EJGO European Journal of Gynaecological Oncology

# Primary management of an extremely large, invasive mucinous ovarian adenocarcinoma: a case report

**L. N. Abaid[1], N. Zekry[2], A. Ronaghi[3], R. C. Kankus[1], B. H. Goldstein[1]**

*[1]Gynecologic Oncology Associates, Hoag Cancer Center, Newport Beach, CA*
*[2]Department of Pathology, Hoag Memorial Hospital Presbyterian, Newport Beach, CA*
*[3]Memorial Care Imaging Center at Orange Coast, Fountain Valley, CA (USA)*

**Summary**

*Background:* Mucinous tumors are typically benign lesions, although they can be aggressive and exhibit stromal invasion. Moreover, studies have reported on extremely large mucinous ovarian malignancies that have infiltrated the abdomen and pelvis. *Case Report:* The authors describe the history of a 52-year-old woman who presented with a 40-cm mucinous ovarian neoplasm that weighed nearly 22 kg. Initially, confirming the diagnosis was complicated and thus, immunohistochemistry (e.g., pancytokeratin, p53, PAX 8, and CK7) staining was indicated. *Conclusion:* Mucinous ovarian tumors are often associated with a favorable prognosis, but when they are extremely large, disease management can be relatively precarious because of the potential for cardiovascular and intraoperative complications. Since there is some difficulty in assessing large mucinous tumors, immunohistochemistry staining and judicious clinical judgment are essential.

*Key words:* Mucinous ovarian tumors; Large abdominal mass; Patient management; Case report.

## Introduction

Mucinous tumors exhibit intracytoplasmic mucin and originate from surface epithelium and stroma [1]. These lesions are composed of distinct, histologic variants, including benign mucinous cystadenoma, mucinous tumors of low malignant potential, and invasive mucinous carcinoma [2]. Invasive mucinous tumors have a particularly aggressive disposition and feature small glands, nests or individual cells that theoretically coincide with extensive stromal invasion [3].

Clinically, invasive mucinous carcinomas are often evaluated within the context of their presenting characteristics, namely bilaterality, stage at diagnosis, potential for lymphatic dissemination, and elevated serum tumor levels [3, 4]. When distinguishing between primary and metastatic mucinous neoplasms, size and laterality can be determining factors [4]; primary tumors tend to be larger and unilateral, compared with metastatic lesions, which have a proclivity for the gastrointestinal tract, pancreas, cervix, breast, and uterus [5].

Mucinous ovarian malignancies can be decidedly large at presentation and occupy the entire abdominopelvic cavity [1, 2, 6, 7]. Select reported cases have documented lesion sizes ranging from 22-26 cm in diameter, some of which exceeded 15 kg [8]. In the current study, the authors report the history and management of a woman diagnosed with an immense, complex, and cystic mass that was revealed to be an invasive mucinous ovarian cystadenocarcinoma.

## Case Report

A 52-year-old, nulligravid woman presented to the Emergency Room with pelvic pain and distension in July 2017. An ultrasound revealed a large, right-sided complex mass suspicious of an ovarian carcinoma (Figure 1). Her preoperative CA-125 was 75 U/mL, CA-19-9 was 643 U/mL, and CEA was 2.9 ng/mL. The patient's medical history was significant for type II diabetes.

In August 2017, she underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy, infra-colic omentectomy, bilateral pelvic lymph node dissection, right para-aortic lymph node dissection, and complete surgical debulking. Intraoperatively, a cystic and solid mass containing thick mucinous material was encountered; thereafter, the intact mass was extracted through the incision with no spillage into the abdominal cavity. Hemostasis was attained with an estimated blood loss of 250 mL. The patient tolerated the procedure well and no intraoperative complications were observed.

The ovarian mass, which measured 40×35×30 cm (Figure 2) and weighed 21.5 kg, was submitted for pathologic evaluation. Frozen section diagnosis indicated invasive adenocarcinoma and immuno-histochemical staining demonstrated that the malignant cells were positive for pancytokeratin, p53, PAX 8, and CK7; the final diagnosis revealed a Stage IIIA, poorly differentiated, invasive mucinous adenocarcinoma (Figure 3). Ovarian surface involvement and non-invasive implants were absent; the pelvic and para-aortic lymph nodes were negative for malignancy.

Revised manuscript accepted for publication December 13, 2018

Eur. J. Gynaecol. Oncol. - ISSN: 0392-2936
XL, n. 6, 2019
doi: 10.12892/ejgo5018.2019

7847050 Canada Inc.
www.irog.net

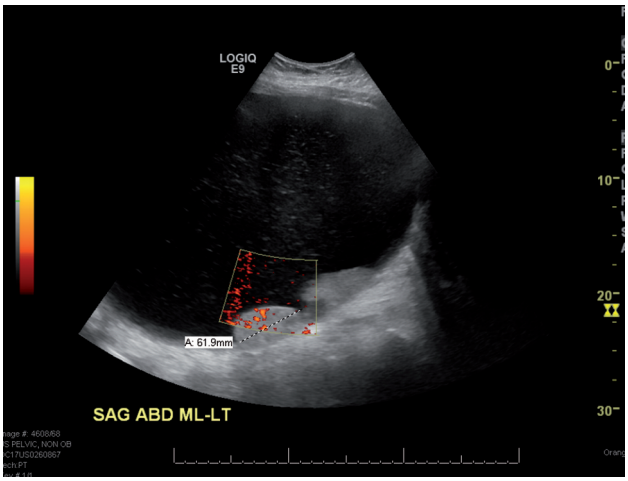


Figure 1. — Ultrasound demonstrates a 35-cm complex cystic lesion with several solid nodular components, the largest measuring 6.2 cm in diameter. Flow within the solid component is seen.

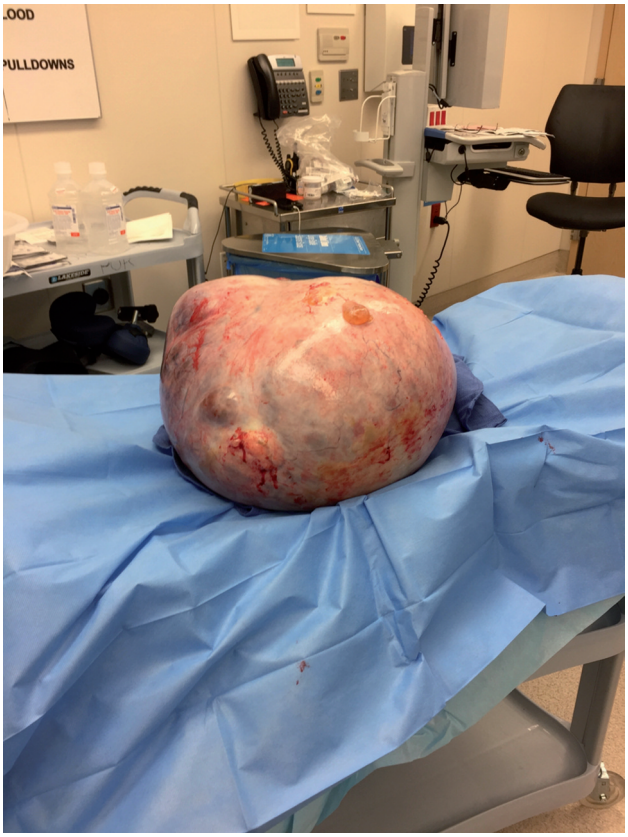

Figure 2. — Gross photograph of the mucinous ovarian cystadenocarcinoma.

Postoperatively the patient was doing well with limited follow-up. In September 2017, she commenced with six cycles of intravenous paclitaxel (80 mg/m$^2$) on days 1, 8, and 15 of a 21-day cycle and carboplatin (AUC 6) on day 1. Her gastrointestinal tract was evaluated with an upper endoscopy and colonoscopy, which excluded a primary gastrointestinal malignancy.

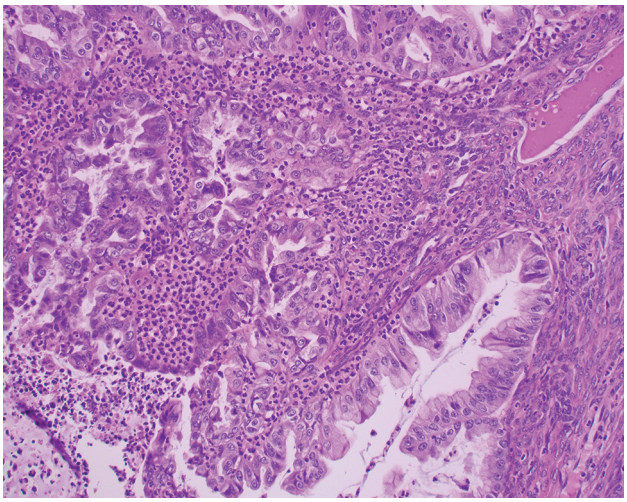

Figure 3. — The epithelium lining the cysts is proliferative and highly atypical, with acute inflammation and necrotic debris present in the lumen.

## Discussion

Invasive mucinous ovarian adenocarcinomas are very rare and comprise less than 2% of surface epithelium ovarian malignancies [9]. The neoplasm's precise incidence is indeterminate because of the varying appearance of metastatic mucinous adenocarcinoma, which is potentially misclassified as the primary disease [4]. Primary mucinous adenocarcinomas of the ovary are usually large, unilateral, smooth, and viscous masses [10, 11]; additionally, they may be diagnosed at an earlier age and characterized by an expansile growth or complex papillary pattern with attendant, luminal necrosis [12].

Metastatic mucinous adenocarcinoma is often bilateral with a multinodular, external surface [13]. The neoplasm ranges from entirely solid to multi-cystic. However, there is some difficulty in assessing large mucinous tumors, especially via frozen section [14]. Hence, when attempting to confirm the diagnosis, representative immuno-histochemical staining (e.g., pancytokeratin, p53, CK7, and PAX 8) and judicious clinical judgment are essential. Moreover, when affirming the presence of metastatic disease, one may scrutinize the presence of ovarian capsular implants, vascular invasion, and infiltrative glandular growth [4, 15].

We describe the unusual history of a woman who presented with a 21.5 kg invasive mucinous adenocarcinoma of the ovary that measured 40 cm in diameter. Previous studies have reported on large mucinous ovarian tumors although rarely have they approached the size of the patient in the current study. Jagtap *et al.* documented the management of a 26 cm mucinous cystadenocarcinoma of the ovary that weighed 9.5 kg [1]. Similarly, Forster *et al.* [7] recounted their experience with a 26-cm, borderline mucinous ovarian tumor that was 3.9 kg. A further review of the literature revealed a patient with a 50.75 kg mucinous ovar-

ian carcinoma, which was metastatic to the coxa and femur [2].

Initially, primary mucinous ovarian carcinomas are treated with surgical debulking and staging. Adjuvant chemotherapy presumably accords some clinical benefit and consistent with the Gynecologic Oncology Group/Gynecologic Cancer Intergroup Study [16], we utilized carboplatin and paclitaxel with our patient; alternatively, bevacizumab, oxaliplatin, and capecitabine have been evaluated in the management of this disease [3].

## Conclusion

Patients with mucinous ovarian carcinoma generally have a favorable prognosis [12]. However, when these neoplasms are considerably large, they are increasingly difficult to manage due to the attendant risk for cardiovascular and intraoperative complications [2, 8]. Ultimately, since primary mucinous ovarian neoplasms may occasion significant diagnostic and clinical difficulties, histopathology is essential to determining the appropriate therapy for this disease [1]. Additional investigation that further evaluates the precise manner on which to diagnose and treat mucinous ovarian carcinomas is warranted.

## Acknowledgement

This study was supported by the Women's Cancer Research Foundation.

## References

[1] Jagtap S.V., Dhawan S.D., Jagtap S.S., Kshirsagar N.S.: "Mucinous cystadenocarcinoma of the ovary presenting as a giant pelvic-abdominal mass". *Int. J. Med. Sci. Public Health*, 2014, *3*, 1305.

[2] Nakamura M., Saitoh M., Miyamoto S., Kubo Y., Tomita H., Andoh A.: "Case of a giant mucinous ovarian carcinoma with bone metastasis". *J. Obstet. Gynaecol. Res.*, 2005, *31*, 576.

[3] Brown J., Frumovitz M.: "Mucinous tumors of the ovary: current thoughts on diagnosis and management". *Curr. Oncol. Rep.*, 2014, *16*, 389.

[4] Seidman J.D., Kurman R.J., Ronnett B.M.: "Primary and metastatic mucinous adenocarcinomas in the ovaries: incidence in routine practice with a new approach to improve intraoperative diagnosis". *Am. J. Surg. Pathol.*, 2003, *27*, 985.

[5] Rouzbahman M., Chetty R.: "Republished: mucinous tumours of appendix and ovary: an overview and evaluation of current practice". *Postgrad. Med. J.*, 2015, *91*, 41.

[6] Katke R.D.: "Giant mucinous cystadenocarcinoma of ovary: A case report and review of literature". *J. Midlife Health*, 2016, *7*, 41.

[7] Forster A., Samaras N., Graf C., Tille J.C., Samaras D.: "Giant mucinous ovarian tumor". *Am. J. Med. Sci.*, 2013, *345*, 400.

[8] Kim Y.T., Kim J.W., Choe B.H.: "A case of huge ovarian cyst of 21-year-old young woman". *J. Obstet. Gynaecol. Res.*, 1999, *25*, 275.

[9] Heintz A.P., Odicino F., Maisonneuve P., Quinn M.A.., Benedet J.L., Creasman W.T., *et al.*: "Carcinoma of the ovary. FIGO 26th Annual Report on the Results of Treatment in Gynecological Cancer". *Int. J. Gynaecol. Obstet.*, 2006, *95*, S161.

[10] Lee K.R., Young R.H.: "The distinction between primary and metastatic mucinous carcinomas of the ovary: gross and histologic findings in 50 cases". *Am. J. Surg. Pathol.*, 2003, *27*, 281.

[11] Guerrieri C., Högberg T., Wingren S., Fristedt S., Simonsen E., Boeryd B.: "Mucinous borderline and malignant tumors of the ovary. A clinicopathologic and DNA ploidy study of 92 cases". *Cancer*, 1994, *74*, 2329.

[12] Simons M., Ezendam N., Bulten J., Nagtegaal I., Massuger L.: "Survival of Patients With Mucinous Ovarian Carcinoma and Ovarian Metastases: A Population-Based Cancer Registry Study". *Int. J. Gynecol. Cancer*, 2015, *25*, 1208.

[13] Boger-Megiddo I., Weiss N.S.: "Histologic subtypes and laterality of primary epithelial ovarian tumors". *Gynecol. Oncol.*, 2005, *97*, 80.

[14] De Decker K., Speth S., Ter Brugge H.G., Bart J., Massuger L.F.A.G., Kleppe M., et al.: "Staging procedures in patients with mucinous borderline tumors of the ovary do not reveal peritoneal or omental disease". *Gynecol. Oncol.*, 2017, *144*, 285.

[15] Jung E.S., Bae J.H., Lee A., Choi Y.J., Park J.S., Lee K.Y.: "Mucinous adenocarcinoma involving the ovary: comparative evaluation of the classification algorithms using tumor size and laterality". *J. Korean Med. Sci.*, 2010, *25*, 220.

[16] Frumovitz M., Schmeler K.M., Malpica A., Sood A.K., Gershenson D.M.: "Unmasking the complexities of mucinous ovarian carcinoma". *Gynecol. Oncol.*, 2010, *117*, 491.

Corresponding Author:
B.H. GOLDSTEIN, PH.D.
Gynecologic Oncology Associates
351 Hospital Road, Suite 507
Newport Beach, CA 92663 (USA)
e-mail: bram@gynoncology.com