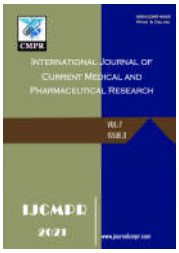
INTERNATIONAL JOURNAL OF CURRENT MEDICAL AND PHARMACEUTICAL RESEARCH
ISSN: 2395-6429, Impact Factor: 4.656
Available Online at www.journalcmpr.com
Volume 7; Issue 03(A); March 2021; Page No.5641-5643
DOI: http://dx.doi.org/10.24327/23956429.ijcmpr202103980



Research Article

A RARE CASE OF SYNCHRONOUS GIANT MUCINOUS CYSTADENOMA OF LEFT OVARY WITH MUCINOUS CYSTADENOMA OF APPENDIX

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ARTICLE INFO

Article History:
Received 10th December, 2020
Received in revised form 2nd January, 2021
Accepted 26th February, 2021
Published online 28th March, 2021

Key words:
Mucinous Cystadenoma, mucocele, appendix, mucinous neoplasms of ovary

ABSTRACT

Primary mucinous cystadenoma of the appendix and ovary are commonly encountered in routine histopathology practice and several published classification schemes for appendiceal mucinous neoplasms have been proposed with resultant inconsistent use of terms and clinical doubt. While the nomenclature for ovarian mucinous neoplasms is more settled, the difficulty lies while distinguishing synchronous mucinous tumours of appendix and ovary to whether the primary is ovary or appendix or have a common origin. The synchronous or metachronous occurrance of multiple primary carcinomas in the genital tract is well recognized. However, simultaneous occurance of mucinous ovarian and appendiceal tumors is very rarely seen. Here, we report a rare case of giantmucinous cystadenoma of ovary and synchronous mucinous cystadenoma of the appendix in a 45 year old lady who presented with a large left abdomino-pelvic mass.

INTRODUCTION

Mucinous neoplasms of the ovary account for 10%-15% of all ovarian neoplasms. They may be benign, borderline, or malignant with large majority constituting benign and borderline, that accounts nearly75% and 10% respectively of all mucinous ovarian neoplasms.[1]

The clinical manifestation is nonspecificbutmost mucinous ovarian neoplasms present as large unilateral pelvic masses. The synchronous or metachronous occurrence of multiple primary cancers in the genital tract is well recognized. The overall incidence of synchronous female genital tract malignancies is 0.63% with most frequently observed synchronous neoplasms being those of the ovary with the endometrium.[2]

Mucinous cystadenoma of the appendix occur rarely and are most often incidentally discovered, mostly present in the form of a mucocele. Mucocoele of the appendix is an obstructive dilatation of the appendiceal lumen due toexcessive mucus accumulation resulting in cystic dilatation of the lumen. It may be related to a variety of pathological conditions.

The coexistence of mucinous ovarian and appendiceal tumors in association with pseudomyxoma peritonei is well established.[2] However, considerable debate has been there regarding the origin of the tumor in such cases. The importance of this case report lies in the fact that we describe two truly independent synchronous primary mucinous tumorsinvolving the appendix and ovary with absence of any evidence of pseudomyxoma peritonei. Metastatic mucinous tumors, particularly those of gastrointestinal or appendiceal origin, should be excluded when primary mucinous carcinoma of the ovary is considered.

We report an unusual case of 45 year old lady who presented with coexisting large ovarian mass with an independent primary appendicular mass without any evidence of pseudomyxoma peritonei.

Case Presentation

A 45-year-old Hepatitis C positive lady presented with large left sided abdomino-pelvic mass noted since few months. Ultrasonography of abdomen revealed an ill-defined semi solid heterochoic mass lesion seen to be arising in urinary bladder with space occupying lesion in pelvis. CECT of abdomen and pelvis revealed a large abdomino-pelvic mass measuring 35.5x18.6x13.9cm with multiple septae and multiloculation causing compression on adjacent structures likely arising from left adnexnal complex cyst suggestive of ovarian cystadenoma. No mural nodule/calcification/fat seen. No ascites was observed. Serum markers CA-19.9 was 1300 U/ml and CEA was 750 ng/ml. Leftsalpingoopherectomy was performed and specimen was sent for intraoperative frozen section examination.

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Pathological findings

We received an already opened up single left sided large globular mass with smooth surface and focal areas of hemorrhage and prominent vessels running on the surface**.** (Figure 1a) Inner surface was multiloculated with thick yellowish gelatinous mucinous material. (Figure 1b) Cyst wall thickness varied from 0.2 to 0.8 cm. No solid areas were seen. Attached tubal tissue measured 4x1cm.

Frozen sections from ovarian massshowed features of benign mucinous cystadenoma. Paraffin sections showed a cyst wall lined by cuboidal to tall columnar with intracytoplasmic mucin vacuoles. (Figure 1c) Few cystic spaces were seen lined by flattened epithelium containing amorphous eosinophilicmaterial. Underlying fibrocollageous tissue shows chronic inflammatory infiltrate. No nuclear stratification/ atypia seen. Uterus and cervix showed proliferative glands and chronic cervicitis respectively. Gross examination of appendix showed enlarged (Figure 2a) and cystically dilated lumen filled with white jelly like material.(Figure 2b) Appendix measured 5x3.5cm showing smooth external surface with foci of hemorrhage. Sections from appendicular cyst showed complete replacement of epithelium lining by cuboidal to tall columnar cells with mucin vacuoles at places showing nuclear stratification upto four cell layers.(Figure 2c) The underlying wall was thinned out and showed fibrosis. A histopathological diagnosis of Mucinous Cystadenoma of Ovary and Appendix was made.

DISCUSSION

Ovarian mucinous cystadenomas are benign epithelialtumors characterized by itsunilateral presentation, multilocularity, smooth outer and inner surface that tend to attain large size and contain mucinous fluid. Mucinous tumors comprise 12% to 15% of all ovarian tumors and almost 75% of all mucinous tumors are benign, 10% are borderline and 15% are invasive carcinoma.[3]

Mucocele of appendix constitute about 0.2-0.3% of all appendectomies. It can be caused by a variety of non-neoplastic and neoplastic pathological conditions of the appendix like mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma (11%), and rest as retention cyst.[4] Mucinous cystadenomas of appendixshow higher frequency in women compared to men with a M:F ratio of 4:1,commonly in patients over 50 years of age.[5]Most of the benign mucoceles including mucinous cystadenomas are asymptomatic and are mostly incidentally detected during ultrasonography, computed tomography and other radiographic examinations of gastrointestinal tract or during a laparotomy.

Unlike the plethora of terms for mucinous lesions in the appendix, the terminology and nomenclature for primary ovarian mucinous tumours is settled that includes: mucinous cystadenoma, mucinous borderline tumor andinvasive mucinous carcinoma. Although the histological criteria and nomenclature for primary mucinous ovarian tumours appears to be well defined and standardised, excluding a metastatic lesion from a primary ovarian mucinous neoplasm still remains a major diagnostic challenge. Appendiceal tumours are typically low-grade mucinous neoplasms and oftentimes do not show obvious invasion.

Seidman *et al*. studied concurrent mucinous ovarian and appendiceal tumorsand found that immunoperoxidase reaction for four epithelial antigens revealed complete concordance between ovarian and appendiceal lesions in only five of the totalof 15 cases.[6]

Chuaqui *et al*. using genetic analysis of microdissected tissue samples concluded that some synchronous mucinous tumors of the ovary and appendix were appendiceal in origin and others represented independent primaries and there was independent origin of the ovarian and appendiceal tumors in most cases and did not favour an origin in a single site but a multi-focalneoplastic process.[7]

In addition, several criteria have been advanced to help distinguish primary ovarian mucinous tumors from metastatic mucinous tumors. A large size, unilaterality, presence of benign and borderline areas, expansile invasion pattern, smooth surface and absence of extraovarian disease, all favour a primary ovarian neoplasm. By contrast, bilateral ovarian involvement, smaller size, ovarian surface involvement, multiple nodules and an infiltrative pattern of stromal invasion favour an extraovarian origin. However, there are cases that do not follow the aforementioned patterns and thus pose significant diagnostic difficulty. This is seen particularly in cases with a primary appendiceal mucinous neoplasm. Our patient also presented with a large unilateral multicystic ovarian mass without surface involvement.

The most commonly used markers to better characterise the origin of ovarian mucinous tumours are CK7, CK20 and CDX-2.Primary mucinous tumors of ovarian origin demonstrate CK7 and CK20 positivity. If CK7 is negative, CK20 and CDX-2 is diffusely positive a colorectal origin is favoured.[8] In the available literature, synchronous tumors of the ovary and appendix are an uncommon yet well-recognized occurrence in the setting of pseudomyxoma peritonei. Most common ovarian tumors in these patients are mucinous cystadenomasor borderline mucinous tumors and the associated appendiceal neoplasms are mucinous cystadenoma.[9]

The origin of such synchronous tumors is widely debated with most evidence favoring a primary appendiceal tumor with the ovarian tumor representing a metastatic process. However in our case patient had no evidence of pseudomyxoma peritonei. In conclusion, the simultaneous occurrence of synchronous primary cancers in female genital tract is well known, but most of them are malignant in nature. In the present case, however, the presence of mucinous cystadenoma in one ovary and mucinous cystadenoma in appendix was observed. Hence based on clinical and histopathological features, this case can be regarded as an example of two truly independent primary mucinous tumors involving the appendix and ovary.

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How to cite this article:

Monal Trisal *et al* (2021) 'A Rare Case of Synchronous Giant Mucinous Cystadenoma of left ovary With Mucinous Cystadenoma of Appendix', *International Journal of Current Medical and Pharmaceutical Research,* 07(03), pp 5641-5643.

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