CASE REPORT

Incidental Finding of Pseudomyxoma Peritonei in a Patient Presenting with Umbilical Hernia.

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Abstract:

Background :

Pseudomyxoma peritonei (PMP) was first described by Rokitansky in 1842.[1] Werth described the association with ovarian tumors in 1884 and Franckel reported its association with appendiceal cystic tumours1 in 1901.

CaseHistory: Acase of peritoneal adenomucinosis in 32 year male patient who presented with umbilical hernia.

Key Words: umbilical hernia, pseudomyxoma peritonei, peritoneal adenomucinosis.

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Introduction:

Pseudomyxoma peritonei (PMP) is a poorly understood condition characterized by mucinous ascites and mucinous implants diffusely involving the peritoneal surfaces. There is considerable debate regarding the definition, pathology, site of origin, and prognosis of PMP.[2]

Case Report:

32 year male presented with a historyof umbilical hernia.There was no history of hematemesis or fever. On examination there was diffuse abdominal distension, it was tender however no abdominal mass was palpable. Swelling in umbilical region measuring 6 x 5 cm was present which was reducible and had positive cough impulse.Ultra-sonography revealed anterior wall defect at the level of umbilicus with small bowel loop as herniating content. With this provisional diagnosis of umbilical hernia the patient was posted for laparotomy.

Intraoperatively abundant mucinous material was seen coming out from peritoneal cavity. The peritoneal cavity was explored and found that it was involving whole of omentum. Approximately 3 litres of mucinous fluid was drained. The appendix was traced and found to be normal. A largemucinous mass with omentum was excised. No other organs were found giving origin for the mucinous mass and the abundant mucoid material. Post-operative period was uneventful.Resected tissue was sent for histopathological examination.

Gross:

A cut open umbilical hernial sac and huge omental mass was received. The hernial sac measured 6.5 x 5.1 cm and on cut section revealed multiple thin walled mucin filled cysts. Omental mass measured 50 x15 x 10 cm. and was composed of large amount of mucinous material covering the mass with grey white solid areas and multilocular cysts filled with mucin on cut section.The cyst had smooth grey white shiny inner surface.

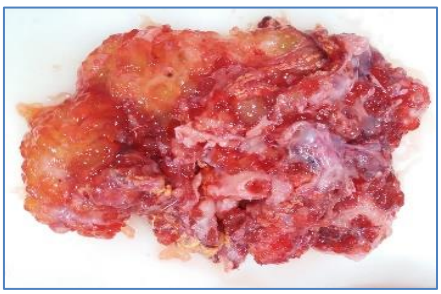


Figure:1

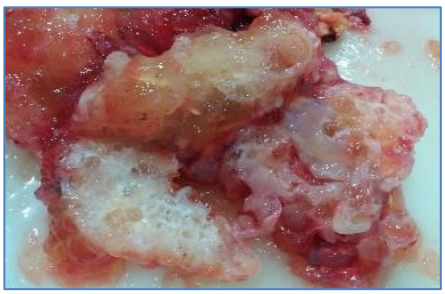


Figure:2

Figure1: Showing large mass with abundant mucoid material.**Figure 2:** Shows multiple uni and multilocular cysts containing mucoid material.

Microscopy: Sections from the cystwall and omental mass showed multiple cysts lined by low columnar to cuboidal epithelium and filled with mucin.

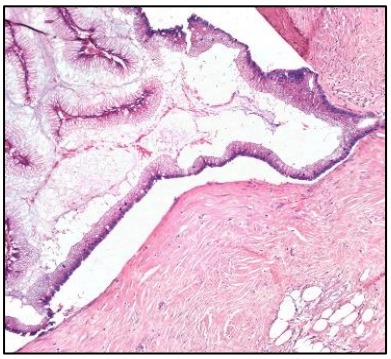


Figure: 3

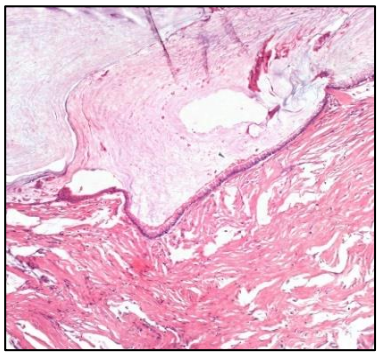
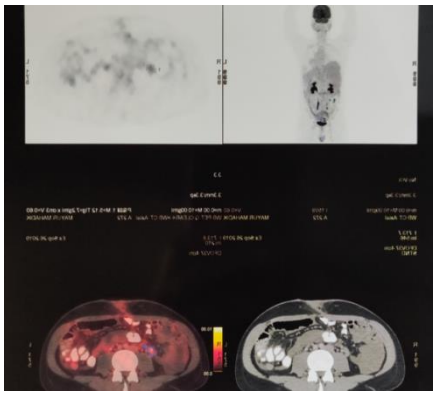


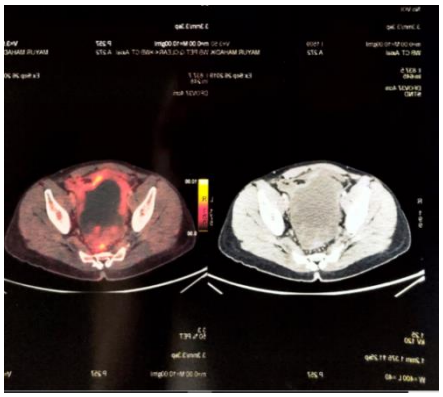
Figure:4

Figure 3 & 4: Show variably sized cysts lined by cuboidal to columnar epithelium with luminal mucinous material

Follow up:

Post-operative Pet scan was done which reported possibility of solid cystic pelvic lesion representing a neoplastic process with peritoneal dissemination, however no mention of appendix was made on Pet scan. Follow up IHC was reported as low grade mucinous neoplasms positive for CD 20, CDX 2 and CEA suggesting appendix as possible primary.Patient was comfortable on subsequent OPD visit.





Discussion:

The incidence of PMP, 2/10000 laparotomies, and its rarity make the study of the disease and its optimal treatment difficult. It is four times more common in women with median age of 54 years. The disease is progressive and is characterized by disseminated intraperitoneal mucinous tumors with peritoneal implants and mucinous ascites. The origin of PMP is controversial but most cases are associated with ruptured cystic adenoma of appendix and ovarian mucinous tumours.[1]This condition was originally described for benign ovarian cystadenomas and appendiceal mucocele.[3]According to some reports PMP also occurs in pancreatic carcinoma, carcinoma of breast and bile duct carcinoma.

According to Gough et al site of origin in their study was from appendix 52%, ovary 36%, colon 4%,endometrium 2% and pancreas 2%.3 It is more common in women than in men.[4,5] Common presentation is progressive abdominal pain, increased abdominal girth and weight loss. Patient may also present pain in lower abdomen on right side mimicking appendicitis.[4,6] Ronnet BM et al. analysed the 109 patients with pseudomyxoma peritonei and based on clinicopathological features divided it into three categories i.e., Disseminated Peritoneal Adenomucinosis (DPAM) , Peritoneal Mucinous Carcinomatosis (PMC) and PMCA with features intermediate between DPAM and PMCA or with discordant features. Disseminated Peritoneal Adenomucinosis (DPAM) was characterised by abundant extracellular mucin with scanty proliferative mucinous epithelium, showing little cytological atypia ormitotic activity. Peritoneal Mucinous Carcinomatosis (PMC), shows abundant mucinous epithelium with cytological feature so fcarcinoma.[4,2]Appendiceallesions seen with pseudomyxoma peritonei include mucosal hyperplasia, benign cystadenoma and cystadeno carcinoma. Primary ovarian cystadenocarcinoma or mucinous borderline tumors were earlier thought to bethesite of origin. Site of origin is controversial as both of organs are involved in the majority of female patients[4,7].

Several studies showed that ovariesare involved secondary to appendix origin which is supported as 25-50% of the patients are male and ovarian lesions are usually bilateral or mainly on right side with tumor implants were foundtobeon the surface rather than within ovarian tissue in75% of patients.[8,9] Apart from appendix and the ovary, rare sites of origin of pseudomyxoma peritonei includes colon, stomach, gall bladder,pancreas,urachus,urinarybladder,breast,fallopian tube, uterine corpus and lung.[2,4,10,12] Recently Bradley et al proposed a three tier grading system, which comprised of Grade I

lacking any significant adverse histologic features, Grade II high cellularity, high-grade cytology, or a high-grade invasion pattern (including the underappreciated “small mucin pool pattern”), Grade III with signet ring features.[13]

On sonography, pseudomyxoma peritonei may present as echogenic ascetic fluid with nonmobile echoesandechogenic septations. Echogenic masses secondary to omentum and parietal peritoneum involvement may also be seen. Bowel loopsare displaced centrally surrounded by echogenic ascitic fluid.[4,11] Scalloping of hepatic and splenic margins may also be present.[4]

Conclusion:

Pseudomyxoma peritonei commonly arise secondary to ovarian/appendix malignancy. These patients may require repeated removal of mucinous material. Knowledge of this rare condition is an important for early diagnosis and proper management. Despite its indolent, bland nature as a neoplasm, PMP is a debilitating condition that severely impacts quality of life.In diagnosing PMP, CT appears to be the most useful diagnostic tool. Diffuse loculations with mucin deposits exhibiting densities higher than non-mucinous or typical ascites are suggestive of PMP.

However in this case the appendix was normal and no other abdominal lesion which causes peritoneal adeno-mucinous was identified intra-operatively. Although there are case reports with varied clinical presentation this case presented with umbilical hernia which is not common presentation and hence reported.

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