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Pseudomyxoma Peritonei or Jelly Belly – Varied Presentations

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Abstract

Pseudomyxomaperitonei is a rare gynaecological condition characterised by mucinous ascites. It has been described as the presence of gelatinous ascites and implants that involve the peritoneal surfaces and omentum. Though essentially associated with mucinous cystadenomas of the appendix and the ovaries , the proliferative nature and gross mucinous ascites associated with PMP demand the need for an optimal debulking surgery. Newer modalities of treatment for PMP are still being studied, intra peritoneal chemotherapy has been found to be the most useful after debulking surgery, to prevent recurrence.

Keywords: Pseudomyxoma Peritonei; Jelly Belly; Mucinous Ovarian Tumours.

Introduction

Pseudomyxomaperitoneiis a rare gynaecological condition which was first described by Werth, as the presence of gelatinous ascites and implants that involve the peritoneal surfaces and omentum.[1]In the western world the average incidence is 1 to 2 million per year. Mucinous tumours of the ovary can very often present as pseudomyxomaperitonei and these tumours are often labelled as mucinous low malignant potential (LMP) tumours. It is also associated with mucinous cysts of the appendix and rarely with malignant cystadenomas.

This case report deals with four interesting and varied presentations of PMP (pseudomyxomaperitonei) that were seen over a period of three years in a tertiary institute and their management.

Case Series

Case 1

A 55 year old postmenopausal lady , presented with abdominal bloating and distension since two months , that kept progressively increasing. She did not have any associated bowel or bladder difficulties and also did not show any features of fever , jaundice or breathlessness. She was tachyapneic on examination and other vitals were normal. It was noted that she had a vague yet large palpable abdominal mass extending from the pelvis to both the iliac fossa. The borders of the abdominopelvic mass could not be differentiated. The uterus and bilateral adnexa felt unusually normal in position and consistency.

Her haematological work up was normal and tumour marker was as follows : CA 125 : 43.6 IU

An ultrasound and CT revealed bilateral cystic masses right side 11cm x 17 cm and left side 11 x 9 cm with multiple septations , displacing the bowel on both sides. Mild omental thickening was noted and an impression of a malignant cysic ovarian mass was made.

The RMI (risk of malignancy scoring) for the patient was 392.4. This suggested moderate risk of malignancy , intermediate chances of survival and the need for intervention of an oncosurgoen with the gynaecologist.

The patient was posted for a staging laparotomy. Intra operatively the abdominal cavity was distended with mucin. On further examination bilateral ovaries were enlarged and cystic of about 11cm x 9 cm masses and

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mucin was found over the appendix and omentum. A total abdominal hysterectomy with bilateral salphingo ophorectomy was done. In addition an appendicectomy and infracolic omentectomy was done.

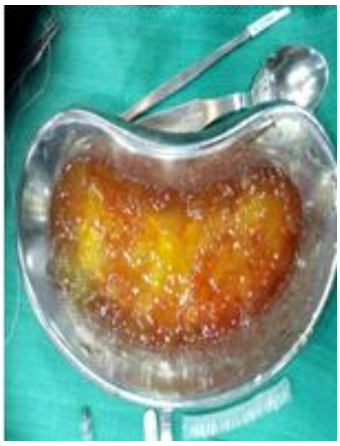

Fig. 1: Mucin found in the abdominal cavity

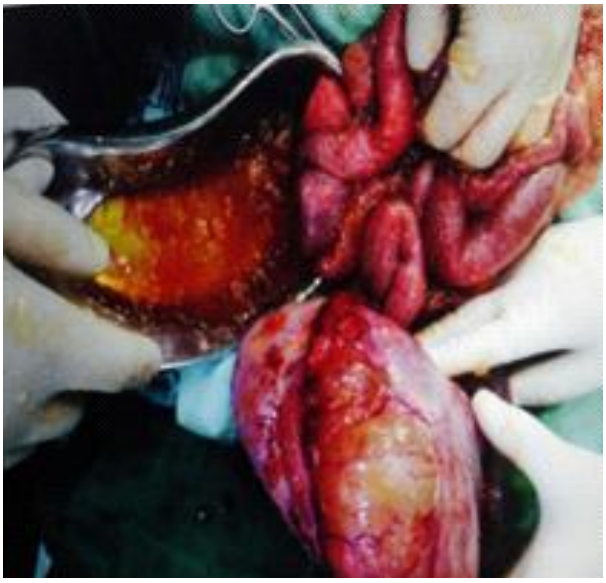

Fig. 2: Mucin stained bowel and appendix

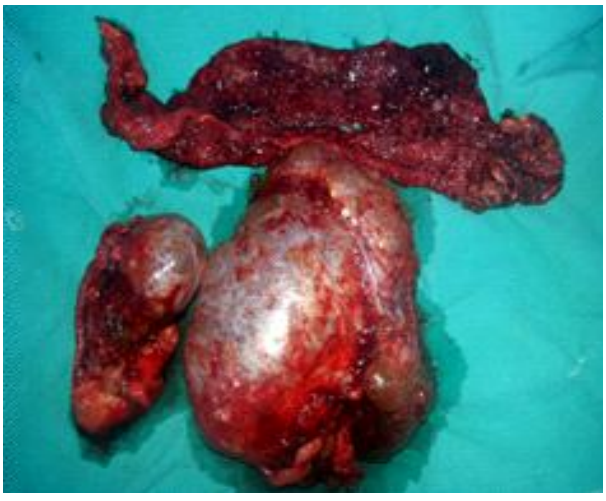

Fig. 3: Specimen of Uterus, bilateral ovarian masses and omentum

Post operative course of the patient was uneventful and histopathology confirmed the diagnosis of a mucinous cystadenoma.This case of PMP was associated with a benign ovarian neoplasm.

Case 2

A 48 year old male patient presented with abdominal distention since 1 yrassociated with vague abdominal pain . He had a significant past historypf undergoing surgery for mucinous cystadenoma of appendix with PMP 11yrs prior. When he was examined he was found to have multiple , firm masses in epigastrium , right iliac fossa and hypogastrium.

A CT scan revealed the following: CT scan**-** Multiple cystic masses with septations involving the mesentry, omentum, lesser sac and perigasrtium indenting the gastric wall and scalloping the body and the tail of the pancreas. Exploratory laparotomy, suboptimal debulking of the mucin and a right hemicolectomy was done. Nodular mucoid cysts were found in the colon and mucoid cystic lesions in the hepatic flexure.

Fig. 4: Specimen of Hemicolectomy with mucin and nodular mucoid cysts

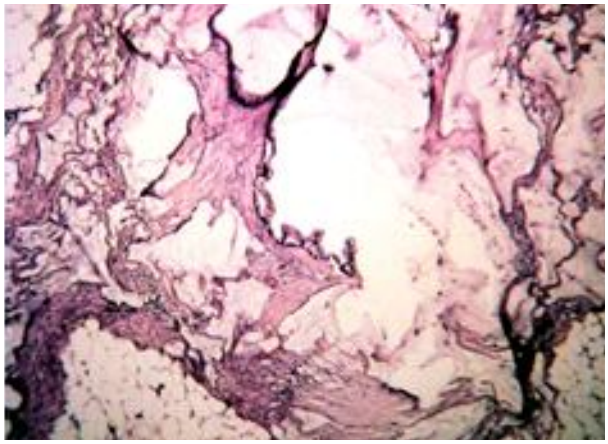

Fig. 5: Histology showing fibrosis in the bowel wall with mucin deposits

Postoperatively , the patient recovered well. The histology confirmed the benign nature of the disease.

In this casepseudomyxomaperitonei was associated with mucinous cysts of the colon.

Case 3

A 68 year old postmenopausal lady presented with progressivedistention of abdomen since 3 weeks. He also had a past history of pseudomyxomaperitonei and was operated 3 yrs back for the same. When examined he appeared to have gross ascites and a fluid thrill could be elicited.

An ultrasound of the abdomen gave the following impression: Gross ascites with fluid collection causing scalloping of the diaphragmatic surface of the liver.

On followup a CT Scan was done and it showed gross ascites with omentalthickening and also fluid collection along the diaphragmatic surface causing scalloping. Following these reports a provisional diagnosis of Pseudomyxomaperitoneiwas made.

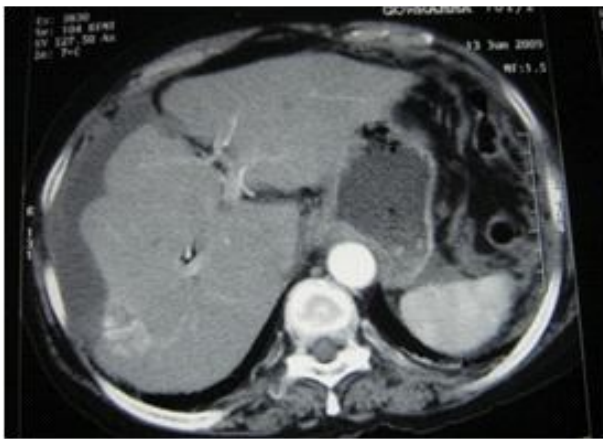

Fig. 6: Fluid collection along the diaphragmatic surface causing scalloping

Folllowing investigations the patient underwent a laparotomy. Further a total abdominal hysterectomy ,omentectomy and debulking of mucin was done. Histopathology showed diffuse areas of fibrosis and mucin filled globules in the omentum.

Cytokeratin profile showed negativity for CK-7. which is a marker for ovarian tumors. In this case PMP was associated with mucinous omental deposits and benign ovarian tumours.

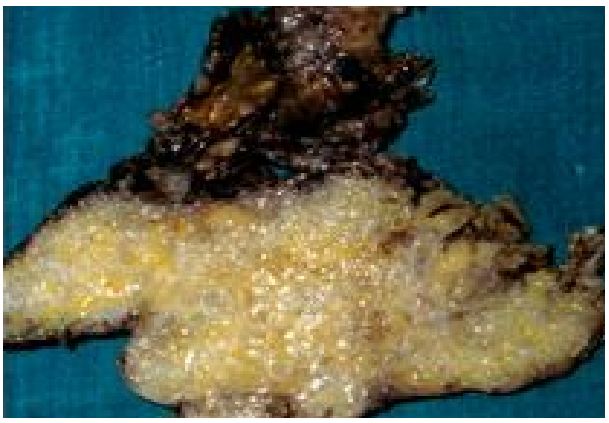

Fig. 7: Omental thickening with mucin.

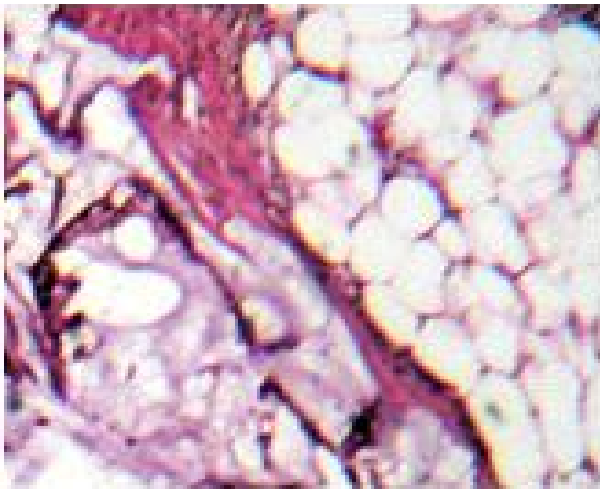

Fig. 8: Magnified view: Mucin globules

Case 4

A 69 year old male presented to the ER with pain abdomen and fever since 3 days. On examination he had diffuse abdominally tenderness and a clinical impression of acute colitis was made. He had a past history of diabetes mellitus. A CT scan of the abdomen showed multiple pockets of free intraperitoneal air and peripherally enhancing pelvic collection with air-fluid levels with ascites.

Patient was prepared for an emergency laparotomy. Intra operatively,multiple peritoneal, omental mucinous deposits over the bowel. There was also thick purulent fluid collection and it was found dilated appendix with perforation. A resectionanastamosis of tumor with segment of ileum was done.TheCytokeratin profile showed a positivity of CK 20.

Post operative histopathology confirmed the diagnosis of pseudomyxomaperitonei.

In this case PMP was associated with mucinous adenoma of the appendix, that presented with perforation.

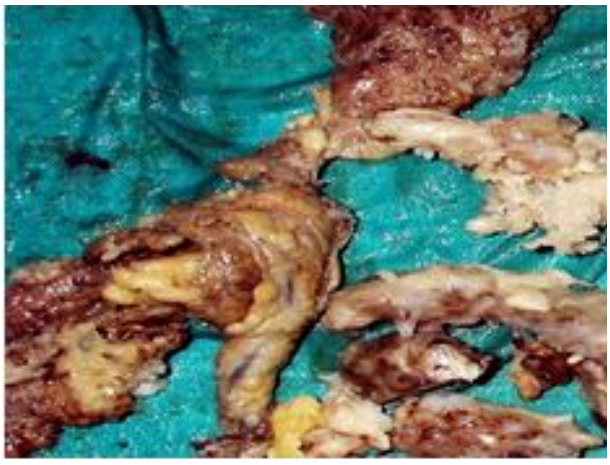

Fig. 9: Peritoneal deposits of mucin

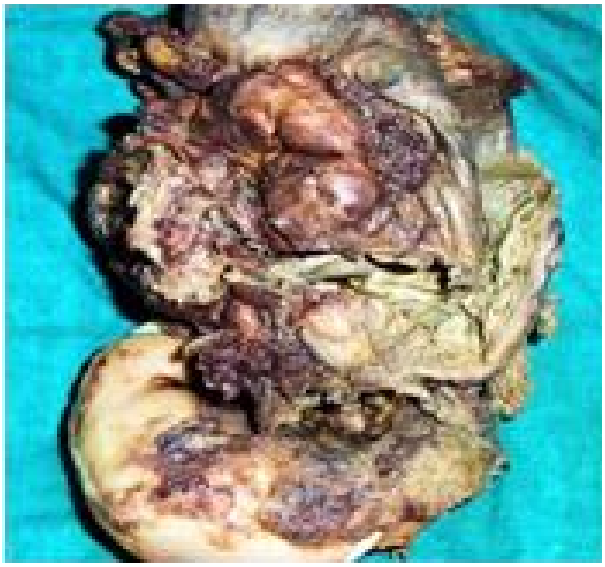

Fig. 10: Perforated appendix

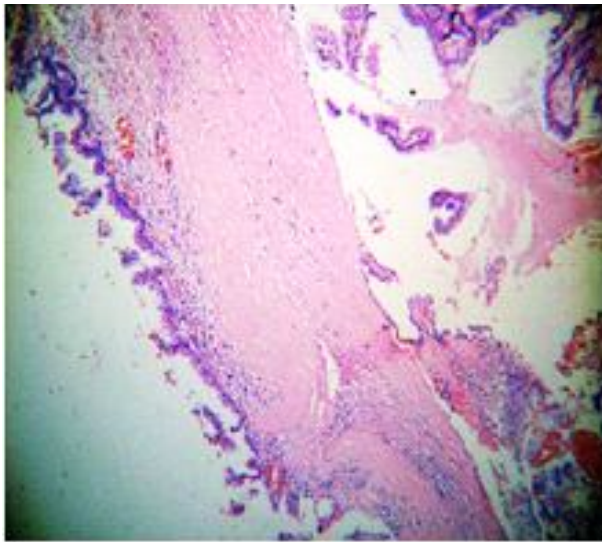

Fig. 11: Histology showing mucn deposits beneath the epithelium

Discussion

Pseudomyxomaperitonei means false mucinous tumor of the peritoneum. It is most commonly applied to a slowly progressive disease process characterized by copious amounts of mucus that, over time, fills the peritoneal cavity. Associated tumors are not considered biologically aggressive because it does not invade or metastasize, although it is a deadly process.[2]

Psuedomyxomaperitonei is most commonly associated with benign, borderline or malignant mucinous tumors of the ovary and appendix and rarely with mucinous tumors of the urachus, bowel, pancreas and common bile duct. [3]

The etiopathogenesis is explained by the " redistribution phenomenon" which suggests that abdominal surfaces that absorb peritoneal fluid (such as the greater omentum and the undersurfaceof the diaphragm) are coated by these tumor cells. These tumour cells do not invade visceral tissue but only extensive proliferate on the surface. This leads to gross ascites in PMP and minimal visceral invasion. Some authors also suggest mucinous metaplasia in the peritoneal mesothelium to be a cause [4].

The most common presenting symptom ofPMP is abdominal distension that progresses rapidly over a short duration. Other symptoms could be of abdominal pain, fever, vomiting, anorexia and weight loss. All four cases in our series presented with abdominal distension. Its median age of onset has been noted to be 55 years and the male to female ratio of occurrence of pseudomyxomaperitonei is 7:3.

We noted that two of our cases were in postmenopausal women and two were in males above the age of 40.

An exploratory laparotomy is the only key to a diagnosis and confirmation of PMP. However this condition should be suspected in patients when radiologically there is evidence of distended abdomen with air fluid levels, displacing the bowel or scalloping the diaphragm are seen and the patient is stable. No laboratory tests have found to be diagnostic though CT scan can be of great help in clinching the diagnosis.

As the disease progresses rapidly , a good number of patients present with emergency obstructive symptoms and require an emergency debulking surgery.

The mainstay of treatment for pseudomyxoma peritonei remains cytoreductive surgery, removing the primary disease, ie, oophorectomy and / or total abdominal hysterectomy, followed by removal of mucinous nodules from the omentum and peritoneal surfaces. Appendectomy must be done and submitted for through sampling by microscopic analysis [2].

Newer modalities of therapy include laser cauterisation of peritoneal implants, peritoneal stripping, adjuvant mucolytic agents like dextran sulphate, photodynamic therapy and radiotherapy. These however have not been proven to be of great use and are still in the phase of research [5].

Cisplatin-based regimens have become the standard of treatment in cases of ovarian epithelial neoplasms and researchers have reported using single cisplatin or cisplatinum-based regimens to treat pseudomyxoma. However the efficacy of these modalities in preventing recurrence is questionable. Two of our patients underwent revision surgery after being diagnosed previously with PMP and were cases of recurrence.

Intra peritoneal chemotherapy is a popular method used in the treatment of PMP. Though varying amount of success in preventing recurrence has been noted, it has a large number of side effects like abdominal pain, seizure, neutropenia and thrombocytopenia.

Patients with ovarian tumors of low malignant potential have a significantly better prognosis than patients with adenocarcinoma. The overall 5- and 10- year survival rates for patients with borderline tumors are 85-90% and 75-80% [6,7].

Conclusion

Pseudomyxomaperitonei or jelly belly is a rare gynaecological condition and there is a lot of debate revolving around its origin and optimum treatment. It is commonly associated with benign mucinous cystadenoma of the ovary and intestine. Though essentillay benign, it can be associated with malignancy and the aggressive nature of spread makes it a condition to watch out for.

An optimal debulking procedure done by skilled surgical expertisecould be the key to preventing recurrence in PMP. Post operative intra peritoneal chemotherapy is a recommended treatment option. Newer methods to tackle its recurrence are still being researched.

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