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Case Report

Carcino-sarcoma of ovary in young female: a rare presentation with review of literature

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ABSTRACT

Carcino-sarcoma is a malignant mixed mesodermal neoplasm accounting for less than 1% of all ovarian tumors. It is an aggressive tumor, composed of both malignant epithelial and mesenchymal components. To the best of our knowledge, less than 400 cases of ovarian carcino-sarcomas have been reported in the literature and only 10% of them are bilateral. In this paper, we report a new case of unilateral ovarian carcino-sarcoma in a 20-year-old adolescent girl with the aim to analyze it with its possible differentials, treatment and prognosis due to rarity of its occurrence in young age.

Keywords: Carcinoma, Ovary, Young age, Sarcoma

INTRODUCTION

Ovarian carcinosarcoma is responsible for 1-4% of all ovarian tumors.[1] Carcinosarcoma is found not only in the ovary but also in other organs of the genito-urinary tract, including uterus, fallopian tubes, breast, and urethra.[2] The peculiar feature of this rare malignancy is that it involves both cellular components of epithelium and mesenchyme of the affected organ. The contribution of each element in the development of the malignancy is different and for this reason, the preoperative diagnosis of the disease cannot be reliably made by core biopsy or FNA.

CASE REPORT

A 20-year-old non-diabetic and non-hypertensive female presented with distension of abdomen since 7 months duration. She also had a complaint of amenorrhea and pain in the lower abdomen since 5 month. There was no history of per vaginum discharge, loss of weight and appetite. There was no history of trauma, past treatment or surgery. On examination, her abdomen was distended. She had a vague lump of size about 9×8 cm present in the lower abdomen. The lump was extending up to the epigastrium and was mobile. The lower part of the lump could not be palpated. Per rectal examination did not reveal any abnormality. Chest X-ray was within normal limits. The ultrasound examination of the abdomen showed an ill-defined heterogenous lesion with multiple cystic spaces in pelvic region. The contrast-enhanced computed tomography (CT) of the abdomen showed a large, hypodense, rim enhancing, multilobulated lower abdominal mass of 12x10x9 cm with enhancing mural component, arising from the right ovary. The left ovary and fallopian tube appeared to be normal. The level of cancer antigen (CA-125) was elevated while routine hematological and biochemical investigations were within the normal limits. Suspicion for ovarian malignancy prompted an exploratory midline laparotomy.

The intra-operative findings showed a right side ovarian cyst measuring 12×10×9 cm which was adherent to mesentery and part of the small bowel. There were no significant para-aortic or pelvic lymph nodes. No peritoneal and omental deposits along with free fluid was identified per operatively. Tumour appeared reddish-grey

in colour with complex solid cystic and heterologous components.

Grossly right ovarian cyst with attached fallopian tube was received in multiple pieces, of which largest measured 10×8×8 cm and fallopian tube measured 2.5 cm in length (Figure 1). Cut surface shows predominantly solid component with focal areas of haemorrhage as well as multiple cystic spaces filled with mucoid gelatinous material. Microscopy revealed tumour disposed predominantly in solid areas composed of elongated or spindle shaped cells disposed in bundles or fascicles. Other areas showed glandular pattern or varying sized cystic spaces comprising of focally stratified columnar cells with moderate anisocytosis and filled with mucinous material (Figure 2).

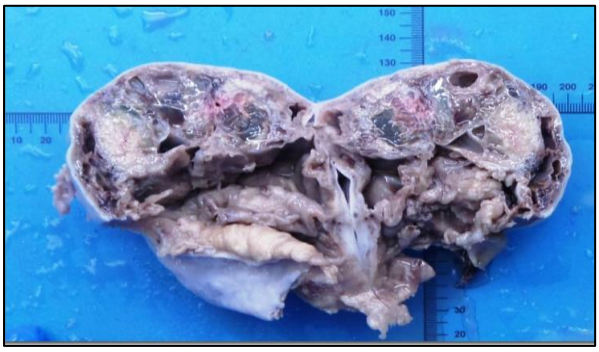

Figure 1: Cut surface shows predominantly solid grey-white component with focal areas of haemorrhage as well as multiple cystic spaces filled with mucoid gelatinous material.

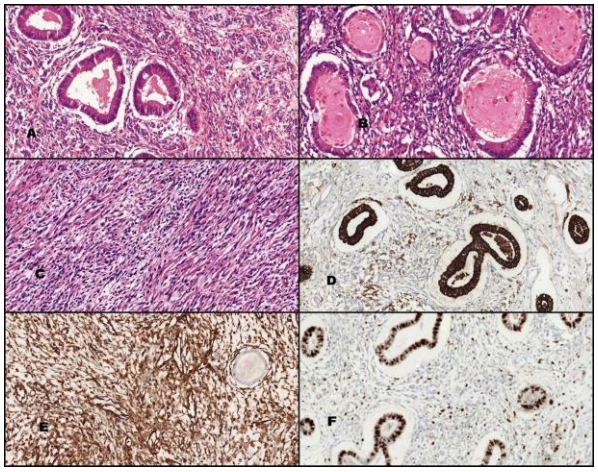

Figure 2: (A) Section show glandular pattern comprising of columnar cells (20X H and E); (B) section show varying sized cystic spaces showing similar cells (20X H and E); (C) pre-dominant fascicular pattern of elongated spindle cells (20X H and E); (D) strong membranous PanCK positive glandular component (10X, IHC); (E) diffuse vimentin positive sarcomatous component (10X, IHC); and (F) high Ki67 index ~35-40% in both areas (10X, IHC).

With the above histo-morphology; high grade mucinous cysta-denocarcinomas with mural nodules, adenosarcomas and carcinosarcomas were kept as possible differentials and immuo-histochemistry panel was applied.

On immunohistochemistry, the epithelial component stained strongly with pan CK and mesenchymal component showed strong vimentin and desmin positivity. Ki 67 index was high ~30-35%. It was negative with CK7, CK20 and CEA thus ruling out above differentials. Negative staining reaction was also seen with calretinin, CD10, myogenin, CD34 and S100. Following surgery patient underwent six cycles of chemotherapy and was responding well to the treatment.

DISCUSSION

Malignant mixed mullerian tumours **(**MMMT's) or ovarian carcinosarcomas (OCS) are rare and highly aggressive cancers of the female genital tract with poor long-term prognosis.[3] OCS is usually encountered after menopause, with a median age of 65 years. The most common symptoms are abdominal pain, distension, early satiety, bloating, nausea, vomiting, and weight loss. The risk factors are advanced age, obesity, nulliparity, exogenous oestrogen, radiotherapy and long-term tamoxifen usage. Patients presents with advanced stage (III–IV) and spread beyond the ovary (>90%) at the time of diagnosis.[4] Recent study suggests a monoclonal theory of histogenesis for ovarian carcinosarcomas. It states that there is metaplastic transformation of the epithelial component, which gives rise to the sarcomatous component. In ovarian carcinosarcoma, there is the presence of both intermixed malignant epithelial and stromal component. Boucher et al in their series of ovarian carcinosarcomas, have described equal representation of the epithelial endometrioid and serous component types. The mesenchymal component was largely heterologous, composed of chondromatous and rhabdomyoblastic differentiation.[5] However in our case epithelial component comprised of mucinous carcinoma predominantly and mesenchymal component showed fibrosarcomatous differentiation.

There is no existing consensus for the treatment of ovarian carcinosarcomas. However, the mainstay of treatment relies on optimal surgical cytoreduction followed by platinum-based chemotherapy. Combination chemotherapy with ifosfamide and cisplatin or taxol and carboplatin is favored. This results in improved progression-free survival according to large retrospective series.[6,7] The prognosis of ovarian carcinosarcoma is poor with a median survival reported in the majority of the studies around 11 months.[8]

CONCLUSION

Ovarian carcinosarcoma is an extremely uncommon tumor with an aggressive course and dismal prognosis. Such cases must be meticulously recorded for future reference

with regard to the treatment provided and outcome achieved.

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