

# **De novo primary squamous cell carcinoma of the ovary: A case of a rare malignancy with an aggressive clinical course**

Ali Imran Amjad, Inam Pal

Department of Surgery, Aga Khan Univeristy Hospital, Karachi, Pakistan.

### **Abstract**

Ovarian squamous cell carcinoma is a rare malignancy and the occurrence is attributable to malignant 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. The case of a 31 year old Asian woman, evaluated for abdominal distension and discomfort is presented. Abdominal CT was suggestive of a malignant neoplastic process. Laparotomy confirmed a malignant tumour with involvement of the right adnexa and extension into the omentum and bowel. Surgical debulking, hysterectomy, bilateral salpingo-oophorectomy and total omentectomy and bowel resection was performed. Histopathology demonstrated squamous cell carcinoma arising from the right ovary with no co-existing ovarian dermoid. The postoperative period was significant for disease progression despite adjuvant chemotherapy.

### **Introduction**

Squamous cell carcinoma of the ovary is a rare clinical entity, accounting for less than 1% of all malignant tumours of the ovary.<sup>1</sup> Malignant transformation of a pre-

existing mature cystic teratoma (dermoid cyst) is appreciated as the underlying pathophysiological mechanism, and this phenomenon is considered rare, as only 1-2% of teratomas demonstrate this change.<sup>1</sup> The de novo development of a primary squamous carcinoma, in an otherwise healthy ovary is an extremely rare occurrence.<sup>2-5</sup> We present a case of this rare malignancy, not been previously reported in the Southeast Asian population, with a review of literature.

### **Case Report**

A 31 year old, para 2, Asian female (Pakistani heritage) was referred to the general surgery clinic with a one year history of progressive abdominal discomfort and distension. She had a history of weight loss with no change in appetite, bowel habit or menstrual flow. A recent pap smear was negative for dysplasia. On examination, she was anaemic with no jaundice, lymphadenopathy and had normal breast examination. Abdominal examination revealed a firm, mildly tender, mobile mass occupying most of the lower abdomen. There was no clinical evidence of ascites. Pelvic and rectal exams were unremarkable.

Pre-operative evaluation showed anaemia with normal cell counts, and normal kidney and liver functions.



Figure 1. Contrast-enhanced CT scan of the pelvis showing the tumour mass.

An abdominal and pelvic CT scan with contrast demonstrated a heterogenous mass (5.5 x 10 x 13 cm) occupying the lower abdomen appearing to arise from the bowel mesentery (Figure 1).

The patient presented to the emergency room with increased abdominal pain and fever before the planned elective surgery and an exploratory laparotomy was performed. Intra-operative features suggested peritonitis with contamination by large bowel contents. On exploration, the solid mass was seen to be arising from the right adnexal area. An area of the sigmoid colon was noted to have a perforation where it was adherent to the mass. There was evidence of peritoneal metastatic disease with omental involvement and deposits on the small bowel mesentery. An intra-operative frozen section on one of the tumour deposits suggested a malignant tumour of squamous origin.

Surgical removal of all grossly visible tumour was undertaken, including performance of a total abdominal hysterectomy and bilateral salpingo-oophorectomy, sigmoid colectomy with primary closure of the rectal stump and exteriorization of the proximal bowel as a mucus fistula. Total omentectomy and limited small bowel resection with an end ileostomy was also performed. The immediate postoperative course was remarkable for abdominal wound infection requiring periodic debridement and antibiotics.

The histopathology confirmed a well differentiated squamous cell carcinoma arising from the right ovary, with evidence of keratinisation. The pathology was remarkable for an absence of any normal ovarian parenchyma, a concomitant teratoma or features suggestive of endometriosis. The histopathological features of the retroperitoneal tumour deposits and the resected ileum were identical to that of the resected adnexa. All the resection margins and the three recovered lymph nodes were tumour

free. The cervix, uterus, fallopian tubes and the left ovary were histologically unremarkable; the clinical staging for ovarian tumour was thus consistent with FIGO stage IIIc.

Despite initiation of adjuvant chemotherapy with cisplatin and etoposide, over the course of the next three months, there was clinical and radiographic evidence of tumour progression with metastases to the liver, mesenteric and pelvic lymph nodes.

## Discussion

Ovarian cancer remains the second most common female genital tract malignancy. Mature cystic teratomas, commonly known as dermoids, represent over 10% of the ovarian tumours.<sup>1</sup> On rare occasions (1-2%) the dermoids provide a background for malignant transformation within components of the teratoma; the majority of such malignancies arising within an ovarian dermoid (80-90%) are squamous cell carcinomas.<sup>1</sup> Malignant transformations have also been reported in endometriotic ovaries.<sup>3,6</sup> Reports of primary squamous cell carcinomas arising in otherwise healthy ovaries are markedly rare<sup>2-5</sup>; such an occurrence has not previously been described in a patient of Southeast Asian heritage.

In contrast to the described presentation, carcinomas reported to arise within a background of an ovarian dermoid are likely to be an incidental histological finding. A few predictive risk factors associated with such malignancy are recognized, namely advancing age, tumour size (higher likelihood appreciated with larger tumour sizes), radiographic characteristics and tumour markers (CA-125).<sup>1</sup>

Of the reported cases of pure squamous cell carcinoma of the ovary in the literature, the majority were associated with cervical dysplasia.<sup>3,7</sup> Such an association however was not apparent in our case as demonstrated by a negative pre-operative pap smear and confirmed by post operative surgical pathology.

Due to the rarity of the condition, as well as an incidental nature of the diagnosis, the optimal management approach to primary ovarian squamous cell carcinomas remains unclear. Reported literature suggests that the surgical management for the described cases is akin to that for adenocarcinomas of the ovary, entailing performance of total abdominal hysterectomy, bilateral salpingo-oophorectomy and omentectomy, with additional steps as needed to ensure surgical debulking of all grossly visible disease. Indeed, optimal cytoreduction and the surgical stage at presentation has been correlated with a significantly improved survival for squamous cell carcinomas arising from dermoid cysts.<sup>8</sup> While a grossly optimal surgical debulking was achieved at the primary surgery, the existing

peritoneal contamination is likely to have contributed to a failure to achieve optimal cytoreduction in the case presented.

The role of adjuvant therapy in the management of ovarian squamous cell carcinomas is similarly unclear. Limited experience suggests that surgical management with close follow up alone would suffice for early stage IA disease.<sup>7</sup> Stages IC and above have been targeted with a variety of adjuvant regimens with variable outcomes. Relatively poor disease control has been noted with the conventional regimens to treat epithelial ovarian cancers, i.e cisplatin, vincristine, mitomycin C, and bleomycin (POMB).<sup>9</sup> On the other hand remarkable responses have been achieved with paclitaxel based regimens in some cases.<sup>9,10</sup>

Radiotherapy has also been used with the rationale of squamous cell carcinoma being a radiosensitive tumour; and has brought forth variable results for cancers within antecedent dermoids.<sup>1,8</sup> In the latest case series and review of literature<sup>1</sup>, albeit in cases of squamous cell cancers arising in mature cystic teratoma of the ovary, whole-pelvis radiation and concurrent weekly platinum-based chemotherapy following aggressive cytoreduction has been shown to be of benefit. It however remains unclear whether patients with de novo primary squamous cell cancer of the ovary would benefit as much from similar adjuvant therapy.

To conclude primary squamous cell cancers arising in the ovary is an extremely rare entity, and has not previously been described in patients of Southeast Asian heritage. In the absence of well designed trials, and based on the existing albeit sparse data, the primary management

approach at present is surgical debulking, akin to the principles underlying the management of ovarian adenocarcinoma. Combination chemotherapy with newer drugs has shown some benefit and pelvic radiotherapy may have a role to play for local control. However, in the absence of quality data, except for the very early stages of presentation, a role for adjuvant therapy is at present unclear.

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