ORIGINAL RESEARCH PAPER

Volume-7 | Issue-11 | November-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PSEUDOMYXOMA PERITONEI - A CASE REPORT AND REVIEW OF LITERATURE

Pathology

Radhika S* — Assistant professor, Department of Pathology, Karuna Medical College, Karuna Medical College, Chittur, Palakkad, Kerala – 678103.*Corresponding Author

Geetha N — Professor/HOD, Department of obstetrics and gynaecology, ESI Medical College Hospital, Coimbatore.

Karpagam Janardhan — Head of the department/Professor, Sri Ramakrishna Dental College and Hospital, Coimbatore. Consultant histopathologist, Histolab, Coimbatore 641 0037.

ABSTRACT

Background:

Pseudomyxoma peritonei (PMP) is a very rare tumour characterized by deposition of gelatinous mucin in the abdominal cavity and peritoneal surface of the abdominal viscera. It usually arises from primary mucinous neoplasm of the appendix. The primary lesion can also be present in colorectum or rarely urachus. According to WHO 2010, PMP is classified into low grade and high grade. Most common primary tumour is low grade appendiceal mucinous neoplasm (LAMN). The histological appearance can range from paucicellular lesion with bland cellular features to those exhibiting frank adenocarcinoma. This case is presented due to its rarity.

KEYWORDS

Pseudomyxoma peritonei, mucinous, appendix,

INTRODUCTION:

The term "pseudomyxoma peritonei" (PMP) was first described by Rokitansky in 1842.1 It is a clinical entity. The incidence 1-2 per million/year.2 The median age of presentation is 54 years. The term "pseudomyxoma" means pseudo- "false, lying", myx- "mucus" and oma- "process" or "action"/ tumour. Pseudomyxoma peritonei (PMP) is a very rare tumour characterized by deposition of gelatinous mucin in the abdominal cavity and peritoneal surface of the abdominal viscera. It commonly arises from the mucinous tumour of the appendix and occasionally from the ovary, colon, rectum, stomach, gall bladder, bile duct, small intestine, urinary bladder, urachus, lung, breast, pancreas and fallopian tube.3 The primary lesion can also be present in colorectum or rarely urachus. According to WHO 2010, PMP is classified into low grade and high grade. Most common primary tumour is low grade appendiceal mucinous neoplasm (LAMN). The histological appearance can range from paucicellular lesion with bland cellular features to those exhibiting frank adenocarcinoma. PMP can be classified into low grade and highgrade according to WHO 2010.4 The prognosis depends on the associated mucinous neoplasm. Its important to know the clinical features, classification, morphology and the immunohistochemical features of this entity. Whenever, there is suspicion of pseudomyxoma peritonei, it's is necessary that appendicectomy is done and the entire appendix is subjected to histopathological examination to arrive at a proper diagnosis.This case is presented due to its rarity.

CASE HISTORY:

55 year old female presented with complaints of abdominal distension since 2 months. History of appendicectomy 10 years back, reports not available.

O/E - bilateral pitting pedal edema present, P/A - 32 weeks flank full. Investigations showed elevated levels of CA 125 and CEA.

CA 125-43.1U/ml(0-35), CEA -136.2 (<5), CA19-9- 14.6u/ml(0-37)
CT abdomen and pelvis revealed a large cystic ovarian mass measuring 23.3x24x20cm, probably mucinous cystadenocarcinoma with omental involvement and moderate ascites, no enlarged lymphnodes in pelvis and abdomen.

Total abdominal hysterectomy with omentectomy and pelvic lymphnode dissection was done. Peritoneal fluid showed mesothelial cells and epithelial cells with atypia in a haemorrhagic background with mucinous material.

Gross examination of right ovarian cyst showed a smooth external surface covered with mucoid material. Sectioning showed a multiloculated cyst containing mucinous material. (Fig:1)

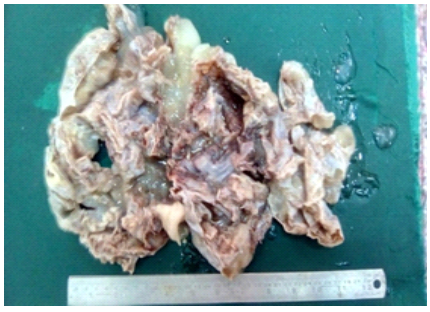

Fig:1 - Multiloculated cyst containing mucinous material.

Histological examination of right ovarian cyst showed multiloculated cystic spaces lined by columnar epithelium containing goblet cells and abundant extravasated mucin. (Fig:2-3) Omentum, peritoneal biopsy and pelvic lymphnodes showed mucinous deposits. (Fig: 4-5)

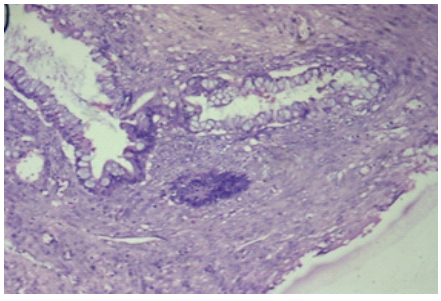

Fig:2 - Histology of right ovary showing mucin secreting tall columnar cells (4x)

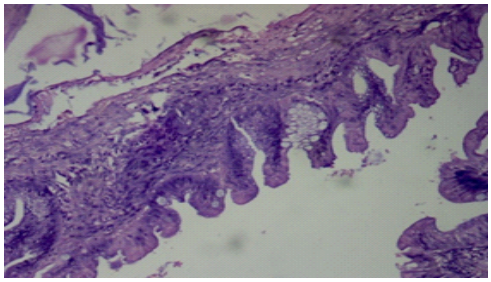

Fig:3 - Histology showing mucin secreting tall columnar cells with goblet cells(4x)

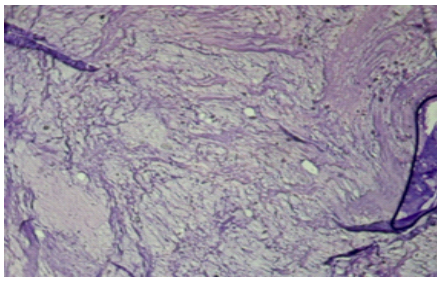

Fig:4 - Abundant extracellular mucin (10x)

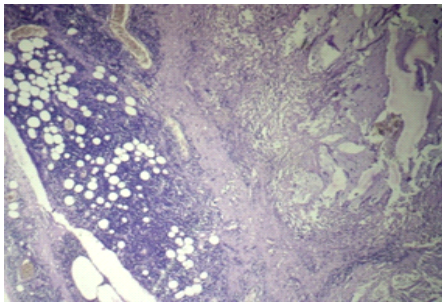

Fig:5 - Lymphnode showing mucinous deposits (4x)

IHC showed tumour cells positive for CK 20 and CDX2(Fig: 6-8) and negative for CK 7 and PAX 8. A diagnosis of metastatic mucinous adenocarcinoma was made and patient was started on chemotherapy FOLFOX.

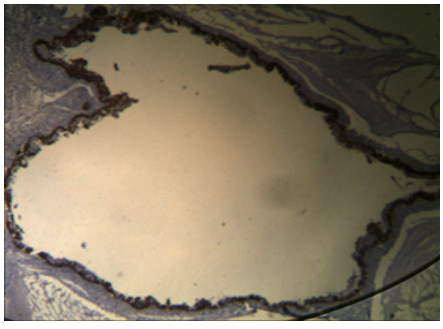

Fig:6 - Immunohistochemistry showing showing positivity for CK20 (10x)

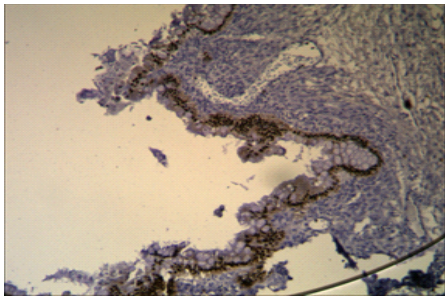

Fig:7 - Immunohistochemistry showing showing positivity for CDX 2 (4x)

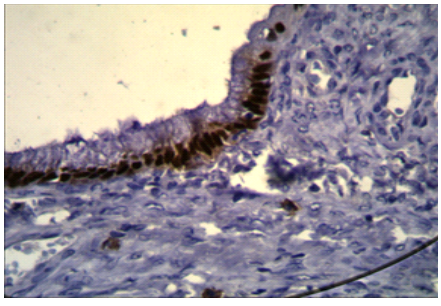

Fig:8 - Immunohistochemistry showing showing positivity for CDX 2 (10x)

DISCUSSION:

PMP is characterized by mucinous ascites and mucinous deposits involving the peritoneal surfaces. The incidence 1-2 per million/year. The median age of presentation is 54 years. The most common presentation is abdominal pain and abdominal distension.

CLASSIFICATION:

WHO 2010 classified pseudomyxoma peritonei into two types.
Low grade pseudomyxoma peritonei (peritoneal adenomucinosis) and High grade pseudomyxoma peritonei (peritoneal mucinous carcinomatosis)4

LOW GRADE PSEUDOMYXOMA PERITONEI:

Low grade PMP is characterized by mucin containing epithelium which exhibits only mild cytological atypia.5 In case of low grade PMP, cellularity is usually <10%) and shows nonstratified simple cuboidal epithelium.

HIGH GRADE PSEUDOMYXOMA PERITONEI:

High grade PMP is characterized by the presence of epithelium displaying moderate to marked atypia. Cribriform or signet ring morphology with desmoplastic stroma can be seen in high grade PMP.5

Most cases of PMP, have their primary lesion in the appendix. Rarely, the primary lesion can be seen in ovary, colon, rectum, stomach, gall bladder, bile duct, small intestine, urinary bladder, urachus, lung, breast, pancreas and fallopian tubes also.

Primary appendiceal lesions are classified as low-grade appendiceal mucinous neoplasm (LAMN) or mucinous adenocarcinoma.

LAMN is related to low-grade PMP, whereas mucinous adenocarcinoma is related to high-grade PMP.

IMMUNOHISTOCHEMISTRY:

Immunohistochemistry, to some extent can help in differentiating the origin of the primary lesion.

Primary ovarian lesions - CK7 +ve, CK 20-ve/weak focal positivity
Appendiceal primary – CK7 negative, CK 20 strongly positive
CK20& CDX2 positivity and CK7&PAX8 negativity favours primary from appendiceal origin.[6,7]

DIFFERENTIAL DIAGNOSIS :

The differential diagnosis of pseudomyxoma peritonei includes ruptured mucinous cystadenomas of appendix or ovary, soft tissue neoplasms with myxoid changes and endometriosis with myxoid change.

TREATMENT:

The gold standard treatment for PMP is complete cytoreductive surgery, followed by hyperthermic intraperitoneal chemotherapy. The prognosis depends on the associated mucinous neoplasm.9
In this case, there is history of appendicectomy, and only the immunohistochemistry helped in arriving at the final diagnosis.

PROGNOSIS:

Circulating tumour markers like CA 125, CEA and CA 19-9 have a prognostic value in PMP.[8]

The patients with normal marker levels have higher mean disease-free (DFS) and overall survival (OS) than those who had elevated levels of all three markers.

CONCLUSION:

Due to the low incidence of pseudomyxoma peritonei, its important to know the clinical features, classification, morphology and the immunohistochemical features of this entity. Whenever, there is suspicion of pseudomyxoma peritonei, it's is necessary that appendicectomy is done and the entire appendix is subjected to histopathological examination to arrive at a proper diagnosis.

REFERENCES:

1. O'Connell, Jerome T., et al. "Pseudomyxoma peritonei is a disease of MUC2-expressing goblet cells." The American journal of pathology 161.2 2002: 551-64.
2. Bevan, Katharine E., Faheez Mohamed, and Brendan J. Moran. "Pseudomyxoma peritonei." World journal of gastrointestinal oncology 2.1 2010: 44.
3. Carr, Norman J., and Leslie H. Sobin. "Unusual tumors of the appendix and

pseudomyxoma peritonei." In Seminars in diagnostic pathology, 1996:vol. 13, no. 4, pp. 314-25.
4. Carr, Norman J., et al. "Pathology and prognosis in pseudomyxoma peritonei: a review of 274 cases." Journal of clinical pathology 2012: jclinpath.
5. Carr, Norman J., et al. "The histopathological classification, diagnosis and differential diagnosis of mucinous appendiceal neoplasms, appendiceal adenocarcinomas and pseudomyxoma peritonei." Histopathology 71.6 2017: 847-58.
6. Strickland, Sarah, and Carlos Parra-Herran. "Immunohistochemical characterization of appendiceal mucinous neoplasms and the value of special AT-rich sequence-binding protein 2 in their distinction from primary ovarian mucinous tumours." Histopathology 68.7 2016: 977-87.
7. Harrison, M. L., C. Jameson, and M. E. Gore. "Mucinous ovarian cancer." International Journal of Gynecological Cancer18.2 2008: 209-14.
8. Baratti, Dario, et al. "Prognostic value of circulating tumor markers in patients with pseudomyxoma peritonei treated with cytoreductive surgery and hyperthermic intraperitoneal chemotherapy." Annals of surgical oncology 14.8 2007: 2300-308.
9. Wang, Huan, et al. "Clinicopathological features and prognosis of pseudomyxoma peritonei." Experimental and therapeutic medicine 7.1 2014: 185-90.