Case Report:

De Novo Ovarian squamous cell carcinoma : A case of rare malignancy.

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Abstract

Pure primary squamous cell carcinoma of the ovary, not associated with pre-existing ovarian lesions like dermoid cyst, Brenner tumours, or endometriosis is extremely rare. A 28 year old nulliparous female presented with acute pain in left lower abdomen and weight loss. Ultrasonography showed a mass arising from left ovary. An ovarian mass adhered to lateral pelvic wall was found on laprotomy. Histopathology confirmed the diagnosis of squamous cell carcinoma of left ovary with no co-existing ovarian dermoid. Postoperatively patient died within 8 weeks despite adjuvant chemotherapy.

Keywords: De Novo, Primary Squamous cell carcinoma

Introduction

Squamous cell carcinoma (SCC) of the ovary is a rare clinical entity, accounting for less than 1% of the malignant tumours of the ovary.[1,2] Pure primary SCC of the ovary not associated with pre-existing ovarian lesions like dermoid cysts, Brenner tumours or endometriosis is extremely rare.[3] Herein we report the18th case of exceedingly rare malignancy of ovarian tumour (pure,denovo) SCC of ovary.[4]

Case Report

A 28 year old nulliparous female was admitted with history of pain in abdomen for one week and weight loss since six months. Patient was poorly built and nourished. Per abdomen examination revealed mass in the left lower abdomen which was tender on palpation. Per speculum vaginal and rectal examination were unremarkable. A pap smear examination was negative for dysplasia/malignancy.

Abdominal ultrasonography (USG) revealed left ovarian mass. Fractional curettage revealed normal cervical mucosa and proliferative phase endometrium. Complete blood count and chest X-Ray were within normal range. On exploration, solid mass was seen arising from left ovary adherent to pelvic wall with no evidence of ascitis. Right adnexae and uterus were normal. Abdominal lymph nodes were unremarkable. Left ovarian mass along with left fallopian tube were removed.

Gross examination of the specimen received in the surgical pathology section showed greyish solid and cystic mass, approximately 12x8x5 cm, with denudation of capsule at one place. On cut section the tumour was partly solid and partly cystic filled with turbid fluid (Fig1).

Careful examination failed to reveal any teratoid elements. Histological examination of ovarian mass showed undifferentiated cells in sheets and strands with areas showing obvious squamous differentiation with keratin formation (Fig 2,3). No evidence of teratoid elements, endometriosis or Brenner tumour were detected in the sections. Normal ovarian stroma was noted at the periphery. Left Fallopian tube was unremarkable. Diagnosis

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of de novo primary SCC of ovary was rendered. The immunohistochemical(IHC) profile showed CK7, EMA, and high molecular weight keratin positive in tumour cells. Inhibin and CK20 were negative. Since mass was adherent to pelvic wall, FIGO stage IIb was assigned. Patient recovered from primary surgery and was advised for chemotherapy course of carboplatin. Unfortunately patient succumbed to death within 8 weeks.

DISCUSSION

Ovarian cancers remain second most common female genital tract malignancy.[1] Mature teratomas (dermoids), included in germ cell category, are most common tumours that can occur in any age. Squamous elements are most commonly arising in ovaries as a part of mature teratoma. Although squamous component in teratoma are benign, SCC can arise from mature teratoma and is the common malignancy arising from mature teratoma.[5] Squamous component arising in absence of teratomatous component are distinctly rare.[6]

Generally, they are known to occur as metastasis from extra ovarian squamous lesions, especially cervix, as part of metaplastic process in the endometrioid adenocarcinoma or Brenner tumour.[5,6] However, in the present case pap smear and fractional curettage examination revealed no dysplasia or malignancy and no evidence of glandular or transitional differentiation seen in the ovarian tumour on histopathology. Endometrioid adenocarcinoma of ovary typically presents postmenopausally and is characterised by areas of squamous differentiation arising within neoplastic endometrioid glands. Brenner tumours present between the age group of 40-50yrs, show transitional differentiation and may also show areas of squamous differentiation. Still rarer are SCC, showing none of the above features.[5] Malignant transformation have also been reported in endometriotic ovaries.[6,1] Reports of primary (pure, de novo) SCC arising in otherwise healthy ovaries are extremely rare.[6,1] Till date about 17 cases of de-novo SCC of ovary have been published.[4] The present case is 18th in the medical literature[4] and second case in Indian population.

In a series of 37 cases of primary SCC of the ovary, 11 cases were of pure SCC with mean age of 27-73 years and three patients had cervical squamous cell carcinoma in situ, four patients were in stage II. Overall mean survival of these cases were between 4-5 months in the series. The stage of the tumour and its grade correlated well with overall survival for SCC.[6] However, in the present case there was no evidence of cervical dysplasia in pap smear and fractional curettage and patient was in stage II and died within 8 weeks. The role of Ca-125 in de novo SCC has also not been well established.[7,4]

Histopathology remains gold standard in diagnosis of SCC. In it's well differentiated form, SCC shows squamous differentiation, keratin formation and intercellular bridging. In its moderately and poorly differentiated form, few squamous features may be identified, such cases may require ancillary immunohistochemical or electron microscopic study to confirm the diagnosis. In the present case, however, areas showing keratinization and keratin pearl formation were evident and IHC was confirmatory. Similar to other organ system, SCC of the ovary should show high molecular weight (HMW) cytokeratin positivity as well as p63 positivity.[8] In present case HMW cytokeratin(CK) positivity was seen. CK 7 +ve and CK 20 –ve is seen in non-mucinous ovarian carcinoma.[9]

Since the data on options for postoperative adjuvant therapy either radiotherapy, chemotherapy or combination of both are insufficient and conflicting, most of the authors opine the use of

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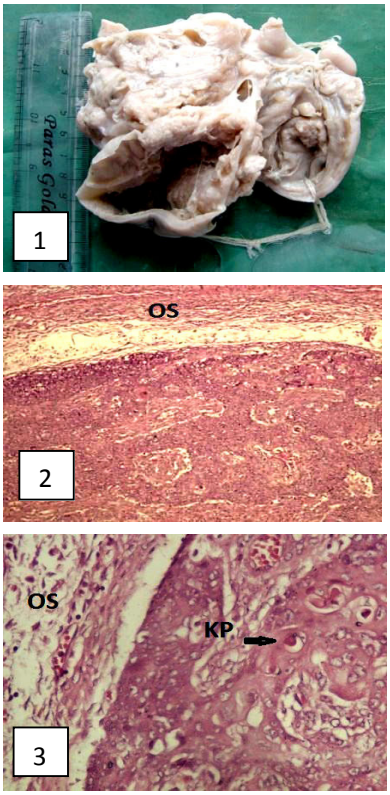
chemotherapy.[4] However, it remains unclear whether patients with de novo primary SCC of ovary would benefit from similar adjuvant therapy.[1] In particular, most of the cases (80% in one study) of SCC of ovary result in death within a few months of diagnosis and radiotherapy is also of limited value.[5,6] In another case of 65 year woman with pure primary SCC, no response to chemotherapy was observed and patient expired 6 months after diagnosis.[10] Etiology of SCC of ovary without any lesion remain obscure; some authors suggest that these lesions may arise due to seeding from occult pre or fully malignant squamous lesions in other location.[5,6]

To conclude the present case of de novo primary SCC of ovary is an exceedingly rare example of ovarian tumour. Histopathological examination remains gold standard for the diagnosis of SCC of ovary. Overall prognosis of the cases with primary SCC of ovary remains poor.

[Fig 1- Gross picture of Ovarian mass showing solid and cystic areas.

Fig 2- Section showing Ovarian stroma(OS) and sheets of malignant squamous cells(H&E, x100)

Fig 3- Section showing ovarian stroma (OS) on left side and malignant squamous cells in sheets with keratin pearl (KP) on right side (H&E, x400)]



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