Case Report

Indian Journal of Obstetrics and Gynecology
Volume 5 Number 2, April - June 2017
DOI: http://dx.doi.org/10.21088/ijog.2321.1636.5217.26

Pseudomyxomatous Peritonei

Anjali M. Chitale*, Agraj Mishra**

Abstract

Pseudomyxomatous peritonei is a clinical condition caused by cancerous cells (mucinous adenocarcinoma) that produce abundant mucin or gelatinous ascites. The tumors cause fibrosis of tissues and impede digestion or organ function, if left untreated, the tumors and mucin they produce will fill the abdominal cavity. The standard treatment for these patients is watchful monitoring and debulking followed by chemotherapy. I report a case of a 54 year old female and review the clinical features and treatment of this disease.

Keywords: Ovary; Pseudomyxomatous Peritonei; cyst.

Introduction

A pseudomyxomatous change of ovary is a rare condition and most of its presentations are benign and rarely descends into malignant changes. In the modern era of medicine, such huge mucinous ovarian tumors have become rare in the current medical practice, as most of the cases are diagnosed early during routine gynecological examinations or incidental finding on the ultrasound examination of the pelvis and abdomen [1]. Most of the patients who have large tumors present mainly with the pressure symptoms over the genitourinary system leading to urinary complaints and also pressure over respiratory system leading to respiratory embarrassment. The role of imaging modalities such as computed tomography (CT) scan and magnetic resonance imaging gives better idea about the extension of the tumor in the various quadrants of the abdomen and consistency of the tumor. Management of ovarian cysts depends on the patient's age, the size of the cyst, and its histopathological nature. Four frozen sections are very important to know the malignant variation of this tumor and that helps in the management of the patient.

Case Report

A 54 yrs old female came with gradually increasing distension of abdomen since last 6- 8 months. The patient was menopausal since last 7 years and there was no significant past medical or surgical history. On examination her general condition was fair, thin built and her weight was 50kgs. Vitals were stable; pulse was 68 beats/min, and blood pressure was 110/70 mmHg. Respiratory and cardiovascular system examination findings were normal. In per abdomen examination and inspection, there was huge distension all over the abdomen and skin over the abdomen had thinned and was shiny. On palpation, the tumor was 32 weeks of uterine size, soft to firm in consistency, mobility like ascitic fluid; margins were ill defined, were extending from the lower pelvis till xiphisternum and laterally upto both sides of iliac fossa. Her hematological investigations were within normal limits. Her liver, renal functions were also in normal values.

Her USG reports were s/o cystic mass of

*Professor **Junior Resident, Department of Surgery, A.C.P.M. Medical College, Dhule, Maharashtra 424001, India.

Corresponding Author:
Agraj Mishra,
Junior Resident,
department of Surgery,
A.C.P.M. Medical College,
424001, India.
E-mail:
mishraagraj37@gmail.com

Received on 01.01.2017,
Accepted on 22.02.2017

size 30 X 12 X 16 cms extending from supraumbilical region to pelvis.

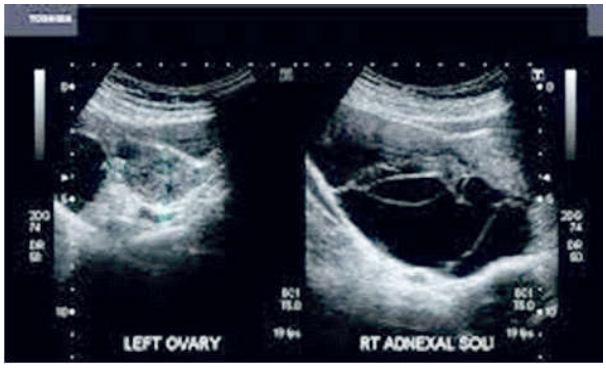

Fig. 1:

CT Abdo+pelvis was s/o multiseptated peripherally partially calcified cystic SOL in lower abdomen and pelvis on right side suggestive of ovarian neoplastic etiology like mucinous cystadenocarcinoma.

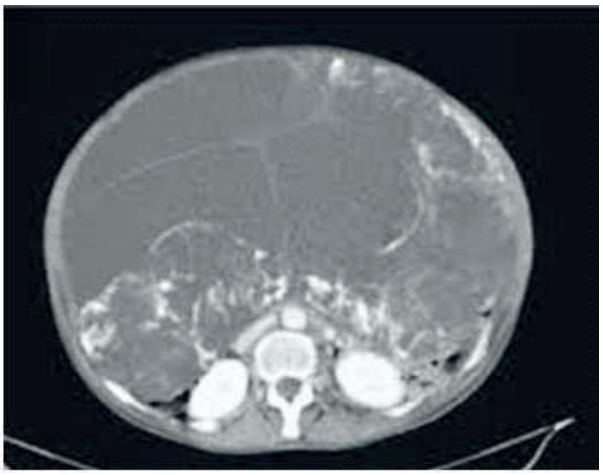

Fig. 2:

Patient was planned for exploratory laprotomy for debulking procedure after fitness. A midline incision was taken and peritoneum was reflected and mucoid masses evacuated completely and appendicecomy along with hysterectomy with bilateral salpingo-oopherectomy was done. Post operative recovery was uneventful. The patient withstood the surgery well with no intraoperative or postoperative complications. On day 10 of surgery, complete suture removal was done and wound was healthy.

Histopathology of specimen sent was consistent with multiple cystic secondary deposits of mucinous adenocarcinoma with calcification and bone formation.

Hence, patient was discharged and referred to oncology department for further management. The patient received four cycles of adjuvant chemotherapy (oral capecitabine) and was discharged in healthy condition.

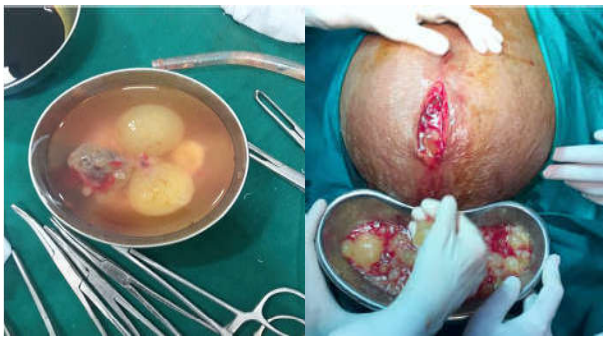

Fig. 3: Intra-Operative Photographs

Literature Review

Pseudomyxomatous peritonei is a rare condition with an incidence of 0.5-1 per cent per million population. The exact origin of this condition point, specially in women, stem from the fact that synchronous appendiceal and ovarian disease is common and overall preponderance for it. The disease is characterized by diffuse intra-abdominal gelatinous collections (jelly belly) with mucinous implants on peritoneal surfaces and omentum. The locoregional progression of the disease results in intestinal failure and malnutrition secondary to raised intra abdominal pressure, fistula formation and infection and hence the associated considerable mortality and morbidity.

In 1995, Ronnett et al proposed separating pseudomyxoma peritonei cases into two diagnostic categories: adenoma (disseminated peritoneal adenomucinosis, DPAM) or carcinoma (peritoneal mucinous carcinomatosis, PMCA) with a third category reserved for cases with intermediate features. The imging and staging modality of choice is CT scanning, in most cases the striking feature is the relative sparring of small bowel and its mesentry, with small bowel content, mucin has similar appearance to water in CT.

In past, treatment compromised of intreval debulking with no realistic long time cure. The natural history of disease had been altered by Sugarbaker who advised debulking followed by HIPEC to eliminate microscopic and residual disease. The majority had (60%) had complete cytoreduction. The uniform treatment approaches has improved shown 10 year survival rate, as compared with past treatment.

Discussion

Pseudomyxoma peritonei is an indolent disease and preferentially affects women with an average age of 53 years. It is traditionally believed that most cases of PMP originate from ovarian tumors. As symptoms remain non-specific the disease presents a great diagnostic challenge to clinicians. Clinical presentation is late and patients usually experience a long course of health deterioration before an accurate diagnosis is made.

Surgical expertise is required to prevent complications as in huge tumors the anatomical planes get distorted. Giant mucinous cystadenocarcinomas are very rare with a huge abdominal enlargement. The main objective of this report is to call attention to ovarian epithelial cysts in the outpatient clinics and primary care services, contributing to a decrease in any underdiagnosis, misdiagnosis, and underreporting that might occur.

References

1. Werth R. Pseudomyxoma peritonei. Arch Gynaecol. 1884, 24:100-118. 10.1007/BF01837425.
2. Fann JI, Vierra M, Fisher D, Oberhelman HA, Cobb L: Pseudomyxoma peritonei. Surg Gynecol Obstet. 1993; 177:441-447.
3. Ronnett BM, Zahn CM, Kurman RJ, Kass ME, Sugarbaker PH, Shmookler BM: 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'. Am J Surg Pathol. 1995; 19:1390-1408.
4. Galani E, Marx GMF, Steer CB, Culora G, Harper PG: Pseudomyxoma peritonei: the 'controversial' disease. Int J Gynecol Cancer. 2003; 13:413-418. 10.1046/j.1525-1438.2003.13318.
5. Hinson FL, Ambrose NS.: Pseudomyxoma peritonei. Br J Surg 1998; 85:1332–1339.
6. van Ruth S, Acherman YIZ, van de Vijver MJ, Hart AAM, Verwaal VJ, Zoetmulder FAN: Pseudomyxoma peritonei: a review of 62 cases. Eur J Surg Oncol. 2003; 29:682-688. 10.1016/S0748-7983(03)00149-5.
7. Sherer DM, Abulafia O, Fliakim R: Pseudomyxoma peritonei. A review of current literature. Gynecol Obstet Invest. 2001; 51:73-80. 10.1159/000052897.
8. Ronnett BM, Shmookler BM, Diener-West M, Sugarbaker PH, Kurman RJ: Immunohistochemical evidence supporting the appendiceal origin of pseudomyxoma peritonei in women. Int J Gynecol Pathol. 1997; 16:1-9.
9. Pandey A, Mishra AK. Pseudomyxoma peritonei: disseminated peritoneal adenomucinosis variant. BMJ Case Rep 2011.p.1136.
10. Pseudomyxoma peritonei. Cancer Treat Res 1996; 81:105–19.
11. Pai RK, Longacre T: Appendiceal mucinous tumors and pseudomyxoma peritonei: Histologic features, diagnostic problems and proposed classification. Advances in Anatomic Pathology. 2005; 12:291-311. 10.1097/01.pap.0000194625.05137.51.
12. Foster DR. Ultrasound findings in pseudomyxoma peritonei. Australas Radiol 1985; 29:39–41.
13. O'Connell JT, Tomlinson JS, Roberts AA, McGonigle KF, Barsky SH: Pseudomyxoma peritonei is a disease of MUC2-expressing goblet cells. AmJ Pathol. 2002; 161:551-564.
14. Sugarbaker PH. Peritonectomy procedures. Ann Surg. 1995; 221:29-42.
15. Dhage-Ivatury S, Sugarbaker PH: Update on the surgical approach to mucocele of the appendix. J Am Coll Surg. 2006; 202:680-684. 10.1016/j.jamcollsurg. 2005.12.003.
16. Sugarbaker PH, fernandez-Trigo V, Shamsa F: Clinical determinants of treatment failure in patients with pseudomyxoma peritonei. Cancer Treat Res. 1996; 81:121-32.
17. Miner TJ, Shia J, Jaques DP, Klimstra DS, Brennan MF, Coit DG: Long-term survival following treatment of pseudomyxoma peritonei: An analysis of surgical therapy. Ann Surg. 2005; 241:300-308. 10.1097/01.sla.0000152015.76731.1f.
18. Qu Z, Liu L. Management of pseudomyxoma peritonei. World J Gastroenterol 2006; 12(38):6124-6127.
19. Miner TJ, Shia J, Jaques DP, et al. : Long-term survival following treatment of pseudomyxoma peritonei: an analysis of surgical therapy. Ann Surg 2005; 241: 300–308.
20. Fox H, Langley FA. Tumours of the ovary. London: Heinneman, 1976.p.46–7.
21. D Elias, F Gilly, F Quenet , etal: Pseudomyxoma peritonei: A French multicentric study of 301 patients treated with cytoreductive surgery and intraperitoneal chemotherapy Eur J Surg Oncol 2010 March 12; 36: 456–462.
22. Andreason H, T. Lorant, L. pahlman, W. Graf and H. Mahteme. cytoreductive surgery. Eur. J.Surg. Oncol. 2014; 40:930-936.
23. Chua; et al. Early- and Long-Term Outcome Data of Patients With Pseudomyxoma Peritonei From Appendiceal Origin Treated by a Strategy of Cytoreductive Surgery and Hyperthermic Intraperitoneal Chemotherapy". Journal of Clinical Oncology. 2012; 30(20):2449–2456.