. Commuted tomography scan showed a large ill-defined thin walled, cystic and peripherally enhancing space occupying lesion measuring 18x17x7.5 cm, with few thin enhancing septae in pelvis and lower abdomen. Ovaries were not visible separately. Diagnosis on commuted tomography was suspicious of pseudomyxoma peritonei. Patient underwent exploratory laparotomy for definitive diagnosis. The

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abdominal cavity was full of gelatinous fluid and diffuse implantations in the peritoneum. Uterus, cervix with bilateral adnexae, appendix and omentum were received for histopathological examination.

External surface of uterus and bilateral ovaries showed multiple soft gelatinous material covering the entire surface. Cut surfaces of uterus, cervix and fallopian tubes were unremarkable however, ovarian cut surface showed distortion and gelatinous material. Omentum showed fibrofatty tissue covered by gelatinous material. Appendix received separately was grey white with gelatinous areas on the surface and cut surface showed distortion of normal structures filled with gelatinous material [Fig.1]. Microscopy sections from appendix showed a malignant tumor arranged in glandular pattern with complex branching [Fig.2]. Individual tumor cells were columnar showing large hyperchromatic pleomorphic nuclei displaying loss of polarity with moderate amount of cytoplasm. Apical mucin and large amount of extracellular mucin pools were also seen in the stroma [Fig.3]. Immunohistochemistry revealed tumor cells expressing CK-7, CK-20, CEA; immunonegative for Ca-125 and PAX-8. Thus final diagnosis was framed as mucinous adenocarcinoma of appendiceal origin having ovarian and omental metastasis and pseudomyxoma peritonei.

Post operatively the patient received six cycles of chemotherapy and has been followed up for one year without any evidence of recurrence.

Discussion

Pseudomyxoma peritonei is rare, chronic relapsing, diagnostically challenging and poorly understood disease characterized by disseminated mucinous ascites and peritoneal implants [5]. Age of presentation is 50-70 years, have female preponderance with high rate of recurrence [4]. In our case the patient is female and age was 50 years in concordance with other studies.

Fig.1: Gross picture showing uterus with cervix, appendix and bilateral ovaries.

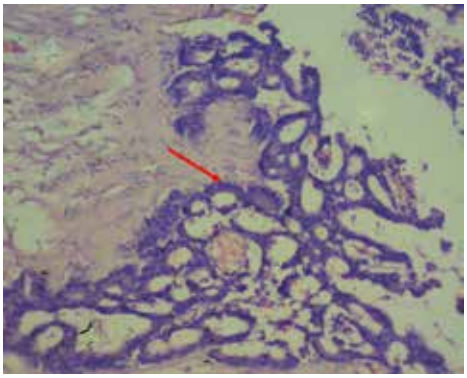

Fig.2: H & E stained section 10x showing malignant tumor in glandular pattern and complex branching (red arrow).

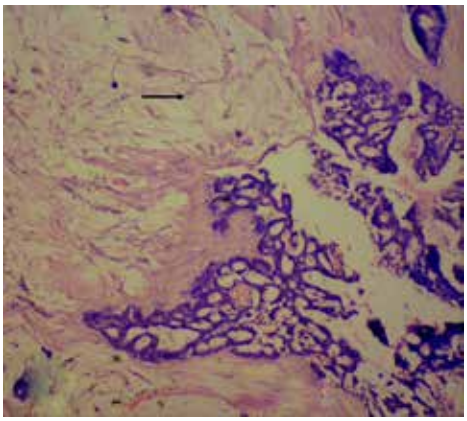

Fig.3: H & E stained section 4x showing mucin pools (black arrow).

Fox and Langely proposed the development of this condition due to foreign body reaction, following spillage of mucus from ruptured cysts into the peritoneal cavity [4]. Benign, malignant and intermediate forms have been described [6]. Our case presented in malignant form. Physiologic principles involved include accumulation and peritoneal absorption of mucinous cells [2]. The myxomatous appearance is attributed to the associated fibroblastic and vascular proliferation that is probably incited by mucin. This results in multifocal, peritoneal, serosal and omental implants resulting in belly full of jelly – "The Jelly Belly". Other terminologies to reflect this spectrum of biological behavior are disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis (PMCA) [5].

Usual features are increased abdominal girth (40%), bilateral or unilateral ovarian tumors (20%), hernia sac tumors (20%), appendicitis like syndrome (10%) and infertility [2]. Our patient presented with increased abdominal girth, ovarian tumor and appendicitis like syndrome. Diagnostic investigations include abdominal ultrasonography supplemented by commuted tomography to reveal the extent of disease. Additional evaluation of tumor markers like CA 19-9 and CEA are used for prognosis [2]. Ultrasonography and tumor markers were not done in our case. Commuted tomography scan will show basic four patterns: (a) posterior displacement of intestine with numerous low density masses and calcifications, (b) diffuse peritoneal infiltration, (c) intrahepatic low density attenuated lesions, (d) scalloping of intra-abdominal organs. Histopathology helps to identify viable epithelial and glandular cells within the mucin pools. A definitive diagnosis of pseudomyxoma peritonei requires presence of (a) mucinous neoplastic cells or epithelium, (b) mucinous ascites [2]. Both of them were found in this case. Advancing abdominal disease caused by intestinal obstruction accounts for majority of morbidity and mortality [4].

Mucinous neoplasms of appendix are uncommon entities associated with variety of underlying pathological processes like simple appendiceal mucoceles, mucinous hyperplasia, serrated adenoma and mucinous neoplasm of uncertain malignant potential. In our case, the origin of the tumor was difficult to ascertain as tumor and mucin pools were diffusely infiltrating appendix and bilateral ovaries. However, the primary site of the malignant tumor was confirmed as appendiceal mucinous adenocarcinoma on immunohistochemistry. Death directly related to pseudomyxoma peritonei is due to extensive peritoneal fibrosis with bowel obstruction rather than lung, liver or lymph node metastasis [5]. Recent studies reveal that pseudomyxoma peritonei is a neoplastic disease of MUC-2 expressing goblet cells. Mucinous tumors of appendix also express MUC-2 [5]. Prognosis is closely related to bulk of the disease evaluated by tumor site, pre-operative tumor volume and completeness of tumor removal by cytoreductive surgery. Sugarbaker has shown that repeated surgeries result in median survival of approximately 2 years [3].

Prompt and aggressive treatment includes drainage of mucus, surgical debulking of primary and secondary tumor implants. Commonly at laparotomy, hemicolectomy is performed. In order to prevent recurrence, resection of both ovaries and appendix must be carried out. Beller *et al*. reported in 1986 that instillation of intraperitoneal mucolytics such as dextran sulphate, urokinase to prevent recurrence [6]. In our case there is no evidence of recurrence.

Conclusion

Pseudomyxoma peritonei secondary to appendicular mucinous adenocarcinoma is an exceedingly rare tumor with a varied presentation and outcomes. Due to paucity of reported cases, prognosis is not known; the continued report of these cases may eventually shed light on prognosis of these tumors.

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