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Case Report

Obstetrics and Gynaecology Section

Pure Primary Squamous Cell Carcinoma of Ovary – A Rare Case Report

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ABSTRACT

Primary Squamous Cell Carcinoma (SCC) is a rare tumour which arises in a mature cystic teratoma, endometrioma or Brenner tumour. The pure variety arises from metaplasia of surface epithelium of ovary and it is the rarest type. For optimal management no definitive treatment protocol is available till date. Also, there is no agreement concerning the postoperative therapy-chemotherapy or radiotherapy. We present a rare case of pure primary SCC of ovary which was managed by aggressive cytoreductive surgery followed by chemotherapy.

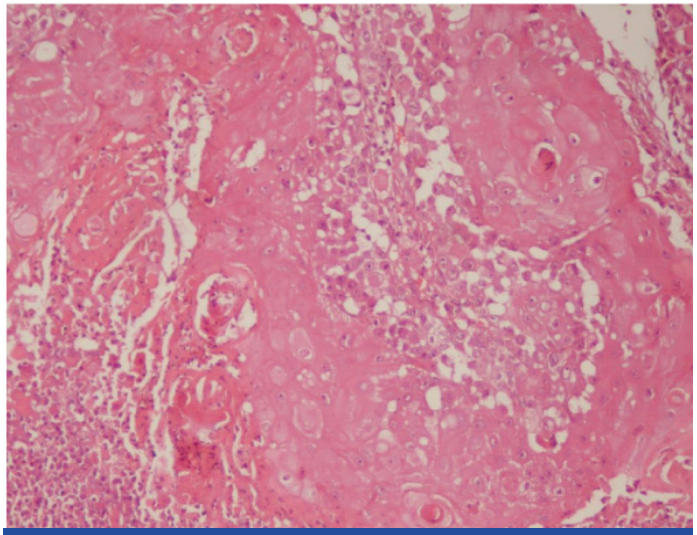
Keywords: Pure primary ovarian squamous cell carcinoma, Squamous cell carcinoma antigen, Squamous cell carcinoma of ovary

CASE REPORT

A 30-year-old multiparous lady presented with mild, persistent pain in right lower abdomen since three months and low grade fever for one month, but no history of loss of weight or appetite. Her bladder and bowel habits were unaltered and menstrual cycles were normal. The gynaecological examination revealed a 6 x 5 cm bilobed, firm-hard, tender mass with restricted mobility in right and posterior fornix. Left fornix was free. Uterus was normal in size but deviated to left side. On per-rectal examination rectal mucosa was free. Haematological, biochemical tests and chest X-ray were within normal limits. CA-125 level was mildly raised to 48.44 IU/ml. Ultrasonography findings were suggestive of right sided inflammatory adnexal mass and mildly hydronephrotic right kidney.

Patient did not respond even after one week of triple antibiotics therapy. There was persistence of pain and the adnexal mass although she became afebrile. Laparotomy was planned as patient did not respond to antibiotic therapy and pain and the mass persisted. Exploratory laparotomy revealed a right sided tubo-ovarian firm-hard 7 x 5 cm mass adherent to posterior surface of uterus, broad ligament and sigmoid colon. Left tube was oedematous and left ovary grossly normal. Abdominal cavity was normal except for few calcified nodules on omentum. There was absence of free fluid or palpable para-aortic lymph nodes. Considering the young age, mildly raised CA-125 levels and probable inflammatory (or tubercular) nature of the mass, right salpingo-oophorectomy with left tubal ligation was done. Removal of the tubo ovarian mass after adhesiolysis left a small raw area on the right pelvic wall. Grossly, the lesion was firm to hard and densely adhered to uterine serosa and pelvic peritoneum. Cut – section of the mass was predominantly solid with few areas of necrosis. The histopathological report revealed SCC of the ovary without evidence of any other associated lesions, including dermoid cysts, endometriosis or Brenner tumour [Table/Fig-1]. Left tubal segment was normal.

Chest x-ray, pap smear, indirect laryngoscopy, USG (whole abdomen, breast and thyroid) was done to exclude any occult primary site of tumour. Ultra sonography revealed moderate hydronephrosis and hydroureter in right kidney. Transvaginal ultrasound (TVS) revealed a homogenous 5 x 3 cm mass on right side with increased vascularity and appearance of a focal metastatic mass (2 x 1 cm) in left ovary. Staging laparotomy with cytoreduction was undertaken after two weeks of primary surgery. A residual growth (5 x 5 cm) adherent to right and posterior surface of uterus and infiltrating into right lateral pelvic wall, pouch of Douglas, recto sigmoid region and terminal ileum was seen per operatively. Uterus was of normal size. Discrete hard nodules were present on omentum and right ureter was dilated. Total abdominal hysterectomy with left salpingoophorectomy with resection of residual tumour (debulking) with infra-colic omentectomy with ileal resection and anastomosis was done. However, a residual tumour (2 x 3 cm) adherent to sigmoid colon and right ureter could not be resected. Histopathological report revealed SCC deposits in left ovary and omentum. The uterus, cervix and tube were unremarkable. Thus, the final diagnosis was primary SCC of ovary (pure variety) – stage III c. Pre chemotherapy CECT scan revealed the residual malignant mass (4.3 x 5.6 cm) invading rectum, sigmoid colon and right ureter resulting in proximal hydronephrosis and hydroureter. Chemotherapy with cis-platinum (75 mg/m^2) and paclitaxel (135 mg/ m^2) every 21 days was started. Seven cycles of chemotherapy resulted in only 50% reduction of clinically palpable right pelvic mass as assessed by CT scan. The patient and her family were prognosticated about the poor response and survival. She succumbed to the disease within one year of her surgery, after three months of the last chemotherapy.



[Table/Fig-1]: Microphotograph of right ovary showing malignant squamous cells with Keratin Pearls and areas of necrosis. (H&E, 40X).

DISCUSSION

SCC is a rare type of ovarian malignancy which generally arises from malignant transformation in a mature cystic teratoma and less often in endometriosis or Brenner's tumour. The pure primary form which arises denovo is the rarest. It results from metaplasia of surface epithelium of ovary [1].

Among all types of ovarian malignancy primary SCC of ovary is a rare entity, with an incidence of less than 2% [1]. Majority of the cases develop from malignant transformation in mature cystic teratoma, endometriosis or Brenner tumour. Very rarely it may arise de novo from ovary where the surface epithelium undergoes squamous metaplasia. Possibly first reported case of pure primary SCC was in 1988 by Ben-Baruch C et al., [2]. Since then only few cases have been reported. Thus no quoted incidence of pure variety is available in literature. Metastatic ovarian carcinoma comprise of 5-6% of all ovarian cancer. Moreover, SCC of the ovary may be metastatic in 2% of all metastatic ovarian cancer [3]. Majority of metastatic SCC originates by a distinct extension from the cervix [4].

Primary SCC of ovary arise from dermoid cysts, Brenner's tumour, mucinous cystadenoma, or endometriosis. Pure primary SCC of ovary not associated with pre-existing ovarian lesion is extremely rare. Till date, only 30 cases of pure primary carcinoma of ovary has been reported in English literature [5].

Various mechanisms have been proposed for origin of these tumours [1]. As tumours arising from the ovarian surface epithelium may contain squamous elements, some tumours may evolve directly from this epithelium. Furthermore, of the reported cases of pure primary SCC, the most significant association identified was with cervical dysplasia. This association could be explained by the following three theories: (i) Contiguous spread along the mucosal surface of female genital tract onto the ovary; (ii) Microscopic angioinvasive cervical carcinoma undetected in the areas sampled, with metastasis to the ovary; (iii) field effect [1].

In one of the largest collection of cases by Pins MR et al., who reported 11 cases of pure primary SCC among a total of 37 cases of primary SCC of ovary – the mean age of patients reported for pure SCC is 56 years (26-73 years). Also, the average size of the tumour is 6-26 cm in greatest diameter and these tumours are usually solid with focal necrosis [1]. Out of these 11 patients, three patients also had cervical SCC in situ. Usually patients with pure variety present with symptoms of abdominal or pelvic pain and/or vaginal bleeding. Sometimes the tumour may be asymptomatic and is found incidentally on physical examination. The mode of spread is transmural with extensive local invasion and so the spread differs from other common ovarian tumours. Recently, a case of bilateral pure SCC of the ovary has been reported [6].

Among tumour markers SCC antigen is significantly raised in SCC arising from teratoma, thus establishing the role of SCC antigen in pre operative diagnosis of malignant transformations and early detection of cancer recurrence [7]. The role of SCC antigen in pure ovarian SCC has not been studied separately due to rarity of this type of malignancy. SCC antigen was not estimated in our case.

No definite treatment guidelines are yet available for effective management of pure SCC of ovary because of rarity of this tumour. Multi modality therapy including aggressive cytoreductive surgery followed by cisplatin based chemotherapy and/or radiotherapy has been proposed to improve survival; however the mean survival in the pure variety of primary SCC is only 11 months [8]. Patients with early disease may respond to optimal debulking. However, advanced stage disease has a very poor prognosis despite postoperative chemotherapy and/or radiotherapy [8,9]. Due to the rare nature of pure primary ovarian SCC, effective adjuvant chemotherapy or radiotherapy regimens have not yet been established. Park J-W et al., described a case which presented with the complaint of cough and was diagnosed as Stage IV primary ovarian SCC with lung metastases, however, the patient died within one year of debulking surgery despite of undergoing postoperative chemotherapy [5]. Another case of a postmenopausal female has been reported who was diagnosed with pure primary SCC of left ovary [9]. Although, postoperative radiation and adjuvant chemotherapy was administered, the patient died in two months.

CONCLUSION

The aim of reporting this case is not only to describe the pathology, but to create awareness among gynaecologists owing to the rarity and poor prognosis of SCC of the ovary. To conclude, primary SCC arising in the ovary is an extremely rare entity. Due to the rarity of the condition and limited data, principles underlying the management and chemotherapy regimens have not been standardized. Therefore, to formulate protocols regarding the management, such cases need to be reported and researched.

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