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Pathology

AN UNUSUAL CASE OF PSEUDOMYXOMA PERITONEI PRESENTING AS AN OBSTRUCTED UMBILICAL HERNIA: A CASE REPORT WITH BRIEF REVIEW OF LITERATURE.

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ABSTRACT Pseudomyxoma peritonei (PMP) is an uncommon clinicopathologic entity with an unique biological behaviour. It is characterised by disseminated mucinous pool throughout the peritoneal cavity. Mucinous neoplasms of appendix, ovary, pancreas, gall bladder and urachus are implicated in the causation of this dreaded complication. Early diagnosis of PMP is challenging, owing to its variable clinical symptoms and signs. However, it is imperative to diagnose PMP at the earliest, because early treatment with cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC) have satisfactory survival rates.

Herein, we present a case of a 70yr old elderly lady with an incidental finding of diffuse mucinous pool in the peritoneal cavity presenting as an obstructive umbilical hernia on laparotomy. We also discuss a brief review of literature with the emphasis on the latest PMP classification and terminology.

KEYWORDS : Disseminated Peritoneal Adenomucinosis, Gelatinous ascitis, Low grade appendiceal mucinous neoplasm, Mucinous neoplasia.

INTRODUCTION:

Pseudomyxoma peritonei (PMP) is a rare clinicopathologic entity with an incidence of 1-2 per million people and is characterized by extensive deposition of mucin containing tumor cells in the peritoneal cavity creating a Jelly- Belly abdomen.[1]The origin of PMP is traceable to the underlying mucinous neoplasia of intraperitoneal organs.[1,2] Early diagnosis of PMP is challenging and patients often present at the late stage of the disease. The wide range of presentation include abdominal distention, intestinal obstruction, malnutrition, cachexia and ultimately death.[3] At times it is an incidental presentation at laparotomy as experienced in our case of an obstructed paraumbilical hernia in a 70 year old lady.

CASE REPORT:

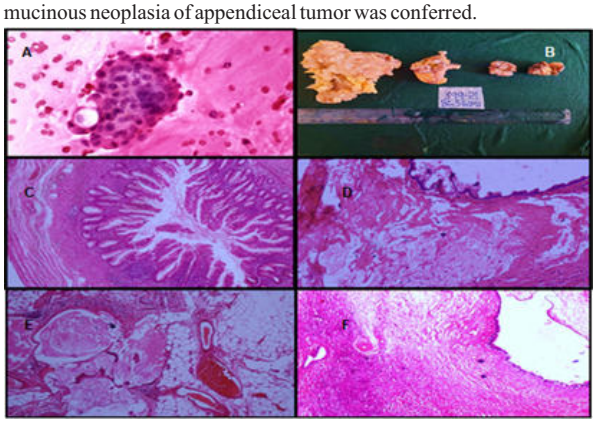
A 70 year female presented with abdominal pain and distension along with swelling below the umbilicus since 3months. There was no history of vomiting, constipation, or secondary changes over the swelling. Abdominal examination revealed ascites. A tense swelling of 5X4 cm was seen 3cm below the umbilicus. The swelling was irreducible. Clinical diagnosis of an obstructed paraumbilical hernia was done and the patient was further evaluated.

Computed tomography (CT) of abdomen revealed right paraumbilical hernia with small bowel loop and fluid within the hernial sac. Mild dilatation of small bowel loops was seen in the abdomen,suggestive of obstruction. Abdominal cavity and pelvis showed massive accumulation of gelatinous free fluid.

Emergency exploratory laparotomy of the abdomen revealed an irreducible infraumbilical hernia. Thick, viscid, mucinous fluid measuring around 1.2-1.5 liters noted in peritoneal cavity. Multiple mucinous deposits were seen over all the abdominal organs including the omentum, mesentery, adnexa, appendix and supracolic compartment. Both the ovaries were atrophic and showed mucinous deposits on the surface. Reduction of hernia, peritoneal exploration, appendicectomy, bilateral salpingo-oophorectomy, omental biopsy and peritoneal lavage was performed.

Cytological analysis of ascitic fluid revealed malignant epithelial cells in sheets, clusters, papillaroid fragments, and in singles on a background of thick mucinous material, numerous reactive mesothelial cells, muciphages, lymphocytes and neutrophils (Fig A). A high index of suspicion for a mucinous neoplasm probably of low grade was considered.

Specimen of omentum, bilateral ovaries, appendix was sent for histopathological examination. Gross examination revealed jelly like mucoid material covering the surface of all specimens (Fig B). Appendix was not distended and ruptured. Histopathologically, all embedded sections from the appendix revealed features of serrated adenoma and low grade appendiceal mucinous epithelium (LAMN) with dissecting pools of mucin extending upto mesoappendix (Fig C, D). Omentum showed wide areas of extracellular mucin pools, few of them were rimmed by tall columnar to pseudostratified columnar epithelium (Fig E). Strips and small islands of tumour cells were seen lying freely within the mucin pool.The tumour cells were bland with minimal atypia at places. Mitosis was absent.Sections from bilateral ovaries showed features of mucinous cystadenoma with dissecting pools of mucin (Fig F). Final histopathological report of Disseminated peritoneal adenomucinosis (DPAM): pM1b,Grade1 with underlying mucinous neoplasia of appendiceal tumor was conferred.



Figure/ Table 1: **(A) Ascitic fluid indicative of low grade mucinous carcinoma (B) Gross specimen of omentum, appendix with bilateral ovaries showing mucinous deposits over surface. (C&D) Microphotograph of appendix showing serrated adenoma and LAMN (H&E, 20x) (E) Microphotograph of omentum showing extracellular mucin pools with low grade mucinous neoplastic**

epithelium (H&E, 20x) (F) Microphotograph of ovary showing mucinous cystadenoma (H&E, 20x).

DISCUSSION:

PMP is a rare clinicopathological malignant syndrome characterised by extensive dissemination of gelatinous mucin containing tumour cells in the abdominal cavity. Mucin deposits in the same pathway as the peritoneal fluid and redistributes in the sites of lymphatic absorption with involvement of omentum, pelvis, paracolic gutters, liver capsule, culde-sac, right retro hepatic space, left abdominal gutter, ligament of Treitz and the serosal surfaces of the visceral organs. [4,5] Small intestine is spared due to the peristalitic movements. It was first described by Werth in 1884.[1,6] Their origin is predominantly traceable to mucinous tumors of appendix (94%) followed by mucinous neoplasia from ovaries, gallbladder, stomach, colorectum, fallopian tubes, lungs, breast and urachus.[4,6] Approximately, 20% mucinous adenocarcinoma of appendix will evolve into PMP.[4]

The pathogenesis in the earlier 19th century was explained on the basis of three major theories as myxomatous peritonitis, foreign body peritonitis and as a metastatic theory.[7]Since then, multiple classification systems across the globe has evolved which employed similar terminology to describe the lesions of variable biological potential or dissimilar terms to describe the same entity creating controversies in the classification system. Three commonly used classification systems followed in 19th and early 20th century included: **The Ronnett classification as** Disseminated peritoneal adenomucinosis (DPAM), Peritoneal mucinous carcinomatosis (PMCA), Peritoneal mucinous carcinomatosis with intermediate/discordant Features (PMCA-I/D), **Bradley system as** Low grade and high grade mucinous carcinoma peritonei, and **American Joint Committee on Cancer (AJCC) and World Health Organisation (WHO) 2010 into** Low grade and High grade mucinous adenocarcinoma.[3,8,9,10]

With lot of controversies in the nomenclature system, only in 2016, a written consensus on PMP pathology classification and diagnostic terminology was published by the Peritoneal surface oncology group International (PSOGI). The latter-developed two taxonomies, the 2017 AJCC staging system and 2019 WHO classification of tumor are similar to PSOGI classification with minor modifications.[3]

PSOGI CLASSIFICATION (2016)	AJCC STAGING SYSTEM (2017)	WHO CLASSIFICATION OF TUMOURS (2019)
Acellular mucin: 1.Mucin without neoplastic epithelium 2.Confined to or distant from organ surface.	MIa	pMIa
Low Grade Mucinous Carcinoma Peritonei: 1.Low grade cytology 2. Few luminal mucinous epithelium (<20% of tumor volume) 3.Rare mitosis	MIb, G1, Well differentiated	pMIb, Grade 1: 1.Hypocellular mucinous deposits 2.Neoplastic epithelial elements have low grade cytology 3.Non-infiltrative type invasion
High Grade Mucinous Carcinoma Peritonei: Features of one or more of the following (At least focally). 1.High grade cytology 2.Infiltration of adjacent tissues 3.Invasion of vascular lymphatic vessels or surrounding nerves 4.Cribriform growth 5.Neoplastic mucinous epithelium (>20% of tumour volume) **Sub classification based on differentiation.** 1.Well differentiated 2.Moderately differentiated 3.Poorly differentiated	MIb.G2 or G3, Moderately or poorly differentiated	pMIb, Grade 2: 1.Hypercellular mucinous deposits as judged at 20x magnification. 2.High grade cytological features 3.Infiltrative type invasion characterised by jagged or stellated stands in a desmoplastic stroma or small cystic pool pattern with numerous mucin pools containing clusters of tumour cells
High Grade Mucinous Carcinoma Peritonei: With Signet ring cells Tumour with signet ring cell component (signet ring cell>20%)	MIb.G3,Poorly differentiated: Peritoneal Mucinous Carcinomatosis-With Signet ring cells	pMIb. Mucinous tumor deposits with signet ring cells

Table/figure2 Shows The Latest Pathological Classification And Terminology Of PMP by PSOGI (2016), AJCC (2017), and WHO (2019).

Even today, PMP is a challenging clinical entity with unique biological behaviour. Its clinical presentation is varied, ranging from an incidental finding on laprotomy to a ruptured appendix, peritonitis, or an ovarian mass, new onset hernia and intestinal obstruction in the advanced stage of the disease.[1,4,6]

Complete sampling of the appendix and the ovary have to be performed to know the origin of the tumour. Extra appendiceal spread will help to prognosticate the disease process in terms of recurrence and survival. High cellularity, presence of signet ring cells, destructive invasion, perineural invasion carries a bad prognosis.[4] Histopathologically detected acellular appendiceal mucin have to be clinically followed up for recurrences. Molecular studies have substantiated the origin with overexpression of MUC2, mucin produced by goblet cells of appendix as a specific marker of PMP.[4,6]

Incidentally detected mucinous ascites on laparotomy in surgical repair of hernia is another important presentation as seen in our case.[4] The cause of the death in cases of PMP is attributed to intestinal failure with increase in intra-abdominal pressure, fistula formation or infections.[3,4,6]

CT scan in PMP is detected as low attenuation mucinous ascites with possible septae or calcification in the paracolic gutters and pelvis with omental implants.[6] Our case showed diffuse mucinous ascites in the entire abdomen. In suspicious cases of appendiceal masses, colonoscopy and CT scan are done to confirm the diagnosis and to predict the success of treatment.

Serum tumour markers CEA, CA-125 and CA19-9 play a role in evaluating the degree of tumor invasion, ascites production and tumor burden and proliferation of cancer cells respectively. It is an invaluable tool in disease survillence and recurrence.[3,4]

Intraoperative evaluation of Peritoneal cancer index (PCI) scoring is used to evaluate the tumor load in the abdominal cavity. Higher PCI score is an independent prognostic factor.[3] This scoring guides the operating surgeon for the amount of removal of peritoneum and to achieve optimal cytoreductive surgery.

In the past, treatment of PMP was only palliative with decreased survival rate due to morbidity and mortality associated with the extensive surgery. Treatment of PMP follows the guidelines given by PCOGI 2016 and is best managed by cytoreductive surgery (CRS) combined with HIPEC and early post operative intraperitoneal chemotherapy (EPIC).[3]This new therapy has increased the chances of survival with reported 5 year survival rate ranging from 53% to 88% depending on the type of the tumor.[4]

CONCLUSION:

PMP is a clinicopathological entity with distinctive clinical presentation and broad spectrum of histological differentiation. Aggressiveness of the disease is reflected in the morphology and histological grade. The consensus guidelines to the classification, treatment of PMP will help the pathologist for standardised diagnostic reporting. A clear communication from the treating physician and adoption of a uniform reporting system plays an important role in decision making regarding its clinical management.

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