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Case Report

Challenges in cyto-diagnosis: a report of 2 interesting cases of pseudomyxoma peritonei

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ABSTRACT

Diffuse intra-abdominal gelatinous or mucinous ascites is commonly known as pseudomyxoma peritonei (PMP). It is a clinical entity rather than a histological diagnosis and usually results from peritoneal dissemination of mucus producing neoplasms most frequently from the appendix and rarely other GIT tumours. Since ascites may be the first symptom, cytologists may face diagnostic challenges in mucinous ascites. We present two cases of mucinous ascites in elderly females, in which thick, sticky, gelatinous fluid was received for cytological examination. After correlation of cytological examination findings with clinical and radiological features, diagnosis of disseminated peritoneal adenomucinosis was made which was later confirmed by fluid cell block with IHC studies.

Keywords: Pseudomyxoma peritonei, Immunohistochemistry, Gastro-intestinal tract, Cytokeratin

INTRODUCTION

PMP is a rare peritoneal malignancy. The annual incidence of PMP is 1-2 per million and more commonly seen in women.¹ PMP refers to intraperitoneal accumulation of mucin secondary to mucinous neoplasms of mainly appendix and far less common from ovarian mature teratomas. Werth introduced term PMP in 1884.¹ It usually involves abdominal cavity, peritoneal surface, omentum, external surface of abdominal and pelvic organs, ovaries and paracolic gutters. It is known by various other names like-Jelly belly and disseminated peritoneal adenomucinosis.³ Peritoneal mucinous carcinomatosis. Clinical presentation of PMP is diverse but the most common presentation is abdominal distension while many patients are asymptomatic and detected accidentally.²

CASE REPORTS

Case 1

A 53 years old female presented with complaint of pain in abdomen and distension since 6-7 months. Abdomen CT

scan findings were suggestive of ill defined multiloculated cystic lesions of size 10×10×13 cm predominantly arising from left adnexa with moderate ascites and multiple omental deposits seen. Past h/o appendicectomy present 2 years back. After ascitic tapping thick, viscous ascitic fluid was received for cytology. Microscopy revealed thick mucoid material in background (Figure 1 A), few groups and clusters of benign epithelial cells (Figure 1 B), mucophages and few reactive mesothelial cells (Figure 1 C and D). Clinicoradiological details and microscopic findings led to suspicion of PMP which later confirmed by fluid cell block with IHC markers. MUC1, CK7, CK20 IHC markers were applied. MUC 1 showed positivity (Figure 1 E). CK 20 positivity (Figure 1 F) and CK7 negativity (Figure 1 G) confirmed lower GI tract origin.

Case 2

A 51 years old female presented with abdomen distension since 4-5 months. Abdominal CT scan was suggestive of gross fluid in peritoneal cavity with multiple subtle ill-defined lesions in peritoneal mucosa and tiny

calcifications at places and septations probably representing PMP.

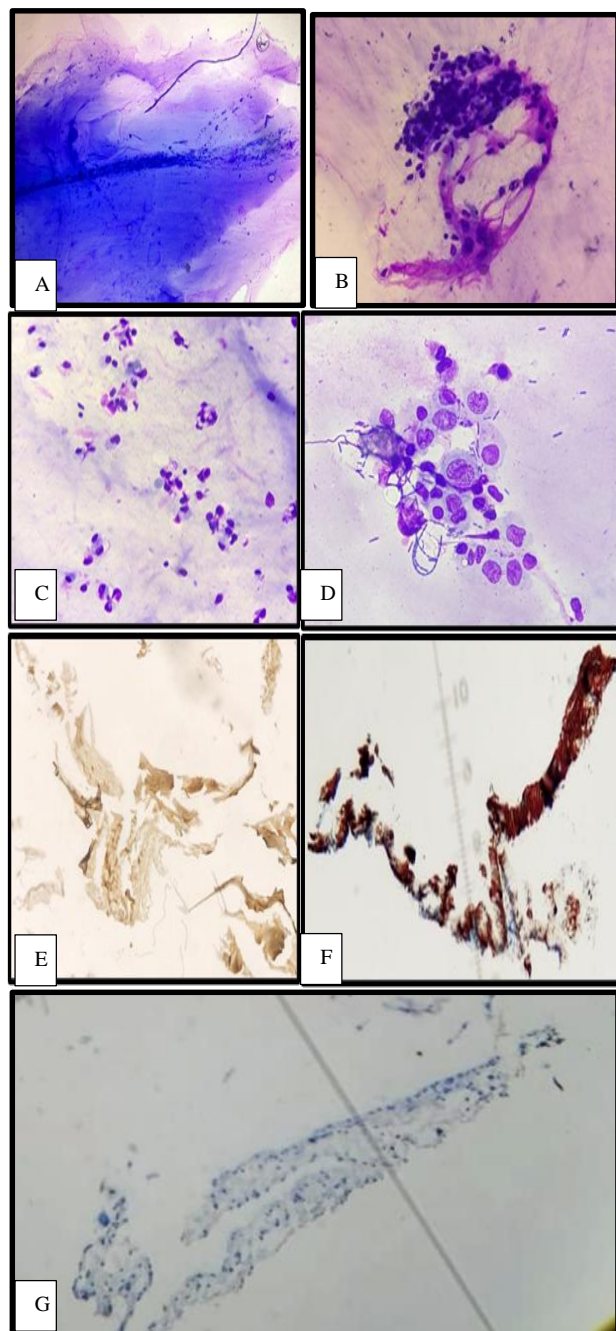


Figure 1 (A-G): Mucinous material (MGG-10x), epithelial cells (MGG-40x), mucophages (MGG-40x), mesothelial cells (MGG-40x), MUC-1 positivity, CK-20 positivity and CK-7 negativity.

Same as first case, a thick viscous jelly like ascitic fluid was received for cytology which also showed similar microscopic features like thick, mucoid material in background (Figure 2 A), few groups and clusters of benign epithelial cells (Figure 2 B), mucophages (Figure 2 C) on cytology. Same IHC markers were applied on fluid cell block in this case which again confirmed lower GI tract origin. In this case other IHC markers like SATB2,

PAX8 were also applied on tissue block. Grossly 1. Omentum with multiple grey white nodules and gelatin like material were received. 2. Ovary and salpinx aggregating to a size of 15×13×4.5 cm and showing solid cystic areas. Mucinous jelly like material seen in the cyst papillary excrescence. Glands were lined by single layer of cells and cells floating in mucinous pool (Figure 2 D and E). Histological features were of mucinous cystadenocarcinoma. SATB2 positivity (Figure 2 F) and PAX8 negativity (Figure 2 G) also confirmed appendiceal origin and ruled out primary ovarian origin.

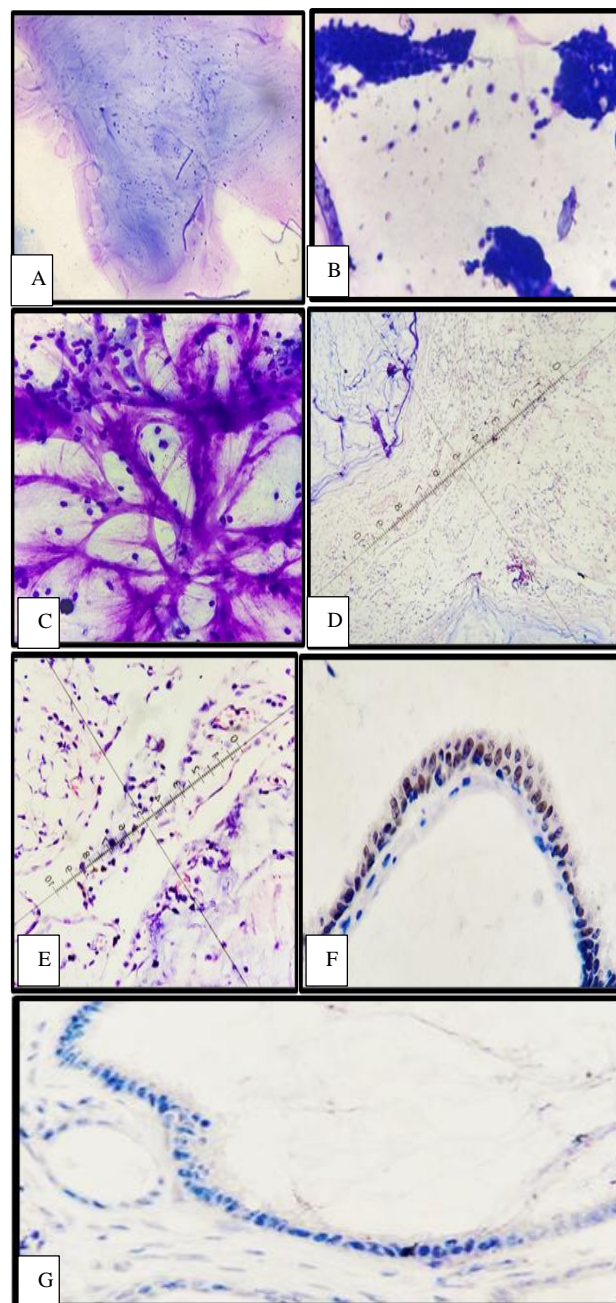


Figure 2 (A-G): Mucinous material (MGG-10x), epithelial cells (MGG-40x), mucophages (MGG-40x), cells floating in mucin pool (H and E10x), cells floating in mucin pool (H and E-40x), SATB2 positivity and PAX8 negativity.

DISCUSSION

There is controversy regarding pathogenesis of PMP and its primary origin in the cases of synchronous neoplasm in the appendix and ovary.³ PMP is a clinical term used to refer the presence of masses of jelly-like mucus in the pelvis and also in the abdomen.⁴ Mucinous neoplasms are mostly seen in adults in the fifth-sixth decade of life.⁵ Mostly all cases of PMP are of appendiceal origin and rare cases are of primary ovarian origin due to an appendiceal-type mucinous tumor arising within a mature cystic teratoma.⁴

In case 1 there was a past history of appendectomy and confirmed on cytology, cell block and IHC. In 2nd case PMP was suspected radiologically and confirmed on cytology, followed by histopathology and IHC. Cytomorphology of both cases showed epithelial clusters, mesothelial cells, mucophages and characteristic mucinous background. On microscopic finding, thick gelatinous fluid was a useful clue for the diagnosis and the microscopic mucinous background on cytology is the hallmark of PMP.³

According to the WHO digestive system tumors 5th edition grading system of peritoneal metastasis of appendiceal neoplasm hypocellular mucinous deposits and low-grade cytology are grade one and hypercellular mucin deposits with high grade cytological features are grade two.⁵ So, our 1st case was grade one and the 2nd case was grade two. In both cases, IHC CK7, CK20, MUC1 were applied. CK 20 and MUC1 showed positive results. CK20 positivity and CK7 negativity confirmed gastrointestinal tract (colorectum) origin.⁶ In view of ovarian mass in second case IHC markers SATB2 and PAX 8 were applied. SATB2 showed positivity which favoured gastrointestinal origin while PAX8 negativity ruled out ovarian origin. So PMP with both appendix and ovarian neoplasm should be treated as primary appendiceal origin.⁷ So non-invasive procedures like fluid cell block combined with IHC, contribute significantly in determining the primary site of tumor, thereby aiding in the accurate diagnosis and therapy.

CONCLUSION

Both cases highlight the role of clinical, radiological, cytological, and IHC findings in diagnosing PMP. Case 1

confirmed an appendiceal origin of PMP with IHC markers (CK20, MUC1 positive, CK7 negative) and low-grade mucinous deposits. Case 2, despite the presence of an ovarian mass, also pointed to an appendiceal origin, with IHC markers SATB2 positive and PAX8 negative, confirming gastrointestinal (appendiceal) origin. These cases demonstrate that non-invasive fluid cell block and IHC are crucial in determining the primary site and guiding treatment.

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