International Journal of Surgery Case Reports 97 (2022) 107415

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Bilateral ovarian mucinous carcinoma (stage III) with omental involvement and incidental hydronephrosis: A rare case report

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ARTICLE INFO

Keywords:
Bilateral
Mucinous ovarian carcinoma
Omental involvement
Case report

ABSTRACT

Introduction: Though ovarian malignancies are common, mucinous ovarian carcinomas of high grade are rare. They usually occur in a young female under 40 years of age. Here, we present a case of mucinous ovarian carcinoma (stage III), with omental involvement and incidental hydronephrosis in a 67-year-old female patient.
Case presentation: A 67-year-old female patient presented to us with a history of lower abdominal pain for 2 months and per-vaginal discharge for the last 6 days. On deep palpation of the abdomen, a nodular mass occupying the suprapubic region was found. Bimanual palpation revealed a mass on the right and left adnexa. After visualization of septate cystic mass bilaterally on CECT, she was planned for staging laparotomy with bilateral salpingo-oophorectomy (BSO) with infra-colic omentectomy with peritoneal cytology. Incidentally, a horseshoe-shaped kidney with right mild hydronephrosis was found. After surgery and histopathologic examination, mucinous ovarian carcinoma (stage III), with omental involvement was confirmed.
Discussion: Mucinous ovarian carcinomas are rare malignancies, with different natural history, molecular profile, and prognosis as compared to other epithelial tumors of the ovary. These carcinomas can be either primary or secondary (those metastasized to the ovary from elsewhere), and this differentiation is essential. The therapeutic approach to the patients depends upon the stage at which these carcinomas are diagnosed.
Conclusion: Mucinous ovarian carcinomas are rare and have unique features among the epithelial ovarian carcinomas. Appreciation of these features will surely make a positive impact in improving the management and thus the prognosis of these carcinomas.

1. Introduction

Among the gynecological malignancies, ovarian carcinomas are the second most common cancers, but the most lethal ones [1]. The most common histological type of ovarian carcinomas is the epithelial ovarian carcinomas under which, the mucinous ovarian carcinomas are a rare variety [2]. Mucinous ovarian carcinomas account for about 3 % of all ovarian malignancies [3]. Mucinous ovarian carcinomas usually present as a multi-cystic mass with a huge amount of intracellular mucin in the majority of the tumor cells, and little extracellular mucin [4]. Mucinous ovarian carcinomas usually occur in young females of age less than 40 years. Most of them are diagnosed at an early stage (stage I, 83 %), and fewer are diagnosed at later stages (Stages II and above, 17 %), conferring a favorable prognosis. However, advanced diseases have a poorer prognosis than as compared to other histological types of epithelial ovarian carcinomas [5,6]. Mucinous histology itself has been regarded as an independent factor responsible for the adverse prognosis [5]. Approximately 14 % of the mucinous ovarian carcinomas are diagnosed in stage III [7]. Treatment of mucinous ovarian carcinomas is usually surgical therapy for early stages and surgical therapy followed by adjuvant chemotherapy for later stages [8].

We, herein present a rare case of bilateral mucinous ovarian carcinoma on stage III, with omental involvement and incidental hydronephrosis, managed in our tertiary care center. The case has been reported in line with SCARE 2020 criteria [9].

2. Case presentation

67 years old female P2L2, housemaker, non-alcoholic, and non-smoker presented to our OPD with the chief complaints of lower

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https://doi.org/10.1016/j.ijscr.2022.107415
Received 29 June 2022; Received in revised form 11 July 2022; Accepted 11 July 2022
Available online 16 July 2022

abdominal pain for 2 months and per vaginal bleeding from previous 6 days. Abdominal pain was a dull aching type progressively increased in severity for the last 10 days, radiating towards groins, aggravated on walking, and climbing stairs. Abdominal pain was not associated with nausea/vomiting/burning micturition/fever and used to be relieved on taking analgesics. Per vaginal bleeding was not associated with the passage of clots or abdominal pain and she used to change 2–3 partially soaked homemade pads per day. There was no history of weight loss and loss of appetite. Bowel and Bladder habits were normal. Her menstrual cycles used to be regular before she attained her menopause 20 years ago. She is a known case of hypertension for 2 years and hypothyroidism for 10 years. There is no prior history of any surgical or gynecological malignancy in the family.

On examination, she was afebrile and with normal vitals BP of 120/70 mm of Hg, pulse 78/min, temp 98.6 °F. Chest examination had equal air entry with normal vesicular breath sound, and normal heart sounds with no murmur.

On abdominal examination the abdomen was distended, umbilicus was central and everted. Bilateral flanks were full and all quadrants moved equally with respiration. There was no sign of engorged vessels or venous prominences. On deep palpation huge non-tender nodular mass of approximately 15 × 12 cm with restricted mobility, irregular surface, and ill-defined margin was found on suprapubic, right and left lumbar region. On percussion, dullness was noted all over the lower abdomen. On bimanual examination, the cervix was posteriorly located, and a mass of approximately 10 cm x12 cm in the left adnexa was noted which was fixed, irregular, nodular immobile, and non-tender. A similar mass of 12 × 12 cm in the right adnexa was also felt. The uterus was bulky and a groove was felt between mass and uterus.

Blood counts were within normal limits however she tested positive for HBsAg/HBeAb. Her tumor markers CA-125/CEA/AFP were 50 U/mL, 193 ng/mL, 1.74 ng/mL respectively.

With the suspicion of malignant pathology CECT Abdomen & Pelvis was done which revealed a 12.9 × 11.6 × 10.9 cm sized thin-walled septate abdominopelvic cystic lesion, with no evidence of solid nodules or calcification. A similar cystic lesion of 11.6 × 9.5 × 8.2 cm was also present. Both cystic lesions were drained by gonadal veins. Both lesions were causing anteroinferior displacement of the urinary bladder, bowel loops, right common iliac artery, and bilateral external iliac vessels and were also compressing the right ureter with upstream dilatation of the ureter and pelvicalyceal system. A mild amount of free fluid was noted in the abdomen and pelvis with omental cacking. Both kidneys were anterolaterally malrotated and fused at the lower pole with isthmus measuring approximately 17.5 mm. Thus, an impression of serous cystic neoplasm of bilateral ovaries with horseshoe-shaped kidneys with mild right hydronephrosis was made from CECT.

As routine FNAC/biopsy of suspected ovarian tumors is contraindicated due to the risk of seeding and upstaging of the tumor, she was planned for staging laparotomy with bilateral salpingo-oophorectomy (BSO) with infra-colic omentectomy with peritoneal cytology. An infraumbilical midline incision was given. The peritoneal cavity was filled with gelatinous mucinous fluid as shown in Fig. 1. B/L ovarian cysts of 15 × 15 cm with smooth surface and no tumor deposits were noted.

The right cyst has a rent of 3 cm releasing thick gelatinous mucinous fluid whereas the left cyst ruptured during surgery releasing mucinous fluid as shown in Fig. 2. Bilateral fallopian tubes were stretched and swollen. Bowel loops were adhered with right ovarian cyst. The uterus was difficult to be visualized as it was covered with bowel and adherent with normal-looking bladder anteriorly. The liver surface and undersurface of the diaphragm contained multiple tumor deposits. Infra-colic omentectomy was done and sent for histopathological examination. BSO was done and 100 mL of cisplatin was instilled intraperitoneally via a red rubber catheter.

Histopathological examination showed multiloculated cyst predominantly showing denuded epithelium partly lined by mucinous columnar epithelium. The columnar epithelial cells show moderate to marked nuclear stratification with mild atypia and hyperchromasia with serosal involvement of both ovaries and fallopian tubes. Section from omentum was positive for metastatic mucinous deposits with the presence of a few small clusters of intact mucinous lining confirming the diagnosis of bilateral mucinous ovarian carcinoma (stage III) with omental involvement. The histologic picture is shown in Fig. 3.

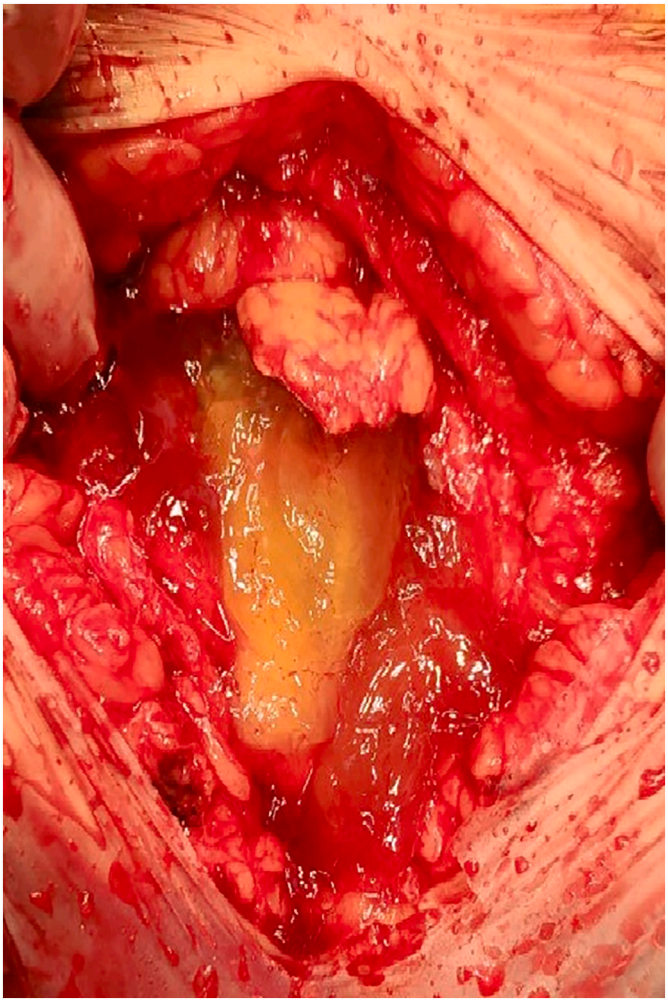

Fig. 1. Peritoneal cavity filled with gelatinous mucinous fluid.

The post-operative period was uneventful and the patient was discharged on the 6th POD with the advice of regular follow-up. The patient is doing well and remains disease-free at one year of follow-up. The patient is happy with the treatment she received.

3. Discussion

Mucinous ovarian carcinomas are a rare variety of epithelial ovarian carcinomas. The mucinous ovarian carcinomas also differ from other types of epithelial ovarian cancers as they have distinct natural history, molecular profile, chemo-sensitivity, and prognosis as compared to others. These tumors differ from the mucinous carcinomas of the GI tract concerning their clinical behavior, pathological features, molecular profile, prognosis, and response to the standard treatment [2]. The commonly known risk factors for other types of epithelial ovarian carcinomas like nulliparity, attaining early menarche or late menopause, absence of breastfeeding, and BRCA mutations, are not associated with mucinous ovarian carcinomas. Cigarette smoking has only been found as a positively correlated risk factor for mucinous ovarian carcinomas [10].

Normally, the ovarian tissues do not contain any of the mucin-

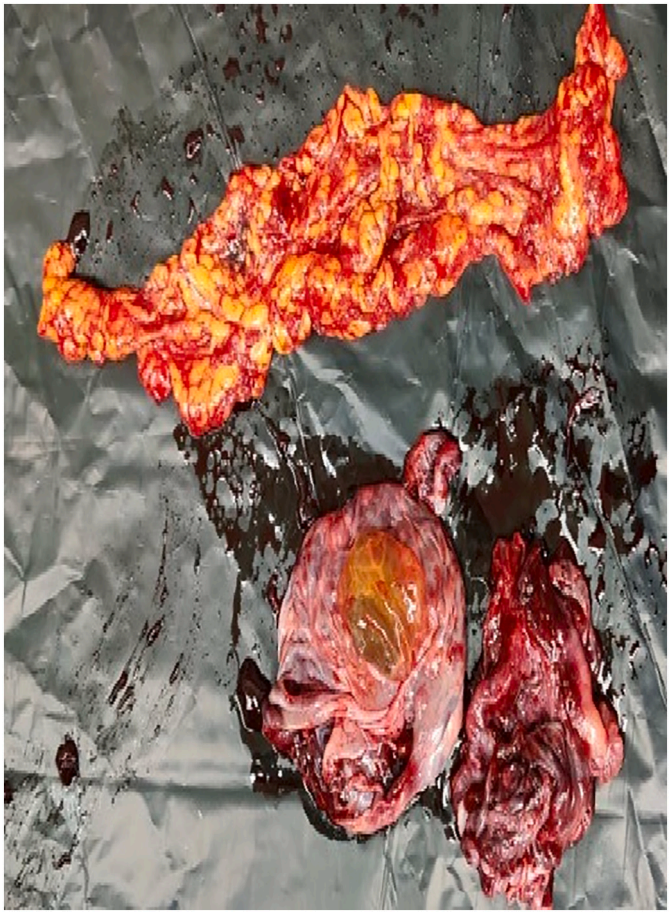

Fig. 2. Picture showing right and left cyst after surgery.

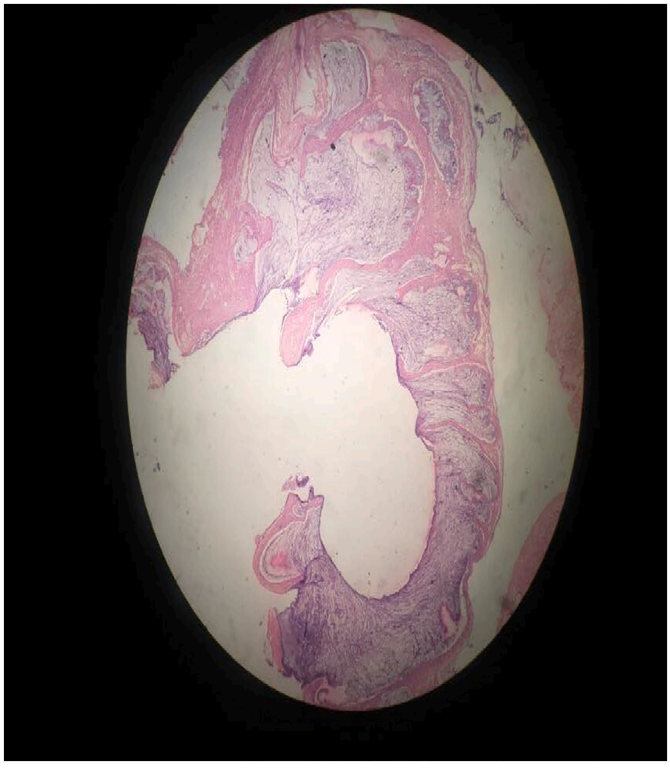

Fig. 3. Picture showing histopathologic image of mucinous ovarian carcinoma.

secreting cells like those present in the stomach, endocervix, or intestine. Multiple theories have been proposed to explain the development of mucinous carcinomas in the ovary [2]. The origin of mucinous carcinomas in the ovary is suggested to have been from the endocervical subtype foci of the Mullerian metaplasia on the surface epithelium or from the cortical inclusion cysts, as explained by Keleman et al. [4].

Primary mucinous ovarian carcinomas need to be differentiated from those metastasized from the GI tract (colon, pancreas, appendix) or those of endocervical or endometrial origin. This diagnosis is also important from the therapeutic point of view. This differentiation can be made based on the clinical profile like bilaterality, involvement of the surface, presence of the signet ring cells, and lymph vascular invasion. These features are usually present in those metastasized to the ovary, and usually rare in primary carcinomas of the ovary [4].

Immunohistochemical profiling of the mucinous ovarian carcinomas has shown them to be CDK7+, CK20 diffuse positive, and co-expression of CDX2. The expression of SABT2, WT1, progesterone, and estrogen receptors, which are generally present in other epithelial ovarian carcinomas, were found to be negative [11]. The frequency of KRAS mutations in mucinous ovarian carcinomas was found to be 76 %, whereas it was only 56 % in a benign form of mucinous ovarian tumors. This suggested that it played role in benign to malignant transformation. ERBB2 amplification has also been reported in about 20 % of the mucinous ovarian carcinomas. These carcinomas have also been found to be associated with CDKN2A loss, IMP3 upregulation, and RNF43 mutations [12].

During workup for the patients during preoperative assessment, besides the detailed history, clinical examination, laboratory investigations, and radiologic imaging, tumor markers are also of utmost importance. Mucinous ovarian carcinomas are frequently associated with elevated CA125, CEA, and CA19-9, the most valuable one being CEA [13]. Besides these investigations, GI investigations are also essential to exclude the metastatic type of mucinous ovarian carcinomas [2].

Though stage I mucinous ovarian carcinomas have an excellent prognosis, there are markedly poor outcomes in advanced diseases when treated with platinum-based chemotherapy. These advanced carcinomas have chemo-resistance, which is the main reason for poorer outcomes [12]. The gold standard operative management of mucinous ovarian carcinomas is staging surgery for early disease and cytoreductive operation for advanced disease. Staging surgery generally includes peritoneal washing, hysterectomy with bilateral salpingo-oophorectomy, lymphadenectomy of surrounding lymph nodes (pelvic and para-aortic), omentectomy, and multiple biopsies from the peritoneum. Removal of all the measurable disease including the microscopic residual disease falls under cytoreductive surgery [14]. Appendectomy is also recommended in all mucinous ovarian carcinomas, however, some authors suggest not removing the appendix if it is normal-appearing [15]. Fertility sparing staging (FSS) surgery is recommended in the early stage of disease as it usually has a good prognosis and occurs commonly in reproductive-age women.

4. Conclusion

Mucinous ovarian carcinomas are a rare variety of epithelial ovarian cancers with unique characteristics. Appreciating the pathologic characteristics and genomic features of mucinous ovarian carcinomas will undoubtedly help in improving the management and thus the prognosis of these carcinomas. Majority of the people who are diagnosed at an early stage do not require adjuvant chemotherapy and can be treated with surgery alone, whereas adjuvant treatment is required in late-stage carcinomas.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Sources of funding

There are no any sources of funding.

Ethical approval

Case reports are exempt from ethical approval in our institution, Tribhuvan University Institute of Medicine, Maharajgunj.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Diptee Poudel (DP), Kshitiz Acharya (KA), and Navin Poudel (NP) = Data acquisition, literature review, and manuscript preparation.

Ashmita Adhikari (AA), Suvana Maskey (SM), and Bishal Khaniya (BK) = Performing the surgical procedure and management of the patient.

All the authors individually did the final proof-reading of the manuscript before submission.

Research registration

None.

Guarantor

Kshitiz Acharya.

Declaration of competing interest

There are no any conflicts of interest.

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