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A CASE REPORT OF PSEUDOMYXOMA PERITONEI IN A 33 YEAR OLD FEMALE

Pathology

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ABSTRACT

Pseudomyxoma peritonei is a rare clinical condition characterized by diffuse intraabdominal gelatinous or mucinous ascites on the peritoneal surfaces.We report a case of PMP in 33 year old female presented with acute abdominal pain for 1 day. USG and CT revealed inflamed appendix ; possibility of rupture with adjacent fluid collection and mild fluid collection in abdomen and pelvis. The patient underwent exploratory laparotomy and the histological features showing pools of acellular mucin, areas of fat with mixed inflammatory cell infiltrates and occasional glands showing atypical nuclei. Also IHC markers related to PMP studied show CK7 & CK20 positivity and SATB2 negativity favoring ovarian origin which is very rare to occur. PMP usually presents with distinctive clinical and pathological features ,always being a challenge in diagnosis and management of the patient. So,its important to make proper diagnosis , to provide appropriate treatment for better prognosis.

KEYWORDS

Pseudomyxoma peritonei(PMP), Ovarian mucinous neoplasm, Mucinous carcinoma peritonei, Tumor markers.

INTRODUCTION

Pseudomyxoma peritonei is a rare clinical condition characterized by diffuse intraabdominal gelatinous or mucinous ascites on the peritoneal surfaces [1,2,3,4,5,6].It was thought to be arising from dissemination of mucinous neoplasms of appendix, ovaries, fallopian tubes, stomach, small and large bowel, but the recent studies confirms that it can also develop from mucinous neoplasms of pancreas, lung, breast, gall bladder[1].It is said to have its incidence in 1 – 4 per million people with female preponderance[1,2,3,4].The pseudomyxoma peritonei exhibits quiescent behaviour, hence it is often discovered at advanced stages during laparotomy, laparoscopy or any radiological studies or have clinical presentation such as increased abdominal distension or sudden severe abdominal pain[1,2,3].

Case Report

We report a case of PMP in 33 year old female came with complaints of acute abdominal pain for 1 day which was insidious in onset, diffuse in nature and eventually progressed to unbearable pain with 2 episodes of vomiting.On examination,the patient was conscious, oriented with elevated blood pressure, increased pulse rate, febrile -100.9^{0}F.On abdomen examination,diffuse tenderness over the abdomen but severe in right iliac fossa and also rebound tenderness noted in right iliac fossa with percussion dull all over abdomen.On ultrasound abdomen and pelvis, the findings were inflamed appendix with periappendiceal fluid noted favoring ruptured appendix and also mild fluid collection in abdomen and pelvis.On CT abdomen findings were inflamed appendix with adjacent fluid collection.The patient was posted for emergency laparotomy.Grossly,we received single globular grey white to grey brown soft tissue mass. External surface of the mass was irregular and congested. On cut surface – cystic and solid areas noted.Also received separately sent grey white gelatinous fibrofatty soft tissue fragments.Histologically,H & E features show pools of acellular mucin with occasional areas of smooth muscle bundles and edema, areas of fat with mixed inflammatory cell infiltrates and occasional glands showing atypical nuclei.Tiny? edematous ovarian stroma also seen.

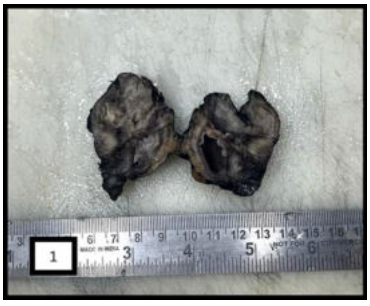

Figure 1: Grossly showing cut surface of single globular mass having both solid and cystic areas.

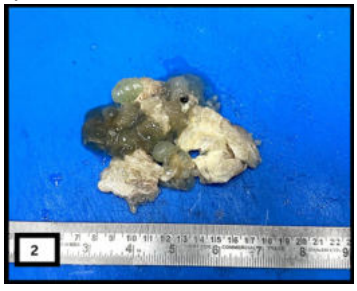

Figure 2 : Grossly showing sent grey white gelatinous fibrofatty soft tissue fragments.

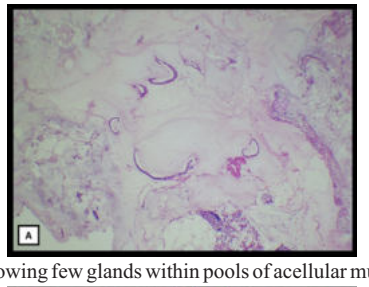

Figure A: showing few glands within pools of acellular mucin.

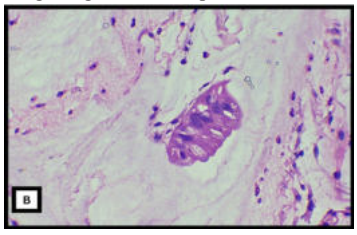

Figure B: showing gland with atypical nuclei.

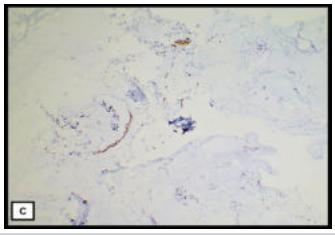

Figure C: showing CK7 membranous positivity pattern.

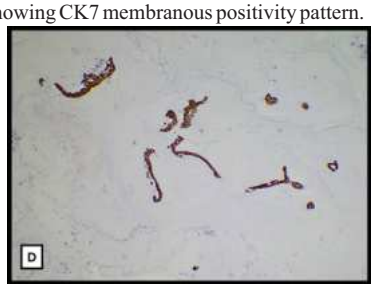

Figure D : showing CK 20 membranous positivity pattern.

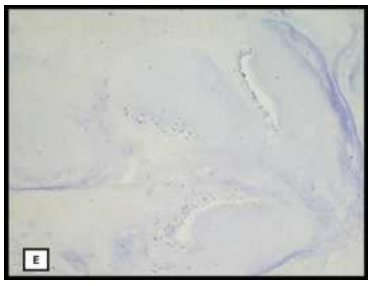

Figure E : showing SATB2 negative staining pattern.

DISCUSSION

Pseudomyxoma peritonei is a rare clinical entity characterized by diffuse intraabdominal gelatinous or mucinous ascites on the peritoneal surfaces producing 'Jelly Belly'[1,2,3,4,6] ; said to occur in 1- 4 out of million per year[1,2,3,4].It is often an incidental finding with female predominance and usually seen in average age between 40 to 55 years[1,2].Exact etiology is not known as it said to be arising from ruptured mucinous neoplasms of appendix, ovaries, fallopian tubes, bowels etc[1].Also genetic mutation with PMP includes KRAS mutations, GNAS mutations & rarely TP53 mutations ,genes related to PI3-AKT pathway [3,1].Clinically,since it has quiescent behaviour,it is likely to be an incidental finding in laparotomy,laparoscopy and in any radiological studies or the patient may present with symptoms depending upon the progress of the disease such as appendicitis like symptoms as localized disease or as an advanced disease like diffuse abdominal pain, abdominal distension, ascites[1,2].The pathogenesis of PMP includes several steps , initially the tumor cells of mucinous epithelium produces mucin continuously into the lumen forming a mucocele most commonly primary tumors of GI tract ; rarely the tumor ruptures.The tumor cells with mucin gets access to peritoneal cavity, gets implanted on various parts of peritoneal surfaces producing large amount of mucins leading to increased intrabdominal pressure. Very rarely lymphatic or hematogenic metastasis to be occurred.The extra peritoneal spread is very rare and mainly in pleural cavity[1,2,3].USG or CT is the preferred imaging study for PMP as it shows scalloping of hepatic surface and splenic surface with mucinous ascites[9,4,5].MRI is useful in assessing peritoneal metastasis[4].Macroscopically, they often show abundant gelatinous or mucinous material with minimal to maximal cystic epithelial deposits in the peritoneal surfaces.These tumor deposits may vary in size from small in mms to enormous size in cms [1].Histologically, in 1995 Ronnet et al. classified PMP as Disseminated peritoneal adenomucinosis (DPAM),Peritoneal mucinous carcinomatosis (PMCA) which was further divided into PMCA-I/PMCA-D)[2,4,5,6].Then in 2010,AJCC and WHO redefined the grading system for PMP. So now according to WHO Classification of tumors of peritoneum (5^{th} edition),instead PMP, it is termed as Mucinous Carcinoma Peritonei and the criteria for grading are 1)No grade if only acellular mucin present.2)Grade1 – mucin with hypocellularity.3)Grade2 -mucin with hypercellularity , marked cytological atypia >10 % of tumor or any angulated glands with desmoplastic stroma ,complex glandular growth or mucin with clusters of tumor cells of any origin.3)Grade3 – mucin with signet ring cells of primary tumor origin[8].IHC study for PMP includes CK7,CK20,MUC2,MUC5AC,CDX2 and SATB2. CK7 positive & MUC2 over expression if appendiceal origin.CK20,CDX2 & MUC5AC over expression if appendiceal or GI neoplasm origin[3,1,5,10]. SATB2 positivity seen if appendiceal or colorectal origin, negative in ovarian origin[7].The prognosis of PMP depends on the grading.The higher the grade ; the poorer the prognosis. Hence, it is really important to look for the clinical, radiological and pathological challenges in diagnosing PMP to provide appropriate treatment for the patients such as CRS/HIPEC[1].

In our case microscopically we observed acellular mucin with occasional glands showing atypical nuclei – Pseudomyxoma Peritonei grade 1 / Mucinous Carcinoma Peritonei Grade 1 and IHCs studied show CK7 – positive,CK20 – positive,SATB2 – Negative.With the above clinical,histopathological findings and SATB2 negativity favoring PMP of ovarian origin in our case.

CONCLUSION

PMP is a rare clinical condition characterized by diffuse intrabdominal gelatinous deposits on peritoneal surfaces. PMP has its distinct clinical and histopathological characteristics that always be a diagnostic challenge till today. Hence, its important to look for clinical, radiological and histopathological examination to arrive at a proper diagnosis at the earliest possible for appropriate treatment and better prognosis of the patient.

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