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PSEUDOMYXOMA PERITONEI: A CASE REPORT OF SYNCHRONOUS LESIONS FROM APPENDIX AND OVARY

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ABSTRACT

PSEUDOMYXOMA PERITONEI (PMP) is a rare tumor well known for its mucin production mainly arising from perforated appendix and/or ovary , which later gets implanted in the abdominal cavity and diffusely involving the peritoneal surfaces. It has an incidence of 1-2 in a million[1]. Classically it is characterized by massive abdominal distension and nutritional compromise in most cases due to diffuse intra-abdominal gelatinous collections (jelly belly) with mucinous implants on peritoneal surfaces and the omentum[2]. Optimal treatment involves a combination of cytoreductive surgery (CRS) with heated intraperitoneal chemotherapy (HIPEC). Following CRS with HIPEC, 5-year survival ranges from 62.5% to 100% for low grade, and 0%-65% for high grade disease. This is a case report which discusses the presentation and treatment options.

KEYWORD

pseudomyxoma peritonei (PMP), Cytoreductive surgery(CRS), Heated intraperitoneal chemotherapy(HIPEC)

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CASE REPORT:

A 81yr old female patient presented with lump abdomen associated with pain since 4 months. She takes mixed diet, has decreased appetite and irregular bowel habits. On clinical examination abdomen is distended, tenderness noted over the lower abdomen. Palpable mass arising from the pelvis. On CECT abdomen the findings were, well defined irregular homogenous peripherally enhancing hypodense cystic lesion with soft tissue component in the left side of pelvis with ill-defined medial cyst wall, with peritoneal, omental, serosal deposits with moderate ascites- possible malignant ovarion lesion. CA125 estimation was 12 U/ml (normal range < 30 U/ml). An exploratory laparotomy was performed, intraoperatively thick jelly like content occupying the entire abdomen was noted (figure 1) with diffuse implantations in peritoneumwith a ruptured ovarian cyst on left side(figure 2). There was even a large cystic appendix of 10x5cms noted. Peritoneal lavage, hysterectomy, bilateral salpingo-oophorectomy, omentectomy, appendicectomy and peritonectomy were performed. HPE reported a well differentiated mucinous adenocarcinoma arising from ovary and appendix infiltrating the peritoneum and omentum.

DISCUSSION:

PMP predominantly originates in the appendix in men and in women, synchronous ovarian and appendiceal disease is common. However, in women after immune-histochemistry and molecular genetic studies reveals that ovarian tumors are due to secondary deposits of perforated appendiceal mucinous tumor[3-6].

Majority of the patients were diagnosed accidentally on laparotomy or laproscopy for suspected appendicitis or ovarian malignancies.

Pathophysiology:

Appendiceal adenoma
↓
Occlude appendiceal lumen
↓
Distension of appendix with mucus and mucinous tumor cells
↓
Eventual rupture of appendix
↓
Subsequent sloe leak of tumor cells into peritoneum
↓
The tumor cells proliferate and produce large quantities of mucus in the peritoneal cavity
↓
Jelly belly

Ronnett and colleagues, have classified the PMP into three categories with low-grade tumours as disseminated peritoneal adenomucinosis (DPAM) and high-grade tumours as peritoneal mucinous carcinomatosis (PMCA), with an intermediate group (IG) demonstrating a mixture of DPAM and PMCA.

Treatment:

Misdraji et al[7] reported on 107 patients with a median survival of 7.5 years, and a 20-year survival of 25% after serial debulking and perioperative intra-peritoneal chemotherapy. Sugarbaker et al[8,9], has introduced the combined treatment of CRS with HIPEC which has increased the 10 year survival rate to 80%.

CONCLUSION:

PMP is uncommon and generally originates from a perforated appendiceal tumour. The optimal treatment involves a

combination of surgery and HIPEC. The treatment strategy is complex, associated with significant morbidity and mortality. The long-term outcomes for CRS with HIPEC in PMP are impressive for patients with low-grade histology amenable to complete cytoreduction.

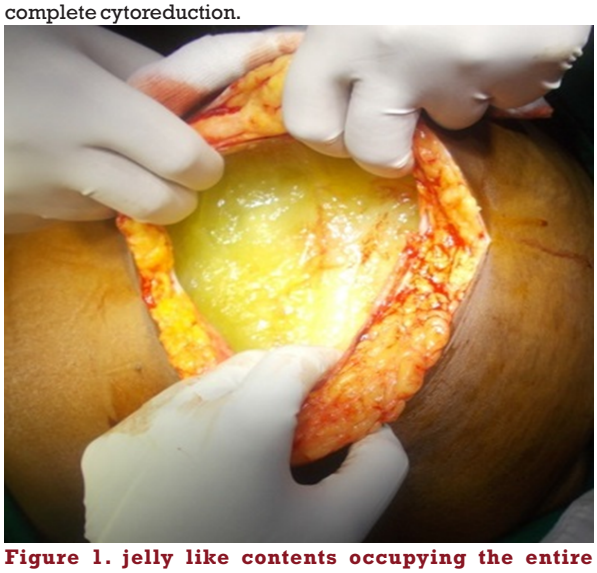

Figure 1. jelly like contents occupying the entire abdomen (jelly belly).

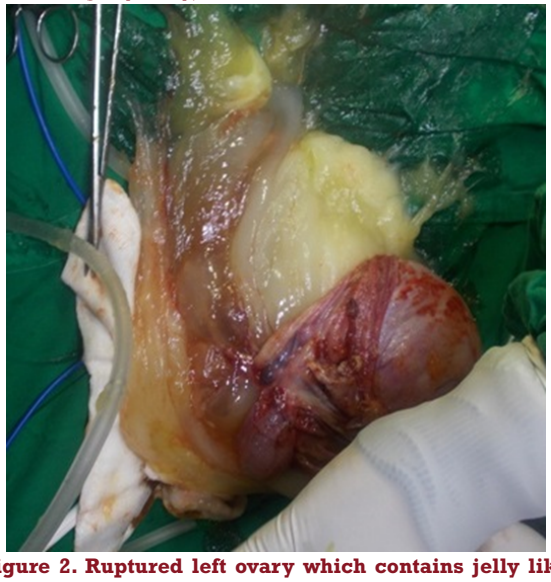

Figure 2. Ruptured left ovary which contains jelly like substance.

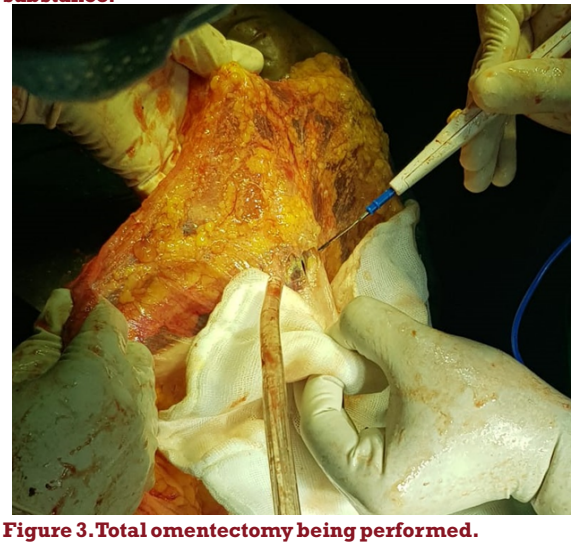

Figure 3. Total omentectomy being performed.

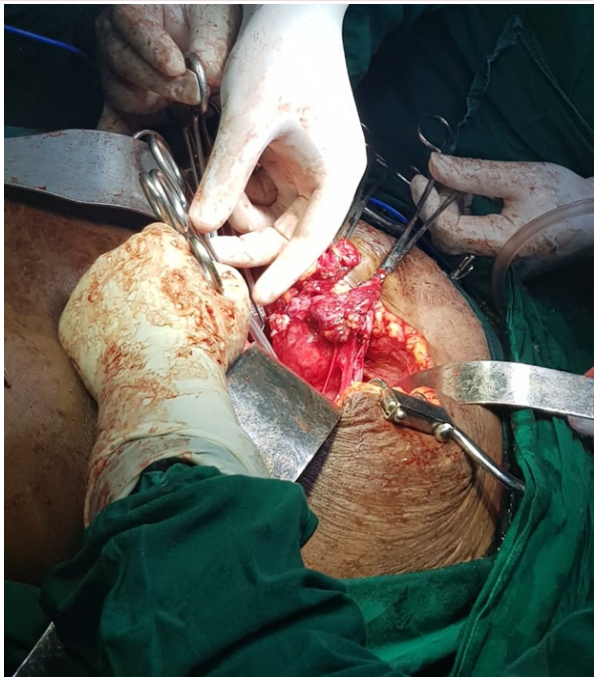

Figure 4 . abdominal hysterectomy with bilateral salpingo- opherectomy being done.

REFERENCES:

1. Smeenk RM, van Velthuysen ML, Verwaal VJ, Zoetmulder FA. Appendiceal neoplasms and pseudomyxoma peritonei: a population based study. Eur J Surg Oncol. 2008;34:196–201. [PubMed] [Google Scholar]
2. Moran BJ, Cecil TD. The etiology, clinical presentation, and management of pseudomyxoma peritonei. Surg Oncol Clin N Am. 2003;12:585–603. [PubMed] [Google Scholar
3. 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. [PubMed] [Google Scholar]
4. Szych C, Staebler A, Connolly DC, Wu R, Cho KR, Ronnett BM. Molecular genetic evidence supporting the clonality and appendiceal origin of Pseudomyxoma peritonei in women. Am J Pathol. 1999;154:1849–1855. [PMC free article] [PubMed] [Google Scholar]
5. Chuaqui RF, Zhuang Z, Emmert-Buck MR, Bryant BR, Nogales F, Tavassoli FA, Merino MJ. Genetic analysis of synchronous mucinous tumors of the ovary and appendix. Hum Pathol. 1996;27:165–171. [PubMed] [Google Scholar]
6. Guerrieri C, Frånlund B, Fristedt S, Gillooley JF, Boeryd B. Mucinous tumors of the vermiform appendix and ovary, and pseudomyxoma peritonei: histogenetic implications of cytokeratin 7 expression. Hum Pathol. 1997;28:1039–1045. [PubMed] [Google Scholar]
7. Misdraji J, Yantiss RK, Graeme-Cook FM, Balis UJ, Young RH. Appendiceal mucinous neoplasms: a clinicopathologic analysis of 107 cases. Am J Surg Pathol. 2003;27:1089–1103. [PubMed] [Google Scholar]
8. Sugarbaker PH. Surgical treatment of peritoneal carcinomatosis: 1988 Du Pont lecture. Can J Surg. 1989;32:164–170. [PubMed] [Google Scholar]
9. Sugarbaker PH, Kern K, Lack E. Malignant pseudomyxoma peritonei of colonic origin. Natural history and presentation of a curative approach to treatment. Dis Colon Rectum. 1987;30:772–779. [PubMed] [Google Scholar]