De-Novo Squamous Cell Carcinoma of Ovary - A Very Rare Tumor Masquerading as Uterine Fibroids

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Short Communication

Pure primary squamous cell carcinoma arising in the ovary is a very rare tumour. De-novo occurrence of an ovarian squamous cell carcinoma (SCC) in absence of a primary ovarian dermoid cyst or Brenner’s tumor occurs even less commonly than the incidence of SCC ovary which is about 2%-3% [1,2]. Some pure SCCs arise in association with a foci of endometriosis and very rarely entirely de novo [1,2]. Mean age of occurrence of pure SCC is 56 years (27-73 years) [2]. Pure primary SCC have been classified by the World Health Organization criteria as surface epithelial-stromal tumours [1] and are believed to arise from metaplasia of the surface epithelium of the ovary. Till date less than 20 cases of de-novo SCC of ovary have been published [2,7].

A 56 year post-menopausal lady, P0L0 who was a known case of uterine fibroids presented to the emergency with acute pain abdomen. She had a prior history of lower abdominal pain and fever off and on for the past 4 months. Clinical examination was suggestive of an 18 weeks palpable mass in lower abdomen with an irregular surface and restricted mobility. Her baseline haemogram, liver and kidney functions were normal. Ultrasound was suggestive of an enlarged uterus with necrotic areas likely fibroids, with multiple coarse interrupted calcifications along anterior wall of uterus. A tubular structure either dilated fallopian tube or dilated appendix on right side of abdomen fused with lower pole of kidney (cross fused ectopic kidney). Bilateral ovaries were not visualized. At this point tumor markers were sent which showed mildly raised CA 125 and raised CEA with the values as follows; CA 125:126.3, HE4:213, ROMA value: 73.70, CEA:>100. With a provisional diagnosis of degenerated fibroids, patient was posted for laparotomy followed by pan-hysterectomy.

At laparotomy, a large mass about 18 weeks gestation uterine size was seen with partly mucoid degenerative material and partly solid component. Uterus, bilateral tubes and ovaries were difficult to delineate separately from the mass. Mass was adherent above to the omentum and bowel loops, laterally to pelvic side wall and anteriorly to bladder. Peritoneal fluid was collected for cytology. Both ureters were traced. Both kidneys were fused together on right side. Sigmoid colon was adherent to the tumor. Adhesiolysis was done and mass was separated all around from its attachments. In view of the above findings, a frozen section biopsy from the mass was sent. This revealed features of a squamous cell carcinoma probably arising in the ovary. Pan-hysterectomy with omentectomy was done following this report. On cut-section, the tumor was firm at places and necrotic at others (Fig 1). Patient made full recovery in the post-operative period and was discharged on 4th post-operative day.

The final histopathology revealed a well to moderately differentiated squamous cell carcinoma – possibly arising from a mucinous cystic tumor of endocervical type in the right ovary. There was no evidence of endometriosis, Brenner’s tumor or any other ecto/meso/endodermal elements and no normal ovarian tissue (Fig 2,3). The omentum was free of tumor. Peritoneal fluid cytology showed nucleated and anucleated squamous epithelial cells in a background of inflammatory cells, RBCs and debris; with a few squamous cells having atypia. Based on the staging, she was diagnosed as well to moderately differentiated squamous cell carcinoma of right ovary arising de novo, stage IIC.

SCC of the ovary are very aggressive tumors, presenting as symptomatic abdominal mass, requiring histopathology for final diagnosis and a multimodality treatment approach. Treatment needs to be structured as per the patient’s clinical status [2,5]. Frozen section has a role in the diagnosis of epithelial or other category of tumours since optimal cytoreduction is the primary management for epithelial tumours [5].

The role of elevated Ca-125 in de novo SCC has not been very well established and CA 125 can be normal to mildly elevated [8] as in this patient. Pure SCC have been found to be positive for IHC
markers -MA903, p63 and ck 5/6.

The data on post operative adjuvant therapy, either radiotherapy, chemotherapy or both are insufficient. Chemotherapy with platinum based agents has shown variable results [4,7,9,10]. SCC is a radiosensitive tumour and concurrent weekly chemotherapy with cisplatin and whole pelvis radiation has shown benefit, extrapolated from results for SCC within antecedent dermoids [11]. However, whether the same concept applies to de novo SCC is not clear.

In advance stages de-novo SCC behaves aggressively with recurrences within 5 weeks to 6 months [4,7], in spite of initial response to cytoreduction and chemotherapy. Overall survival in de-novo SCC ranges from 5 weeks to 24 months [4,7,10].

This patient underwent optimal debulking and was referred for post-operative radiation but was lost to follow up after 5 months of surgery.

Thus, albeit an uncommon tumor, pure primary SCC of the ovary can mimic other benign conditions and confuse the clinical picture. It is important to have a good pathology backup when trying to diagnose this rare tumor, whose incidence seems to be rising and a multidisciplinary management approach to maximise survival.

References


