Massive Mucinous Malignant Cystadenoma – Excision in Toto and Follow up

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Abstract
Mucinous mesenteric neoplastic cysts are extremely rare extra-ovarian cysts that should be considered as a differential diagnosis in women with large abdominal cysts of unknown origin. Complete excision is the only management treatment due to their malignant potential.

Introduction
Mucinous mesenteric cysts are extremely rare, have a female preponderance, and are difficult to diagnose preoperatively. They are regarded as benign with malignancy only reported in three case reports (3%) [1]. Complete excision is the only treatment option for all mesenteric neoplasms because of their malignant potential [2].

Case Presentation
A 26-year-old nulliparous woman attended routine gynaecology clinic with a five month history of weight gain associated with pelvic pain which was worse on the left and associated with deep dyspareunia. There were no associated urinary or bowel symptoms. She had no past medical or surgical history. Clinical examination revealed a palpable pelvic mass above the umbilicus.

Investigations
An abdominal ultrasound (US) demonstrated a simple cyst in the mid abdomen, extending from the upper abdomen to the pelvis measuring 36 x 17.8 cm. The origin of the cyst was unknown. Both ovaries and uterus were normal. Therefore a computerized tomography (CT) abdomen and pelvis was organized. The CT showed appearances of a large benign unilocular cyst that extended from the pelvis into the abdomen. This was thought to be probably ovarian in origin due to its proximity to the left ovary (Figure 1). CT scan appearance of cystic structure extending from pelvis close to the left ovary. Tumour markers Ca125, CEA and Ca19-9 were all within normal limits.

Differential diagnosis
A benign ovarian cyst was considered but as no definite diagnosis could be established a diagnostic laparoscopy was undertaken.

Treatment
At laparoscopy a large mesenteric cyst was identified arising from the transverse mesocolon, extending superiorly to just below the transverse colon itself. The ovaries and uterus were completely normal. Peritoneal washings were taken and the decision at the time was not to proceed to excision of this cyst of unknown pathology as this could not be achieved laparoscopically and...
possible oncological resection could be compromised. At clinic review, peritoneal washings did not demonstrate any evidence of malignant cells. The operative findings were discussed and the patient consented for laparotomy and excision of cystic mass with possible colectomy due to the proximity to the transverse colon and its mesentery. At laparotomy the cyst was found to be contained within the transverse colon mesentery, with no firm adhesions or shared blood supply. It was enucleated in toto. Macroscopically the cyst had a thin and smooth wall and contained serous coloured fluid (Figure 2). Intraoperative photograph of cyst following enucleation of the mass from the transverse colon mesentery.

Outcome and follow-up

Histology demonstrated a thin walled cyst which was predominantly lined by a single layer of mucinous epithelium. In one area the epithelium was stratified and pleomorphic with increased mitotic activity and complex glandular pattern consistent with intra-epithelial carcinoma. There was no evidence of invasion. The epithelium was strongly positive with cytokeratin-7 (CK7) and Ca125. The dysplastic area showed intestinal differentiation and was CK20 and CDX2 positive. Spindle cell stroma was found focally within the cyst wall, reminiscent of ovarian stroma. The overall features were consistent with a mucinous cystadenoma with intraepithelial carcinoma confined to the cyst epithelium (Figure 3). Low power histopathology image showing normal epithelium changing to darker abnormal epithelium.

The patient made a good recovery and subsequent follow up investigations of Year 1 and Year 3 (interval) CT scans have not shown any evidence of disease recurrence or metastatic spread. She also had a transvaginal ultrasound scan, colonoscopy and gastroscopy which did not identify a primary cause, however gynaecological origin was likely.

Discussion

Mucinous cystic neoplasms (MCNs) arise in the ovary and various extra-ovarian sites. Both ovarian and extra ovarian MCNs share histological similarities and therefore a common pathway of development has been suggested [3]. There are only fifteen reported cases of mesenteric MCNs in the literature, with the majority being benign Cystadenoma [1]. Malignancy was found only three cases: one Cystadenocarcinoma [4], one report of incomplete excision of cystadenoma leading to malignant transformation [5] and one describing the development of pseudomyxoma peritonei following rupture of a mesenteric cyst [2]. A greater median age of presentation, observations of malignant transformation from benign neoplasms, and focal occurrence of benign, focally borderline and malignant epithelium suggests possible progression from adenoma to carcinoma. Therefore mucinous cystic neoplasms should be approached as a potential malignancy and complete excision is the only treatment.

Learning Points/Take Home Messages

- Extra-ovarian mucinous cystic neoplasm must be considered as a differential diagnosis in women with cystic masses of unknown origin.
- Although predominately benign, there is malignant potential which dictates resection of the complete cyst. Malignant growth can only be determined histologically.
- Excision of the cyst without rupture is the only treatment for mesenteric cystic neoplasms.

References