De novo Ovarian Squamous Cell Carcinoma. A Case Report and Review of Literature

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Introduction

Malignant ovarian tumours comprise of a spectrum of clinicopathologic entities. Many have comparable presenting clinical features, some are associated with unique and intriguing histomorphological features. Not only does a histopathology define a tumour's ontogeny and biology, they may also predict prognosis and help plan an adjuvant therapy. What follows is a 17th case report of an exceedingly rare primary ovarian tumour (pure, de novo), squamous cell carcinoma (SCC). However, it is known that 2 % of mature teratomas harbour malignant component and up to 80 % will be squamous. Some pure SCCs will arise in association with a foci of endometriosis and very rarely entirely de novo [1–4]. SCC of ovary are aggressive tumours, which may be amenable to multimodality treatment. Due to conflicting evidence from literature, the best therapeutic option cannot be deciphered. Treatment needs to be tailored to patient profile.

Case Report

A 50 year old postmenopausal woman presented with history of pain in the left iliac fossa for 2 months. On examination tenderness could be elicited in the left iliac fossa with

a vague mass, there was no ascites. Per speculum examination ruled out cervical and vaginal cause for pain abdomen. A pap smear was performed. Per vaginal and rectal examination depicted an atrophic uterus and a hard mass measuring 8×8×8 cms behind the uterus and probably adherent to the rectosigmoid area, tender to palpate, thickened left parametrium, normal right parametrium and rectal mucosa. Her haemogram, blood and serum biochemistry were normal, serum antibodies to HIV and HbSAg were negative, normal chest x-ray. Ultrasound of abdomen and pelvis revealed the presence of a solid and cystic heterogenous mass in the left adnexa and pouch of douglas, with compression on the left ureter leading to mild hydronephrosis, Fig. 1. Ca-125 levels was 70U/ml. Pap smear was normal.

During exploratory laparotomy, a left ovarian tumor which was solid and cystic, necrotic and purulent material emanating from it measuring 7×8×7 cms, densely adherent to the pouch of douglas (pod), left parametrium and left ureter and the rectosigmoid junction was encountered. Right ovary had few deposits on surface which was removed, uterus was atrophic. Upper abdomen, omentum appeared normal. No palpable para-aortic or pelvic nodes. On table frozen section revealed the presence of squamous cell carcinoma probably primary or metastatic. A total abdominal hysterectomy, omentectomy, bilateral pelvic lymphnode dissection and excision of tumor deposits on the distal end of the ureter was performed. Post operative period was uneventful.

Final histopathology was moderately differentiated squamous cell carcinoma of the left ovary with involvement of the fallopian tube, ureteric deposits showed tumour, Figs. 2 and 3. The right ovary and tube, uterus, cervix, bilateral parametria and omentum were unremarkable.

She was assigned a stage II C.

She underwent external radiotherapy with a dose of 50 Gy over 28 fractions along with single agent weekly cisplatin chemotherapy. She presented 6 months later with

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Fig. 1 Ultra sound showing atrophic uterus (U), cervix (C), bladder (B), and the heterogenous mass posterior to the uterus (M)

distension of abdomen, obstipation with intestinal obstruction and chest x-ray evidence of metastasis, Fig. 4. She deteriorated and died of respiratory failure.

Discussion

Pure primary SCC have been classified by the World Health Organization criteria as surface epithelial-stromal tumors [1]. To date about 16 cases of de novo SCC of ovary has been published [3–10].

Certain practical findings are noteworthy in the present case whose presentation was similar to any ovarian tumour presenting with postmenopausal status, 50 year old, pain abdomen, with a preoperative diagnosis of a solid cystic ovarian mass, probable adherent in the pouch of douglas and adherent to ureter, mildly elevated Ca-125- 70U/ml, who will need an on table frozen section to categorize the tumor and decide the procedure.

The role of elevated Ca-125 in de novo SCC has not been very well established and some authors are of similar opinion [11].

Fig. 2 H and E X10, showing ovarian stroma (OS) at the upper end and de novo squamous cell carcinoma with keratin pearls (KP) at the bottom of the figure

Fig. 3 High power showing keratin pearls

What is important is the frozen section diagnosis of epithelial or other category of tumours, because optimal cytoreduction is the standard of care for epithelial ovarian tumours. It is not clear from the literature if optimal cytoreduction has any benefit on survival, however authors have opined that when the dilemma exists, perform an optimal cytoreductive surgery [7, 8]. Histopathology remains the gold standard to trace back the tumor to its ontogeny and biology, similar experiences have been shared by many authors [3, 10].

Since the data on options for post operative adjuvant therapy either radiotherapy, chemotherapy or a combination of both are heterogenous and insufficient and also conflicting, most of the authors opine the use of chemotherapy. Various drugs like cisplatin, carboplatin, have been used, but paclitaxel based regimens have produced good results [12, 13]. On the contrary not so good response also have been reported [6–10].

Use of radiotherapy stems from the understanding that squamous cell carcinoma is radiosensitive tumour and

Fig. 4 Chest x-ray showing cannon ball (CB) appearance following tapping for pleural effusion depicting pulmonary metastasis

concurrent weekly chemotherapy with cisplatin and whole pelvis radiation has shown benefit, which has been extrapolated from results for SCC within antecedent dermoids [2]. Whether the same concept apply to de novo SCC is not clear.

However, it has been observed that when it is an advanced stage, they behave very aggressively with recurrences almost within 5 weeks–6 months [6–10] inspite of initial response to first line chemotherapy and optimal cytoreductive surgery similar to the present case.

The survival ranged from 5 weeks to 24 months in de novo SCC. In the present case she was disease free for only 5 months and died of respiratory failure due to pulmonary metastasis compounded by intestinal obstruction and sepsis. Similar experiences have been discussed by other authors [6–10, 12].

With the knowledge that de novo SCC being a rare tumour and available narrow treatment options, it should not deviate a surgeon from performing an optimal cytoreductive surgery. Postoperative adjuvant therapy though not very well established, we can still extrapolate experience from SCC in ovarian dermoid and attempt to materialise its benefits in de novo SCC and hence use weekly concurrent chemotherapy along with radiotherapy as discussed. With the availability of better chemotherapeutic drugs there appears to be a better outcome for these women.

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