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CASE REPORT

Ovarian Carcinosarcoma: Rare Histology Which Never Fails to be Aggressive

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Introduction

In total, 1–4% of all ovarian malignancies are ovarian carcinosarcomas. Uterine sarcomas are more frequent in the pelvis than ovarian primary sarcomas, which are described infrequently [1]. In this publication, along with a brief review, we are reporting a case of ovarian carcinosarcoma that was treated at a non-oncology centre at the outset and referred to us after an incomplete operation.

Case Details

A 44-year-old nulliparous female who had undergone right oophorectomy and subtotal hysterectomy one month prior for an ovarian tumour presented to the outpatient department. Any intraoperative notes could not be traced out. The histopathological analysis of the ovarian mass revealed a malignant spindle tumour with mitotic activity > 6–7/10 HPF. There was a fascicular pattern of mild to moderate atypia, widespread palisaded necrosis, with sporadic fragmented epithelioid components (Fig. 1). Uterus and fallopian tubes were free of tumour. The immunohistochemistry performed at our institute was suggestive of stromal expansion in a carcinosarcoma/mixed mullerian tumour with CK patchy positivity, vimentin focal positivity, calretinin focal positivity, SMA negative, desmin negative, and S 100 negative. The contrast-enhanced computed tomographic imaging showed a 6*3 cm residual lesion in the right adnexal area, near to the hysterectomy stump, with peri-lesional streakiness and bladder wall adhesion (Fig. 2a). The serum CA-125 level was increased to 60 IU/ml, while all other ovarian serum tumour markers were normal. The patient had a completion surgery encompassing left oophorectomy, excision of the remnant of lower part of uterus and cervix and right ovary, right pelvic peritonectomy, removal of deposits from recto-sigmoid and bladder peritoneum, total omentectomy, bilateral pelvic lymph node sampling, and para-aortic lymph node sampling along with removal of anterior abdominal wall peritoneal deposit. Completion of cytoreduction score of 0 was achieved. Microscopic sections from greater omentum, right pelvic peritoneum and pelvic, small bowel and rectal deposit showed the presence of atypical spindle and ovoid cells, consistent with metastatic deposit from ovarian carcinosarcoma. Pelvic and para-aortic lymph nodes were reactive. The platinum-based adjuvant chemotherapy was started after three weeks of surgery and was tolerated well. Within one month of completion of chemotherapy, imaging done for response evaluation showed features of progressive disease. Contrast-enhanced imaging revealed 36*20 mm cystic lesion in right side of pelvis. Another 21*19 mm deposit was noted in left side of pelvis (Fig. 2b,c). The patient was put on second-line lipodox-based chemotherapy thereafter.

Discussion

The average age of presentation of ovarian carcinosarcoma is between sixty and seventy years [1]. Though the pathogenesis of gynaecological carcinosarcoma remains poorly understood, it revolves around following theories: biclonal (collision) and monoclonal (combination and conversion)

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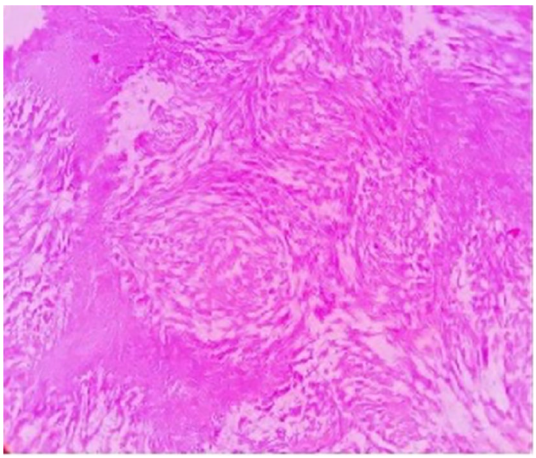

Fig. 1 Histopathological picture of ovarian carcinosarcoma

theories. The collision theory suggests consolidation of two independent histologies that originated in different cells. According to the combination theory, early in the genesis of tumours, a single-stem cell differentiates into two components. The conversion theory which views carcinosarcoma as dedifferentiated carcinomas of the ovary postulates that an epithelial cell passes through metaplastic differentiation to give rise to a sarcomatous component.

The most recent suggested pathway for these tumours' growth is the epithelial–mesenchymal transition and heterologous differentiation [2]. The tumour is frequently unilateral and solid, and the greater omentum is the most common site of metastatic spread. The high incidence of haemorrhagic ascites is one of the most striking clinicopathological findings. Ovarian carcinosarcoma is histologically divided into sarcomatoid and epithelial components. The epithelial component in the ovary might be endometrioid, clear cell, serous, or squamous epithelium. The endometrial stromal, fibro, and leiomyosarcomas that make up the mesenchymal/sarcomatous component may be native to the ovary, also known as homologous. The heterologous elements, being non-native to ovary, include osteosarcoma, rhabdomyosarcoma, liposarcoma, or chondrosarcoma [2].

The newly diagnosed and relapsed ovarian carcinosarcoma is managed in similar lines with epithelial ovarian cancer. The disease is staged according to the revised TNM or FIGO staging (2009) standards [3]. The initial approach to diagnosis includes patient's detailed medical and surgical history, physical examination, routine blood tests, tumour markers (CEA, CA-125), and in addition, if there has been prior histological confirmation, chest radiography, mammography, gastroscopy and colonoscopy, ultrasound, and/or CT scan of the abdomen and pelvis. Hysterectomy and bilateral salpingo-oophorectomy and comprehensive surgical staging (or debulking as needed) is the primary treatment as in line with any epithelial ovarian cancer. Poor surgical candidates are managed initially with neo-adjuvant therapy. There is no well-established consensus regarding chemotherapy and the optimal regimen; platinum-based or non-platinum-based. NCCN 2021 recommends paclitaxel/carboplatin as the preferred regimen. For stage I, monitoring/follow-up with tumour marker is advocated. For stage II–IV, maintenance therapy is considered if BRCA1/2 mutation is known.

Several novel molecular targets, including EGFR, c-Kit, Cox-2, Her-2-neu, and VEGF, are being studied and may provide future treatments [3]. Ovarian carcinosarcomas are aggressive tumours that differ from serous malignancies in their natural history. With a median survival of fewer than two years, the prognosis is worse than

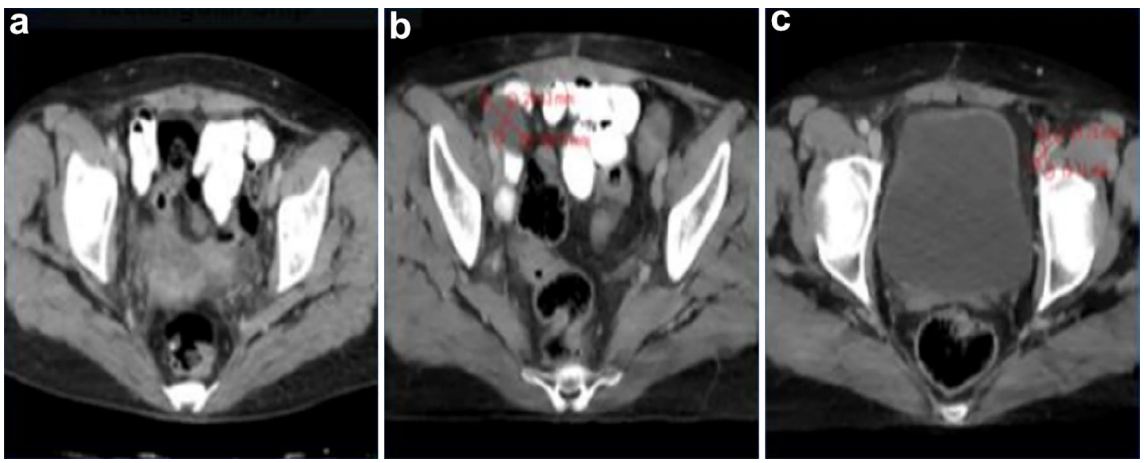

Fig. 2 (**a**) Axial cut of contrast-enhanced computed tomographic imaging showed a 6*3 cm residual lesion in the right adnexal area, near to the hysterectomy stump, with peri-lesional streakiness and bladder wall adhesion (**b**) Platinum-resistant recurrence: 36*20 mm cystic lesion in right side of pelvis. (**c**) Another 21*19 mm lesion in left side of pelvis

the latter [4]. Stage I carcinosarcomas have a 5-year survival rate of 65.2% (95% CI, 58.0–71.4%) compared to serous tumours 80.6% (95% CI, 78.9–82.2%). Similarly, the 5-year survival rate for patients with stage IIIC carcinosarcomas is 18.2% (95% CI, 14.5–22.4%) as opposed to 33.3% (95% 32.1–34.5%) for serous carcinomas. Optimal cyto-reduction, homologous subtype, < 60 years of age, early stage, paclitaxel/platinum therapy are favourable prognostic factors in terms of disease relapse and survival. Negative prognostic factors are: overriding sarcomatoid element more than 25% and especially the heterologous one, disease stage, tumour grade, large size of the initial tumour, residual disease after surgery, p53 mutation, VEGF expression, and the long-term tamoxifen use [4].

Conclusion

Ovarian carcinosarcomas are aggressive tumours that differ in their natural history from serous malignancies. Due to the rarity of carcinosarcoma, prospective data are sparse, and case reports, series, and observational studies help to understand the natural course of the disease. This case study emphasises the need to broaden our perspective in order to accurately diagnose and timely treat the unusual histologies. This case further supports the disease's aggressive behaviour as described in earlier literature.

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Declarations

Conflict of interest The authors declare no conflict of interest.

Informed consent Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

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