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Case Report

Primary retroperitoneal mucinous cystadenocarcinoma: a rare surgico-pathological entity

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ABSTRACT

Ovarian cancer (OC) is the seventh most common cancer for females in the world. Epithelial OC is the most predominant pathologic subtype (85%-90%), with five major histotypes- serous, mucinous, endometrioid, brenner and clear cell tumour. Each have three pathological subtypes ie benign, borderline and malignant. Mucinous cystadenomas and carcinomas of the ovary are well-established and common tumors. Primary retroperitoneal mucinous cystadenomas and carcinomas are being very rare and histopathogenesis of which is still uncertain. Most pathologists suggest their origin through mucinous metaplasia in a pre-existing mesothelium-lined cyst. An accurate preoperative diagnosis of these tumors is challenging due to lack of established effective diagnostic measures. Hereby presenting a 50-year-old woman, who visited to the hospital with abdominal distension and discomfort since two months. Sonography and computed tomography scans were performed and showed large predominantly cystic lesion arising from right adnexa. Patient underwent exploratory laparotomy for removal of the tumor. Histology and immunohistochemistry revealed primary retroperitoneal mucinous cystadenocarcinoma.

Keywords: Ovarian cancer, Primary retroperitoneal mucinous cystadenocarcinoma, Retroperitoneal tumours

INTRODUCTION

Primary retroperitoneal mucinous cystadenocarcinomas (PRMC) are very rare entities, with the first case was reported in 1965.[1] Since then, only 61 cases have been reported in the international literature.[2] Few cases have been reported in males and most of the cases are found in females.

PRMC remains unsuspected preoperatively due to the nonspecific symptoms and inability of radiology to determine the exact site of origin. The exact pathogenesis, behaviour, mode of treatment and prognosis remain controversial due to its rarity.

Authors herein report the case of a 50-year-old woman presented as abdominal mass. Following removal of mass by exploratory laparotomy, we could surgico-pathologically diagnose it as PRMC.

CASE REPORT

A 50-year-old woman reported to our hospital with a 2-month history of abdominal distension and discomfort. She had no past history of any surgery and medical co-morbidities. On per abdomen examination there was a palpable mass up to 36 weeks size with restricted mobility and having slight tenderness on right side. Bimanual examination revealed same mass on right side with uterus felt separately from mass and being deviated towards left side. Pouch of Douglas was smooth and rectal mucosa was free on per rectal examination.

The patient was in good general status. The tumor markers (CA 125, HE4 and CEA) were normal. ROMA being 16.7% in low risk range. The USG and CT scan (abdomin-pelvis) showed a cystic mass measuring 18.4×15.4 cm is seen arising from pelvis appear to be originating from right ovary. Superiorly extending up to level of Morrison's pouch. An enhancing solid area is seen along medial wall of size 6×5 cm. Right ureter compressed by mass and leading to moderate right hydroureteronephrosis. No Ascites or lymphadenopathy were seen. Uterus and left ovary were normal (Figure 1, 2).

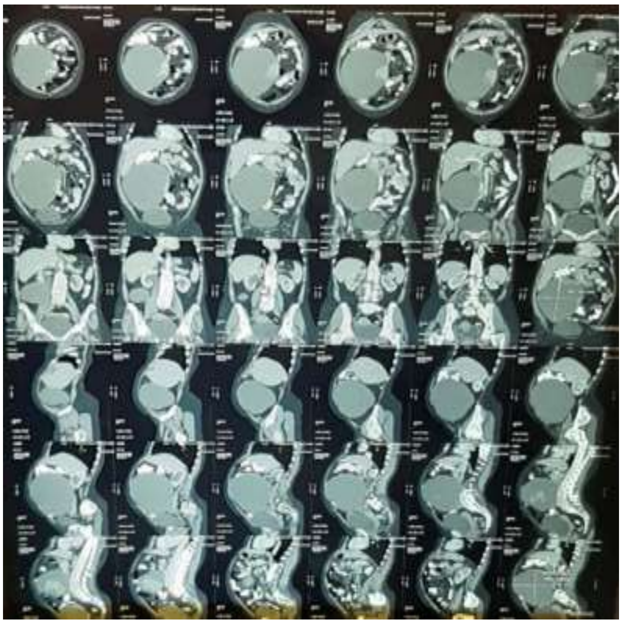

Figure 1: CT scan (abdomino-pelvis) of a cystic mass measuring 18.4×15.4 cm is seen arising from pelvis appear to be originating from right ovary. Superiorly extending up to level of Morrison's pouch. An enhancing solid area is seen along medial wall of size 6×5 cm.

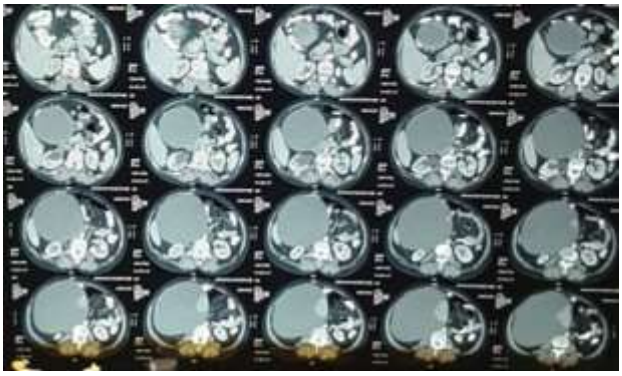

Figure 2: CT scan (abdomino-pelvis) - axial section of the mass in relation to adjacent structures.

The patient underwent exploratory laparotomy. Intra-operatively there was no Ascites. Uterus with its bilateral adnexa and appendix appeared normal and found separately from the cyst. After opening retroperitoneum on right side, ureter was identified which had moderate hydroureter. The ureter was then taken on umbilical tape. Approximately 18×20 cm cystic mass with solid area at its base was seen going behind the ileo-colic junction, small bowel mesentery and ascending colon reaching up to the subhepatic region. The cyst was densely adherent to small bowel mesentery and posterior surface of ascending colon. Intra-op assistance of oncosurgeon was taken. The cyst got ruptured intraoperatively during adhesiolysis and mucinous material came out. Cyst wall was separated from the right I-P ligament, ureter, posterior peritoneum and surrounding structures. Attempt to separate the adherent peritoneum from small bowel mesentery lead to its tear, which was repaired. The cyst wall was sent for frozen and report read as mucinous cystadenoma. Pan hysterectomy and appendicectomy was done. Exploration of abdomen was done at the end of the procedure as the mass was occupying whole of the abdomen. On exploration, no metastatic deposits or nodules were seen elsewhere in the abdomen. The GIT exploration done and no abnormal area was found. Patient's post-operative period was uneventful.

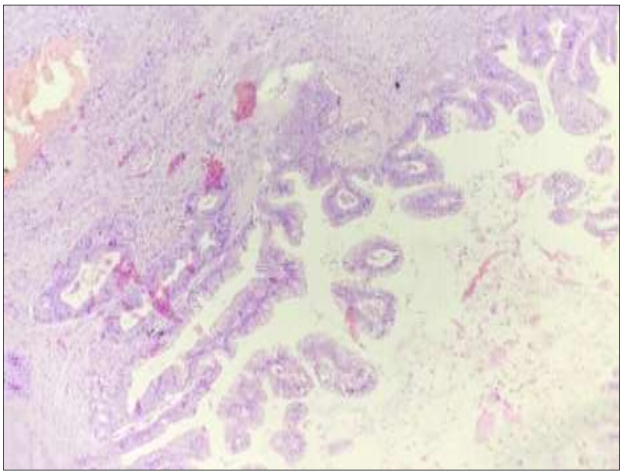

Figure 3: Presence of well differentiated mucous gland with cribriform and papillary architecture lined by mucin secreting columnar epithelium showing pleomorphic nucleus and prominent nucleoli. H and E stain X400.

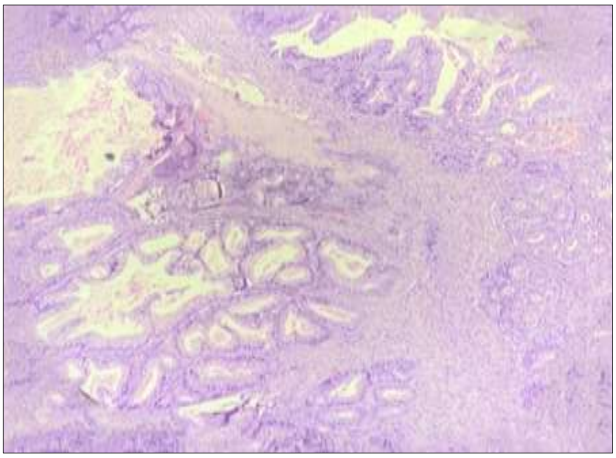

Figure 4: Cyst wall lined by single to pseudostratified malignant mucin secreting columnar epithelium with fibrous cyst wall. Ovarian stroma is not evident. H and E stain X40.

Final histopathology report suggestive of well differentiated mucinous cystadenocarcinoma from retroperitoneum (Figure 3, 4). Uterus, bilateral adnexa and appendix were unremarkable. Upper and lower GI scopies were done postoperatively to rule out GIT primary.

Immunohistochemistry markers like CK7, CK20, CDX2, CEA, PAX-8 (focal) were positive. Since both ovaries are grossly and microscopically free of any tumor, as well as intraoperative findings are suggestive of retroperitoneal origin mass (ovaries normal, appendix normal), features favor mucinous adenocarcinoma of retroperitoneum. positivity of above All IHC markers may be due to secondary mullerian system manifesting its existence in retroperitoneum and show metaplastic and neoplastic lesions that are analogous in all regards to those commonly found in ovary, uterus or other organs of female genital tract.

DISCUSSION

Retroperitoneal tumors constitute about 0.2% of all neoplasms.[3] Primary retroperitoneal mucinous cystadenoma and carcinoma is very rare tumors, as retroperitoneum lacks epithelium. They can be found in any part of the retroperitoneum without attachment to the ovary. The first case of carcinoma with mullerian type epithelium was reported by Roth and Ehrlich.[4]

The pathogenesis and the site of origin of this uncommon entity are ambiguous, with various theories being postulated. As indicated by one hypothesis, ectopic ovarian tissue in the retroperitoneum is the origin. This is supported by the morphological, histochemical and immunohistochemical similarities between PRMC and mucinous ovarian cystadenoma. Subramony et al study showed that the estrogen receptors were present in stromal cells of a PRMC.[5] An IHC analysis concluded that PRMC was similar to ovarian mucinous tumor, as it was positive for CK7 and CK20 antibodies. The other theory is, these tumors are arising from an invagination of the multipotential mesothelium with subsequent mucinous metaplasia of the mesothelial lining cells, which leads to a mucinous cyst with other malignant phenotypes.[6] As seen in epithelial ovarian tumor, peritoneal epithelium has the potential for mullerian differentiation has been shown in other studies.[7] According to some study, mucinous tissue overgrowing other components of a teratoma. The retroperitoneal masses can be classified into 3 pathological types: cystadenoma, borderline cystadenoma and cystadenocarcinoma.

Preoperative diagnosis is important, because retroperitoneal masses are mostly malignant. It’s challenging as there is lack of established effective diagnostic modalities.[3] PRMCs can be found anywhere in the retroperitoneal space and usually presents as multi or unilocular cystic mass of varying size. Radiological imaging, such as USG, CT and MRI helps in describing and assessing the disease characteristics and its involvement with adjacent and distant structures, but malignant potential cannot be excluded. The utility of serum tumor markers (CA-125, CA 19-9, CEA, CA 15-3 and αFP) in the diagnosis or follow-up is questionable.[8] Intra-operative finding and final histopathology helps in the diagnosis.

PRMC must be differentiated from other cystic masses, benign or malignant. Benign cysts include lymphocele, cystic lymphangioma and pancreatic pseudocyst. Malignant tumors include cystic teratoma, pseydomyxoma retroperitonei, cystic mesothelioma, renal and ovarian tumours. The metastatic mucinous tumors from ovaries, intestines, and pancreas have to be excluded.

Surgery is the cornerstone of management. Intact Radical removal of the tumor is the standard therapy. According to Gotoh et al survival is improved if oopherectomy is done.[7] Lee et al did total hysterectomy along with oopherectomy.[9] Kessler et al suggest that if uterus and ovaries are grossly normal and on the grounds that follow-up in most of revealed cases is inadequate, so panhysterectomy can be excluded for the treatment of PRMC.[10] This procedure should be done in patients who have completed their family and are postmenopausal. Law et al advised that removal of the tumor laparoscopically and exploration of abdomen and pelvis and fertility sparing surgery as management in these patients.[11]

Adjuvant chemotherapy following surgery is controversial, as no definite therapeutic guidelines have been laid. Role of adjuvant chemotherapy is debatable and should be given if there is intra-operative spill, or when there is involvement around the tumor or distant metastasis.[10] Another reason for adjuvant treatment is that primary retroperitoneal mucinous cystadenocarcinomas and ovarian mucinous tumor have same histogenesis.[12] As there was intra operative rupture of cyst, this study patient received adjuvant chemotherapy.

CONCLUSION

PRMC is an extremely rare tumor and have an aggressive potential for recurrence. Preoperative diagnosis of PRMC is difficult so large retroperitoneal masses should be kept as a differential diagnosis. Intact Radical removal of the tumor is the mainstay therapy and the most important prognostic tool. The role of lymphadenectomy or adjuvant chemotherapy is still controversial. The long-term management following surgery is still evolving. Due to its rarity and availability of short follow-up of the patients in literatures, the prognosis of these tumors remains uncertain. This case is being presented to emphasize on the rarity of PRMC. Further studies are needed to explain the etiology and effective management.

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