PRIMARY SQUAMOUS CELL CARCINOMA OF OVARY - A CASE REPORT WITH REVIEW OF LITERATURE
Iffat Jamal

1 Tutor, Department of Pathology, All India Institute of Medical Sciences, Patna.

ABSTRACT

BACKGROUND
Ovarian squamous cell carcinoma is a very rare malignancy and its occurrence is generally seen associated with transformation of an existing ovarian dermoid cyst. The de novo occurrence of squamous cell carcinoma of the ovary in the absence of an antecedent ovarian dermoid is extremely rare. We report a case of 35-year-old female patient evaluated for abdominal distension. Abdominal CT was suggestive of a malignant neoplastic mass. Laparotomy was performed, which confirmed a malignant tumour involving the right adnexa and extending into the bowel and surrounding omentum. Surgical debulking, total hysterectomy with bilateral salpingo-oophorectomy, omentectomy and bowel resection was performed. Histopathological examination demonstrated squamous cell carcinoma arising from right ovary with no existing dermoid.

KEYWORDS
Primary, Squamous, Ovary, Dermoid, Neoplastic, Salpingo-Oophorectomy.

HOW TO CITE THIS ARTICLE: Jamal I. Primary squamous cell carcinoma of ovary- a case report with review of literature. J. Evid. Based Med. Healthc. 2016; 3(99), 5486-5490. DOI: 10.18410/jebmh/2016/1136

BACKGROUND
Squamous cell carcinoma of the ovary is a rare clinical entity accounting for less than 1% of all malignant tumours of the ovary.1 Malignant transformation of a preexisting mature cystic teratoma is considered to be the pathophysiological mechanism and this phenomenon is considered rare as only 1-2% of teratomas demonstrate this change. The de novo development of a primary squamous cell carcinoma in an otherwise healthy ovary is an extremely rare occurrence. We report a case of this rare malignancy, which has been very sparsely reported previously in medical field with review of literature.2

REVIEW OF LITERATURE
There are more than 30 different types of ovarian cancer, which are classified by the type of cell from which they start. Cancerous ovarian tumours start from three common cell types-

• Surface epithelium - cells covering the outer lining of the ovaries.
• Germ cells - cells that are destined to form eggs.
• Stromal cells - Cells that release hormones and connect the different structures of the ovaries.

1. Epithelial Tumours
Epithelial ovarian tumours develop from the cells that cover the outer surface of the ovary. Most epithelial ovarian tumours are benign (noncancerous). There are several types of benign epithelial tumours including serous adenomas, mucinous adenomas and Brenner tumours. Cancerous epithelial tumours are carcinomas- meaning they begin in the tissue that lines the ovaries. These are the most common and most dangerous of all types of ovarian cancers accounting for 85 to 90 percent of all cancers of the ovaries. Unfortunately, almost 70 percent of women with the common epithelial ovarian cancer are not diagnosed until the disease is advanced in stage.

There are some ovarian epithelial tumours whose appearance under the microscope does not clearly identify them as cancerous. These are called borderline tumours or tumours of Low Malignant Potential (LMP tumours).

2. Germ Cell Tumours
Ovarian germ cell tumours develop from the cells that produce the ova or eggs. Most germ cell tumours are benign (non-cancerous), although some are cancerous and maybe life-threatening. The most common germ cell malignancies are maturing teratomas, dysgerminomas and endodermal sinus tumours. Germ cell malignancies occur most often in teenagers and women in their twenties. Today, 90 percent of patients with ovarian germ cell malignancies can be cured and their fertility preserved.

3. Stromal Tumours
Ovarian stromal tumours are a rare class of tumours that develop from connective tissue cells that hold the ovary together and those that produce the female hormones, oestrogen and progesterone. The most common types are granulosa-theca tumours and Sertoli-Leydig cell tumours. These tumours are quite rare and are usually considered low-grade cancers with approximately 70 percent presenting as stage I disease (cancer is limited to one or both ovaries). Granulosa Cell Tumours (GCTs) are considered stromal tumours and include those composed of granulosa cells, theca cells and fibroblasts. GCTs account for approximately 2 percent of all ovarian tumours.

Financial or Other, Competing Interest: None.
Submission 21-11-2016, Peer Review 28-11-2016, Acceptance 08-12-2016, Published 12-12-2016.
Corresponding Author:
Dr. Iffat Jamal,
Flat No. D/D, Phase-1, Sapna Apartment,
Nayatola-800004, Bihar.
E-mail: iffatjamal111@gmail.com
DOI: 10.18410/jebmh/2016/1136
4. Primary Peritoneal Carcinoma

The removal of one’s ovaries eliminates the risk for ovarian cancer, but not the risk for a less common cancer called primary peritoneal carcinoma. Primary peritoneal carcinoma is closely related to epithelial ovarian cancer, which is the most common type. It develops in cells from the peritoneum (abdominal lining) and looks the same under a microscope. It is similar in symptoms, spread and treatment.

Ovarian cancer is the most common cause of cancer death from gynaecologic tumours in the United States. Malignant ovarian lesions include primary lesions arising from normal structures within the ovary and secondary lesions from cancers arising elsewhere in the body. Primary lesions include epithelial ovarian carcinoma (70% of all ovarian malignancies). Current research suggests that the majority of these originate from the fallopian tubes.

Stromal tumours of the ovary include germ cell tumours, sex cord stromal tumours and other more rare types. Metastases to the ovaries are relatively frequent. Common sources are tumours in the endometrium, breast, colon, stomach and cervix. See the image below. Ovarian cancer represents the sixth most commonly diagnosed cancer among women in the world and causes more deaths per year than any other cancer of the female reproductive system. Despite the high incidence and mortality rates, the aetiology of this disease is poorly understood. Established risk factors for ovarian cancer include age and having a family history of the disease while protective factors include increasing parity, oral contraceptive use and oophorectomy.

Lactation, incomplete pregnancies and surgeries such as hysterectomy and tubal ligation may confer a weak protective effect against ovarian cancer. Infertility may contribute to ovarian cancer risk among nulliparous women. Other possible risk factors for ovarian cancer include postmenopausal hormone replacement therapy and lifestyle factors such as cigarette smoking and alcohol consumption. Many of the causes of ovarian cancer are yet to be identified. Additional research is needed to better understand the aetiology of this deadly disease.

Up to a quarter of ovarian masses originate from germ cells and many of these are mature cystic teratomas, the secondary development of malignancy is a rare, but well-known phenomenon in patients with ovarian teratomas. Squamous cell carcinoma accounts for 80% of secondary malignant transformations of ovarian teratomas. We aimed to do an up-to-date systematic review of this rare malignant transformation. 64 suitable studies provided information on 277 patients. Squamous cell carcinoma in mature cystic teratoma was mainly found in women aged more than 50 years with high concentrations of squamous cell carcinoma antigen and cancer antigen CA125 and with ovarian tumours more than 100 mm in size. Patients with FIGO stage Ia tumours had better survival than those with more advanced disease. Complete resection together with hysterectomy, bilateral salpingo-oophorectomy and lymphadenectomy for patients with advanced disease followed by adjuvant chemotherapy with an alkylating drug was associated with higher survival, radiotherapy was not. We make proposals for investigation and treatment of this rare disorder.

Intraoperative sampling and frozen section interpretation is often requested at the time of surgery for intraoperative and postoperative planning. A diagnosis of SCC can be made provided that the histological features are present. The underlying aetiology should not be guessed at, however, given that extensive sampling of the surgical specimen is required to rule out the more common squamous lesions. Detailed examination of the cervix is required to rule out a cervical SCC, since ovarian metastases in such cases have been documented. With a diagnosis of ovarian SCC, full gynaecological staging should be undertaken including completion of total abdominal hysterectomy and bilateral salpingo-oophorectomy as well as peritoneal, omental and lymph node biopsies as indicated. Invasion of nearby tissues may require resection and reconstruction. In addition to surgery, adjuvant treatment may also be attempted. Some case reports have noted a response to early paclitaxel in combination with a platinum agent. However, outcomes remain poor in most cases.

CASE REPORT

A 35-year-old female para 3 was referred to General Surgery Department of Patna Medical College and Hospital, Patna, with a one-year history of progressive abdominal discomfort and distension. She had a history of weight and appetite loss. On clinical examination, abdomen was firm, tender mildly and a mobile mass was felt occupying most of the lower abdomen. There was no clinical evidence of ascites. Abdominal and pelvic CT scan with contrast demonstrated a heterogenous mass of size 5 x 3.2 x 1.2 cm in right adnexa with indention of the colon with the mass. She was operated upon and on exploration a solid mass was seen arising from right adnexa. There was evidence of peritoneal metastatic deposits in omentum and adjacent large intestine. Surgical removal of all grossly visible tumour was undertaken including performance of abdominal hysterectomy and bilateral salpingo-oophorectomy, sigmoid colectomy with primary closure of rectal stump and total omentectomy.

Grossly, there was grayish white tumour mass occupying whole of the ovary of size 5 x 3.2 x 1.9 cm. The fallopian tube was grossly looking involved by the tumour mass. (Figure 1 and 2).

Figure 1. Gross Photograph of Ovarian Specimen with Fimbrial End Identified

Microscopy revealed nest of malignant squamous cells with evidence of keratinisation in the form of some keratin pearls and individual cell keratinisation (Figure 3 and 4). The malignant cells were infiltrating the stroma and diagnosis of well-to-moderately differentiated adenocarcinoma of right ovary was given. The histopathology was remarkable for an absence of a concomitant teratoma or features suggestive of endometriosis. The histopathological features of retroperitoneal tumour deposits and the resected colon were identical to that of resected right adnexa (Figure 5). All the resected margins and lymph nodes were free of tumour.

The cervix, uterus, fallopian tubes and left ovary were histologically unremarkable. Adjuvant chemotherapy with cisplatin and etoposide was started and the patient was doing well.

**DISCUSSION**

SCC of the ovary presents an intriguing diagnostic challenge to clinical and pathology teams. SCC more commonly arises from non-ovarian sources making the above diagnosis exceedingly rare.\(^1\) When this entity occurs, however, it presents an interesting insight into the histopathological variation that maybe seen in epithelial malignancies of the ovary. Primary ovarian lesions are classified into the epithelial, germ cell or sex cord stromal categories.\(^3\) Mature teratomas (dermoids) included in the germ cell category are the single most common ovarian tumour and can occur at any age. Squamous elements are most commonly identified in ovaries as part of a mature teratoma Although, the
Squamous component in a teratoma is often benign, SCC can arise from mature teratomas; this entity, in fact is the most common malignant component arising from a mature teratoma. Only one to two percent of mature teratomas harbour a malignant component, but up to 80% will be squamous. Squamous elements arising in the absence of a teratomatous component (i.e., arising as a purely epithelial lesion) are distinctly rare. Generally, these occur as metastases from extra-ovarian squamous lesions or as part of a metaplastic process in an endometrioid adenocarcinoma or Brenner tumour. Endometrioid adenocarcinoma of the ovary typically presents postmenopausally and is typified by areas of squamous differentiation arising within neoplastic endometrioid glands. Brenner tumours are primary ovarian tumours that occur chiefly at age 40-50 and show predominantly transitional (urothelial) differentiation; these more infrequent tumours may also show areas of squamous differentiation. Rarer still are SCCs showing none of the above features. Some primary pure SCCs will arise in concert with foci of endometriosis and rarely others are noted entirely de novo. SCC of the ovary generally behaves aggressively. The lesion is typically identified as an enlarging pelvic mass coinciding with symptoms of abdominal pain in the absence of significant ascites. Other symptoms are related to tumour invasion of neighbouring structures such as the urinary system and other gynaecologic organs and to distant metastases. There are no pathognomonic radiological features in SCC, pelvic imaging remains important in the workup of ovarian malignancies. The radiologic and gross pathologic findings generally show a heterogeneous solid and cystic mass approximately 10-15 cm in maximal dimension. This latter fact may helpful in sorting out the preliminary differential diagnosis given that many aggressive ovarian neoplasms are often much larger. Areas of necrosis are often visible and there are often adhesions to surrounding pelvis. The diagnosis of SCC of ovary can only be made histopathologically. It is identified by the presence of architectural and cytological features that resemble those found normally in squamous elements, but that also show evidence of invasion. As in other SCCs throughout the body, the clinically aggressive character of SCC of the ovary is recapitulated histologically by its tendency to form invasive ribbons and tongues of tumour cells that may extend well beyond the original tumour focus. In its well-differentiated form, SCC will show squamous maturation, keratin formation and intracellular bridging. In its poorly-differentiated form, few normal squamous features maybe identifiable; such cases may require ancillary immunohistochemical or electron microscopic studies to confirm the diagnosis. Infrequently, SCC may show "pseudo gland" formations. These structures may confuse the diagnosis especially in tissue more likely to harbour an adenocarcinoma than an SCC. In the latter scenario, the use of immunohistochemical markers specific for squamous elements can be very helpful. Intraoperative pathology consultation comprising of tissue sampling and frozen section interpretation is often requested at the time of surgery for intraoperative and postoperative planning. A diagnosis of SCC can be made provided that the histological features are present. The underlying aetiology should not be guessed at, however, given that extensive sampling of the surgical specimen is required to rule out the more common squamous lesions. Detailed examination of the cervix is required to rule out a cervical SCC since ovarian metastases in such cases have been documented. With a diagnosis of ovarian SCC, full gynaecological staging should be undertaken including completion of total abdominal hysterectomy and bilateral salpingo-oophorectomy as well as peritoneal, mental and lymph node biopsies as indicated. In addition to surgery, adjuvant treatment may also be attempted. Some case reports have noted a response to early paclitaxel in combination with a platinum agent. However, outcomes remain poor in most cases. In particular, most cases (80% in one study) of SCC arising with or without endometriosis result in death within a few months of diagnosis.

**CONCLUSION**

Although, squamous elements are quite frequently present in ovarian lesions. Primary squamous cell carcinoma of ovary is quite rare in occurrence. There are debates going on the pathogenesis of primary squamous cell carcinoma without any associated dermoid. Some of the authors believe that squamous cell carcinoma of ovary arises is mostly associated with endometriosis and there is neoplastic transformation of the endometrial glands plays the key role in the pathogenesis of squamous cell carcinoma. But, the pathogenesis behind de novo squamous cell carcinoma of ovary is still not known.

**REFERENCES**


