10.5005/jp-journals-10006-1637

CASE REPORT

A Rare Case of Primary Mucinous Adenocarcinoma of the Appendix Presenting as Ovarian Mass: A Diagnostic Challenge

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ABSTRACT

Introduction: Cancer of the appendix is an uncommon disease and is typically identified incidentally during appendicectomy. Bilateral ovarian metastases may occasionally occur in mucinous adenocarcinomas of appendix mimicking primary ovarian malignancies and can present a diagnostic challenge. The prognosis is poor as it is usually diagnosed at an advanced stage, either due to a low threshold of suspicion and also due to difficulties in diagnosis before surgery. Here we are reporting such a case of occult appendiceal adenocarcinoma which presented clinically as primary ovarian malignancy. Staging laparotomy revealed large bilateral ovarian tumors of FIGO Stage III, with presumed appendiceal implants. Histological examination revealed transmurally infiltrating Mucinous Adenocarcinoma of the appendix with mesoappendiceal invasion and serosal involvement, with lymphovascular invasion with secondaries to bilateral ovaries and omentum. Immunohistochemical staining revealed positive for CK 20 and CDX2 and negative staining for CK 7. The diagnosis of a primary mucinous adenocarcinoma of Appendix with bilateral ovarian metastasis was confirmed with pT4aG2pNxpM1c; TNM stage IV. The patient is planned for right hemicolectomy, total peritonectomy and hyperthermic intraperitoneal chemotherapy (HIPEC).

Conclusion: Although adenocarcinoma of the appendix is uncommon, they should be considered in the differential diagnosis of intraabdominal masses, as the treatment modalities vary.

Keywords: Appendiceal malignancy, Ovarian metastasis, Ovarian carcinoma, Primary carcinoma of appendix.

How to cite this article: Sehagal P, Vijay A, Sharma K. A Rare Case of Primary Mucinous Adenocarcinoma of the Appendix Presenting as Ovarian Mass: A Diagnostic Challenge. J South Asian Feder Obst Gynae 2018;10(Suppl 2):425-428.

Source of support: Nil

Conflict of interest: None

Date of received: 10 October 2017

Date of acceptance: 5 May 2018

Date of publication: July 2019

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BACKGROUND

Cancer of the appendix is an uncommon disease and is typically identified incidentally during appendicectomy. Carcinoid is the commonest histological type with mucinous adenocarcinomas accounting for 66% of appendiceal cancers.[1] Bilateral ovarian metastases may occasionally occur in mucinous adenocarcinomas of appendix mimicking primary ovarian malignancies and can present a diagnostic challenge.[2] Here we are reporting such a case of occult appendiceal adenocarcinoma which presented clinically as primary ovarian malignancy.

CASE DESCRIPTION

A 68-year-old lady, para 4 live,[3] postmenopausal, presented to the gynecology OPD with acute urinary retention and abdominal distension of 15 days duration. She also complained of constipation, occasional vomiting, loss of appetite and weight. Abdomino-pelvic examination revealed a firm to the cystic, irregular, nontender mass of 35 × 25 cm, arising from pelvis and extending up to xiphisternum.

Abdominopelvic ultrasonography and contrast-enhanced computed topography revealed a large multilocular cystic lesion of size 33 × 25 × 17 cm, with multiple internal septations, in lower abdomen with minimal vascularity and moving internal echoes. There was no evidence of a solid component or ascites. Evidence of left ureteric compression and left hydro-uretro nephrosis was present. No obvious appendiceal lesion was noted. Tumor markers like CA-125 and CA19-9 were normal with elevated CEA levels (131.28 ng/mL). Endoscopy and colonoscopy were done which showed a normal study.

Based on the clinical examination and investigations, a pre-operative risk of malignancy was evaluated using the risk of malignancy index (RMI) and International ovarian tumor analysis-assessment of different neoplasia of the adnexa (IOTA-ADNEXMODEL). The calculated values showed low-risk for malignancy (RMI-41.7, Iota-Adnex Model–LR 1-0.25, LR 2-0.049).

With all the above findings and observation, a provisional diagnosis of an ovarian tumor with low risk for malignancy was made. A staging laparotomy with prior bilateral ureteric stenting was performed which revealed thick yellowish mucinous material on the

opening the peritoneal cavity. Bilateral ovarian masses seen–measuring 30 × 20 cm on the left side and 20 × 15 cm on the right side (Fig. 1). Capsule was found ruptured with mucinous material oozing out of the cyst. Macroscopic omental and appendiceal implants were noted and these were studded with mucinous material (Figs 2 to 4). Rest of the abdominal organs were found to be normal with no obvious lymphadenopathy. With a probable surgical staging of FIGO III, total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, and appendicectomy were performed.

Histopathological analysis revealed transmurally infiltrating mucinous adenocarcinoma of the appendix of size 1 × 1 cm, grade 2, with mesoappendiceal invasion and serosal involvement, with lymphovascular invasion and positive surgical margin. Bilateral ovarian masses and omentum showed evidence of metastasis from mucinous adenocarcinoma of the appendix (Figs 5 and 6). Endometrium was atrophic. To confirm the origin of tumor cells, immunohistochemical staining was done which revealed positive for CK 20 (Fig. 7) and CDX2 and negative staining for CK 7. With the histological and immunohistochemical results, the diagnosis of primary mucinous adenocarcinoma of appendix with bilateral ovarian metastasis was confirmed with pT4aG-2pNxpM1c; TNM stage IV. The patient is planned for right hemicolectomy, total peritonectomy and hyperthermic intraperitoneal chemotherapy (HIPEC).

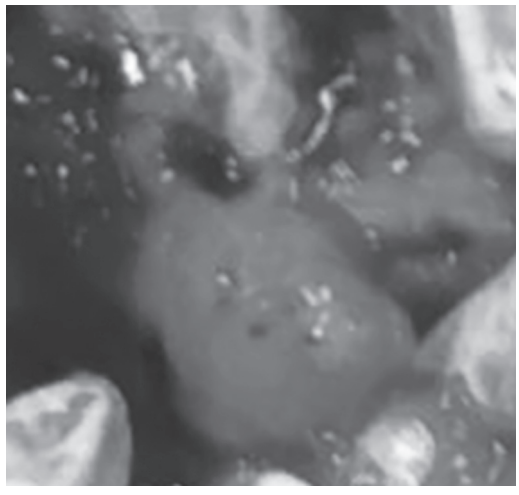

Fig. 2: Appendix studded with mucin

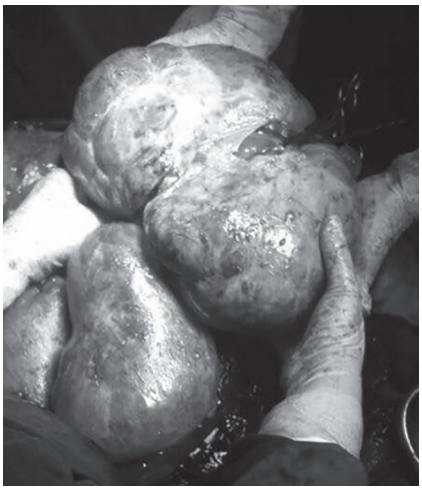

Fig. 1: Bilateral ovarian mass

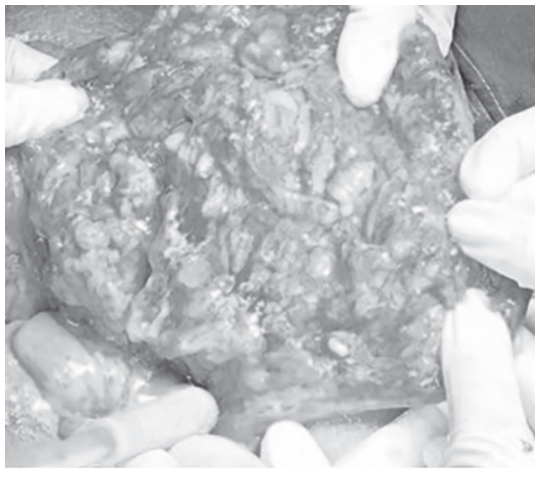

Fig. 4: Omentum studded with mucin

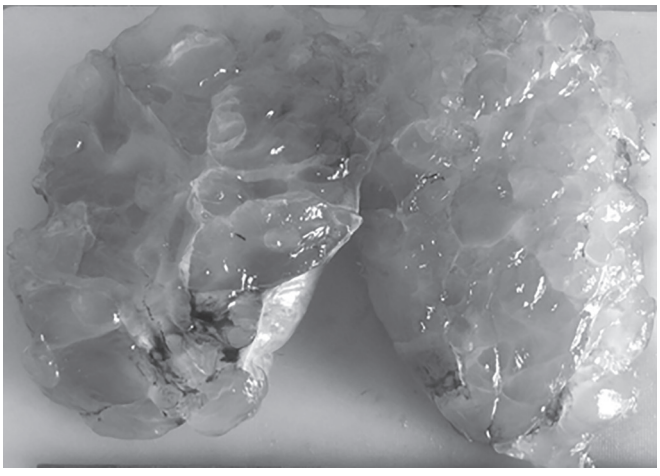

Fig. 3: Cut section of ovary

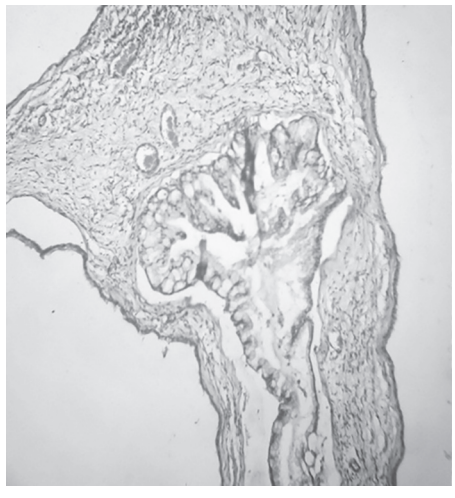

Fig. 5: Histopathological picture of ovary

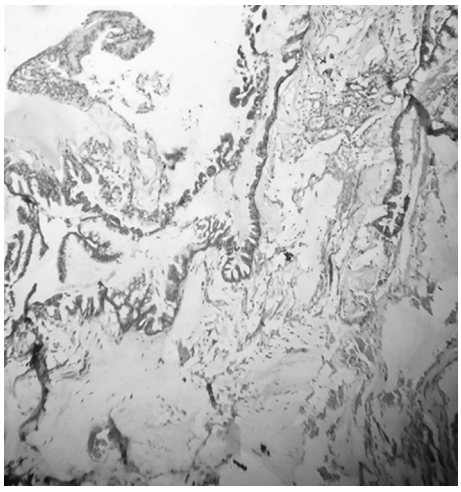

Fig. 6: Histopathology of appendix showing transmural involvement suggesting primary appendiceal carcinoma

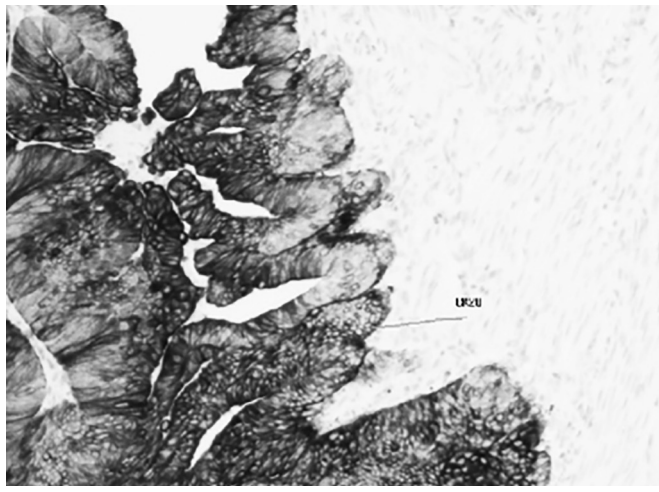

Fig. 7: Immunohistochemistry showing CK 20 positive

DISCUSSION

Primary appendiceal malignancies are very rare and account for approximately 0.4% of GIT tumors[3] and are incidentally diagnosed only in 0.9–1.4 % of appendicectomy specimens.[4] Among these carcinoids comprises of about 66%, cystadenocarcinoma comprises of 15–20% and adenocarcinoma accounts for 10% of tumors.[1] Most common age group is 55 to 70 years (except carcinoid). Men and women are at equal risk.[3] Among these adenocarcinomas can presents in four morphological patterns-mucinous, colorectal, mixed mucinous with a signet ring and signet ring type. Overall 5 years survival is 46% with mucinous adenocarcinomas.[5]

About 7% of lesion presenting clinically as the primary ovarian tumor is of metastatic in origin. Most common sources of metastasis include the stomach, large bowel, appendix, breast, uterus, and lungs.[6]

Ovarian metastasis comprises of 16–37% of adenocarcinomas.[4] Ovarian metastasis usually presents as cystic or solid, well differentiated, mucin-producing and sometimes associated with necrosis and hemorrhage. Features that suggest a metastatic mucinous ovarian carcinoma are (i) bilaterality, (ii) a multinodular surface, (iii) an irregular infiltrative growth, (iv) single cell invasion, (v) lymphovascular invasion and, (vi) an extraovarian spread.[7] In the present case, all features were present and indicate secondary ovarian malignancy.

Ovarian metastases from mucinous adenocarcinomas of GIT mimics primary ovarian mucinous carcinomas. It is very difficult to diagnose it before histopathology. In the case of primary appendiceal carcinoma, there is transmural involvement with a mesoappendiceal spread, whereas in metastasis to appendix will have serosal involvement predominantly.

So for the confirmation of diagnosis and to find the origin of the tumor, immunohistochemistry with cytokeratins and CDX2 is needed.

In any case of adenocarcinoma, CK 7 is positive and CK 20 negative. In cases of mucinous cystadenocarcinoma, CK 7 and CK 20 both are positive. In other cases of ovarian carcinoma, CK 7 positive and CK 20 is negative. In all GIT malignancy, CD X2 is positive. In cases of colorectal carcinoma, CK 20, MUC and CDX2 is positive and CK 7 is negative. In cases of primary appendiceal malignancy, CDX2, CK 20 and MUC positive but CK 7 is negative.[8-11]

In our case CDX2, CK 20 and MUC was positive whereas CK 7 was negative which is suggestive of primary appendiceal carcinoma.

It is very important to have an exact diagnosis, as the management depends on it. An optimal surgical debulking and chemotherapy is the line of management of primary ovarian malignancy[4,12] whereas an aggressive cytoreductive surgery which includes hemicolectomy, total omentectomy, and peritonectomy with hyperthermic intraperitoneal chemotherapy (HIPEC) with mitomycin-C is the definitive treatment of appendiceal adenocarcinomas.[4]

CONCLUSION

Appendiceal adenocarcinomas must always be considered as one of the possibilities in cases of bilateral mucinous ovarian tumors, and in such cases, prophylactic appendicectomies should always be done.

CLINICAL SIGNIFICANCE

Mucinous adenocarcinomas of appendix mimicking primary ovarian malignancies is a diagnostic challenge. The prognosis is poor as it is usually diagnosed at an advanced stage, either due to a low threshold of suspicion and also due to difficulties in diagnosis prior to surgery. Although adenocarcinoma of the appendix is uncommon, they should be considered in the differential

diagnosis of intraabdominal masses, as the treatment modalities vary.

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