

Borderline ovarian mucinous tumor with anaplastic carcinomatous mural nodule: A case report

K. Lakshmi Haridas^{a,*}, Abhilash Menon^a, B. Deepthi^b

^a Department of Medical Oncology, Regional Cancer Centre, Trivandrum 695011, Kerala, India

^b Department of Pathology, Regional Cancer Centre, Trivandrum 695011, Kerala, India

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ABSTRACT

Malignant mural nodules in borderline ovarian tumors are rare. Among them, anaplastic mural nodules are infrequent and only limited case reports are available. Here we report a patient diagnosed as borderline ovarian mucinous tumor with an anaplastic carcinomatous mural nodule. She underwent comprehensive staging laparotomy and six cycles of adjuvant chemotherapy with paclitaxel – carboplatin and has no evidence of disease progression at eight months of follow up. This tumor has an aggressive behaviour and patients with stage $\geq 1C$ have inferior survival.

1. Introduction

Mural nodule (MN) in borderline ovarian neoplasm is a rare entity. The MN can be either benign or malignant with varied sarcoma/carcinoma histologies [Provenza et al, 2008]. The underlying background of ovarian neoplasm is mostly cystic, either mucinous or a serous, with the former accounting for the majority of the cases. Here we report a rare patient diagnosed as ovarian borderline mucinous neoplasm with a MN harbouring anaplastic carcinoma.

2. Case report

A 70 year old lady, para 3 live 3, presented with abdominal distension, bloating sensation and early satiety of one month duration. There was no bleeding per vaginum, breathlessness or other systemic symptoms. She is a known hypertensive on regular medications. She denied any drug allergies or habits. The family history was significant for carcinoma breast in her daughter. At presentation, her Eastern Co-operative Oncology Group performance status was 1. On examination, there was a vague mass palpable per abdomen and the uterus was enlarged. Trans-abdominal ultrasound showed a cystic lesion with fine internal echoes and septations of size 23 cm extending from pelvis to right hypochondrium. Computed Tomography (CT) abdomen and pelvis revealed a multi loculated abdomino-pelvic mass 20x13x17cm, with thick and thin septations, inferiorly extending to right adnexa and superiorly till head of pancreas (Fig. 1). There were para-aortic nodes as well, largest

measuring 2.5x2.1 cm. The serum level of CA125 was 167 IU/L and CEA was 0.5 ng/mL.

She underwent comprehensive staging laparotomy with typeA total abdominal hysterectomy, left adnexal mass excision, right salpingo-oophorectomy, omentectomy and pelvic/para-aortic lymph node debulking. Intra operatively, ascites was mild and liver, spleen, bowel, bladder, mesentery and omentum were normal. The uterus was enlarged to 12 week size with a fundal fibroid. There was a left adnexal mass measuring 25x25cm in size with intact capsule. Right ovary and tube were normal. Para-aortic and bilateral iliac nodes were enlarged and the same were debulked.

On gross histopathological examination, the nodular cystic left adnexal mass was of 29x25x13 cm. Sections studied from left ovarian mass showed predominant portions of a multiloculated cyst lined by mucinous epithelium with stratification of glandular epithelium. Individual cells showed mild to moderate nuclear atypia with basally placed nuclei and mucinous cytoplasm. Atypical cells showed cellular stratification and tufting suggestive of a borderline mucinous neoplasm. A focal nodular area with a MN of 3x2cm was noted in the left adnexal mass. This nodule showed features of high grade poorly differentiated malignant neoplasm composed of round to polygonal cells with high grade nuclei and eosinophilic cytoplasm with brisk mitosis and apoptotic bodies suggestive of a mural nodule. Extensive areas of necrosis and foci of calcification were noted. The right ovary, both fallopian tubes and omentum were free of neoplasm. The borderline mucinous area was immunoreactive for CK7 and immunonegative for CK20, CEA, ER,

* Corresponding author.

E-mail address: lakshmiharidasrcc@gmail.com (K. Lakshmi Haridas).

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vimentin and CDX2; while the mural nodule showed focal immunopositivity for panCK and was immunonegative for CK7, CK20, CDX2, desmin and chromogranin. Correlating the morphology with the immunoprofile, a final diagnosis of borderline mucinous tumour with mural nodule showing anaplastic carcinoma was favoured (Fig. 2). At one focus noted, 9/17 lymphnodes studied from left external iliac node, para-aortic node, right iliac node, right obturator nodes showed metastasis from mural nodule showing features of anaplastic carcinoma with no extranodal extension. Subsequently, the diagnosis of stage III carcinoma ovary was entertained. After multidisciplinary tumor board discussion, she completed six cycles of adjuvant chemotherapy with paclitaxel and carboplatin and has no evidence of progression at eight months of follow up.

3. Discussion

Ovarian MNs are solid masses or nodules which occur in the background of ovarian neoplasms. Majority of the MN are seen in mucinous ovarian tumors with a few occurring in serous counterparts as well. They can either be benign or malignant. The benign variant of MN is termed as reactive sarcoma like lesion and is commonly seen in young patients.

The malignant MN may manifest as a true sarcoma/sarcoma-like and/or as varied carcinoma histologies [Ying Shao et al., 2020]. The malignant tumors have ill-defined borders and they invade vascular structures and capsule unlike their benign counterparts. The reported histologies of malignant MN are fibrosarcoma, rhabdomyosarcoma, carcinosarcoma, undifferentiated sarcoma, clear cell carcinoma and very rarely anaplastic carcinoma [Allende et al., 2010].

Anaplastic carcinomatous MNs are rare with only less than 40 cases reported in the literature. Most of these lesions occur in the background of either borderline mucinous tumor or mucinous cystadenocarcinoma. The largest case series published in 2008 by Provenza et al on anaplastic carcinoma in mucinous ovarian tumors has histologically categorized anaplastic carcinomatous foci into three groups: rhabdoid, sarcomatoid and pleomorphic [Provenza et al., 2008]. The debate on hypothesis regarding the pathogenesis of MN is between collision theory, ie, collision between two independent adjacently placed neoplasms and the progressive dedifferentiation theory of the mucinous cells [Desouki et al., 2015]. The possible presence of totipotent stem cells in the stroma of mucinous tumors and their malignant transformation has also been proposed.

As per Provenza et al, patients with anaplastic carcinomatous MN in mucinous ovarian tumors stage 1a had good prognosis and those with stage $\geq 1C$ had inferior survival outcome [Provenza et al., 2008]. However, no relation was seen between the histological pattern of anaplastic carcinomatous nodules and the stage/prognosis of the disease [Provenza et al., 2008]. These are aggressive tumors with 50% mortality rate [Desouki et al., 2015]. Our patient presented with pathologically

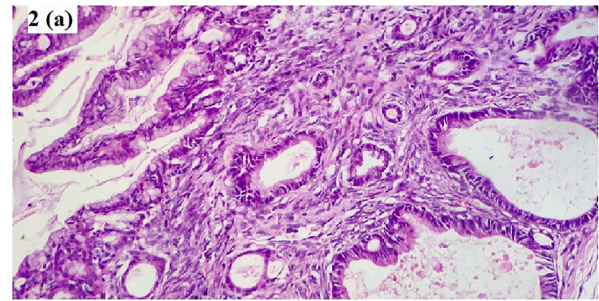


Fig. 2a. Mucinous glands showing focal stratification and mild atypia (400x,H&E).

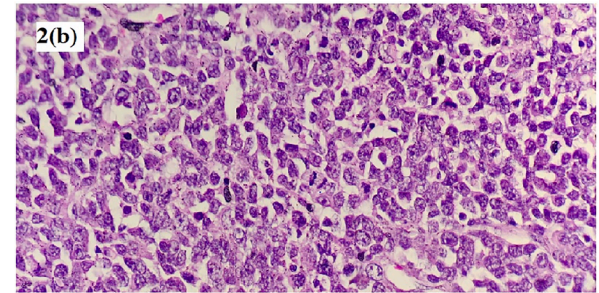


Fig. 2b. Showing a mural nodule with anaplastic areas with high mitosis (400x,H&E).

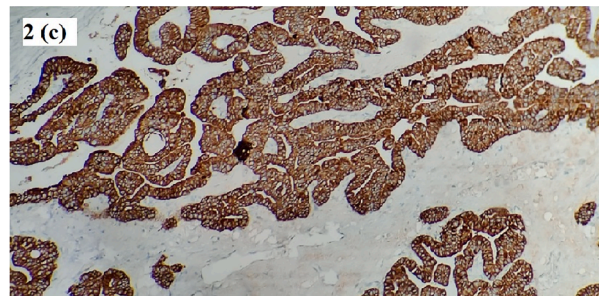


Fig. 2c. CK7 immunohistochemistry showing diffuse strong membranous positivity in borderline mucinous tumor areas.

positive lymph nodes after comprehensive staging laparotomy. She received six cycles of adjuvant chemotherapy and has not progressed at eight months of follow up.

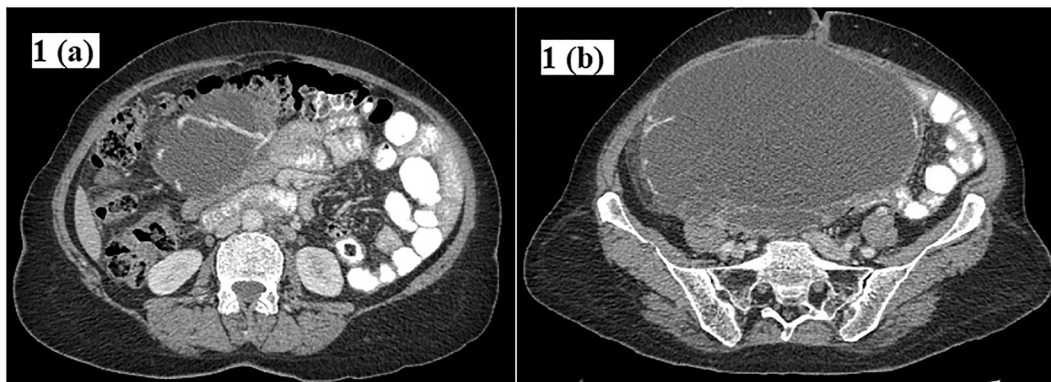


Fig. 1. (a) and (b): Computed Tomography abdomen and pelvis showing a multi loculated abdomino-pelvic mass 20x13x17cm, inferiorly extending to right adnexa and superiorly till head of pancreas.

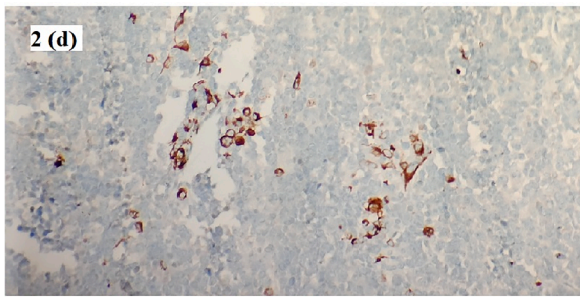


Fig. 2d. CK immunohistochemistry showing focal scattered strong membranous positivity in mural nodule.

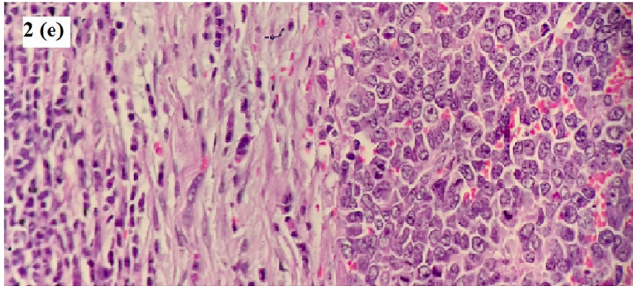


Fig. 2e. Microscopy showing a lymph node with metastases from anaplastic carcinoma (400x,H&E).

Patient consent – obtained.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Author contribution:

Author 1: Was involved in concepts, design, definition of intellectual content, literature search and manuscript editing.

Author 2: Was involved in data acquisition and manuscript preparation.

Author 3: Was involved in histopathological review and manuscript preparation.

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