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Case report

BILATERAL OVARIAN CARINOSARCOMA-A RARE ENTITY

*Sinkar Prachi¹, Pande Pankaj², Yelikar BR.³

¹Post Graduate Student, ²Associate Professor, ³Professor & Head of Department, Department of Pathology, Shri B.M. Patil Medical College, Hospital & Research Centre, BLDE University, Vijayapur, Karnataka, India

*Corresponding author email: doc.prachi@gmail.com / prachi.doc@gmail.com

ABSTRACT

Malignant mixed Mullerian tumor (carcinosarcoma) of the ovary is rare neoplastic condition with an incidence of less than 1% of all ovarian neoplasms. Histologically, carcinosarcomas comprise of epithelial as well as mesenchymal components, which are either homologous (normally found in ovary) or heterologous (not normally seen in ovary). Here, we report a case of a 50 year old female patient who presented with abdominal distension and was diagnosed as malignant mixed mullerian tumor of bilateral ovaries histopathologically. Carcinosarcomas of the ovary are extremely rare and aggressive. We wanted to draw the attention that although it is more frequently unilateral and seen among the postmenopausal nullipara women, malignant mixed müllerian tumors can also be bilateral and seen among multiparas in the reproductive period as with this case report.

Keywords: Bilateral, Carcinosarcoma, Mixed Tumor, Ovarian Neoplasms.

INTRODUCTION

Ovarian carcinosarcoma, also called malignant mixed mullerian tumour (MMMT), is a very rare ovarian neoplasm, with an incidence of less than 1% of all ovarian tumors, and less than 400 case reports in literature¹. Histologically, carcinosarcomas comprise of epithelial (carcinomatous) as well as mesenchymal (sarcomatous) components both, which are either homologous (normally found in ovary) or heterologous (not normally seen in ovary)^{2,3}. These tumors are often seen in the 5th to 7th decade in postmenopausal women who are nulliparous. They are usually asymptomatic. Only 10% of them are bilateral.²⁻⁴ Despite aggressive treatment which includes surgery and chemotherapy, patients have an increased risk of death compared to women with epithelial ovarian cancer⁴. Here we report a case of malignant mixed mullerian tumour of the bilateral ovaries in a 50 year old female with complains of abdominal distension where a total abdominal

hysterectomy and bilateral salpingo-oophorectomy was performed.

CASE REPORT

A 50-year-old woman, gravida 2, para 2, consulted to the outpatient department of Obstetrics and Gynecology with complaints of abdominal distention since two months. Physical examination was unremarkable except for bilateral masses per abdomen. There were no other complaints and she was previously doing well. History of irregular bleeding or any such significant past history was negative. Blood, urine and biochemical investigations of patient were within normal limits. However only an increase in CA-125 (60.2 m/ml) and CA-19-9 (20.4 m/mL) was noted. USG revealed cystic lesions in both ovaries left ovary measuring 14x7 cms and right ovary measuring 9x7 cms.

Ovarian cancer was suspected, so the patient underwent exploratory laparotomy. Optimal debulking surgery was performed, and specimen was sent for histopathological study.

Pathology: On gross appearance, an already cut open unoriented specimen of both ovaries was received which was fleshy, bulky and polypoidal, larger mass measuring 13x9x6cms and smaller mass measuring 9x7x4cms. External surface showed numerous fragmented pieces which were soft, encephaloid, grey, black, glistening with areas of blackish discoloration and focal areas of hemorrhage. Cut section -Solid, grey white fragments showing variegated appearance with areas of hemorrhage, necrosis and multiple small cysts were noted. No normal ovarian tissue was noted.

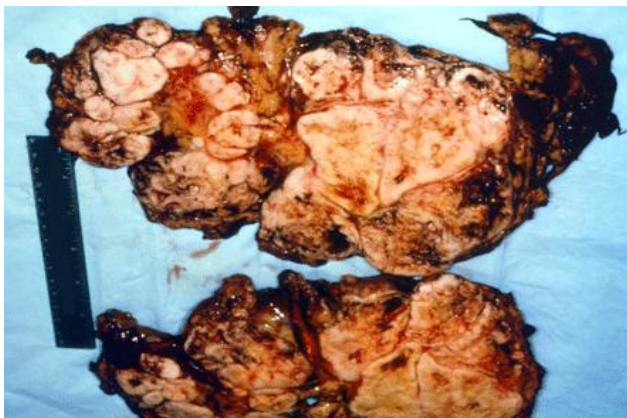


Fig 1: Gross photograph of the ovary showing areas of hemorrhage and necrosis.

On light microscopy, the tumor had a biphasic pattern, which consisted of two components: poorlycarcinomatoid and dominantly sarcomatoid. Carcinomatoid component consisted mostly of glandular formations of pleomorphic large-round cells, polygonal hyperchromatic nuclei and inconspicuous nucleoli with moderate amount of vacuolated light cytoplasm.

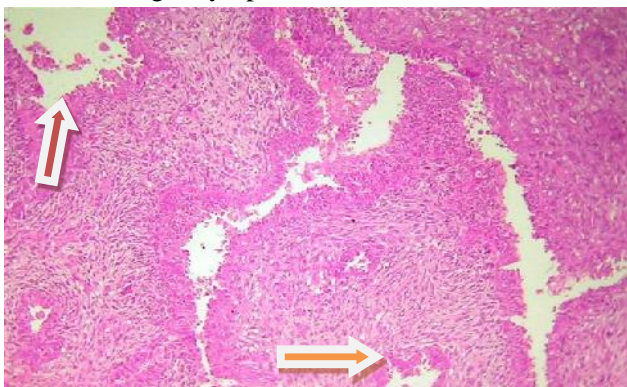


Fig 2:Microphotograph showing both carcinomatous (brown arrow) and sarcomatous components (yellow arrow) components.(H&E 10x)

In the sarcomatoid component, individual and small groups of trapped malignant cells were found, as well as multi-nucleated (bizarre) cells next to the areas of tumor necrosis. High mitotic activity, proliferating

blood vessels, dense areas of necrosis and focal areas of chronic inflammation were seen.

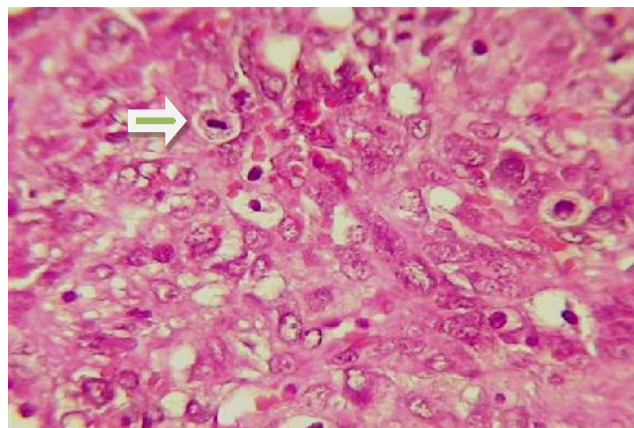


Fig3: Microphotograph showing high grade tumor cells with mitotic activity.(arrow head) (H&E 40x)

Immunohistochemical analysis revealed positivity for vimentin in confirming stromal component and for EMA confirming the epithelial component.

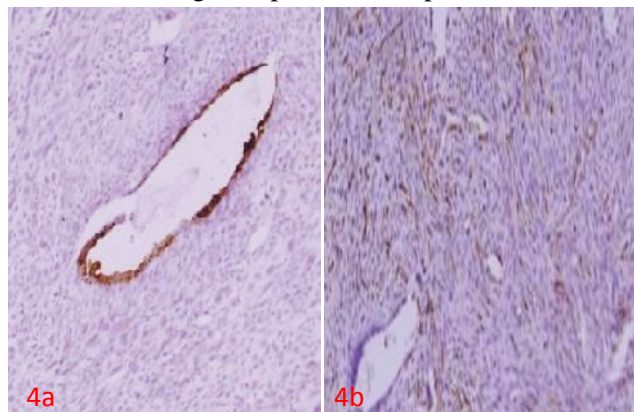


Fig4:(a)Epithelial component showing positivity with epithelial membrane antigen (EMA)4(b)Stromal component showing positivity with vimentin (Vim)

Based on these histopathological and immunohistochemical characteristics of the tumor cells, Malignant Mixed Mullerian Ovarian tumor was confirmed. (Carcinosarcoma)

DISCUSSION

Mixed mullerian tumors are extremely rare tumours of genital system seen in postmenopausal women with a peak incidence in the sixth decade of life. They are often localized in the uterine corpus, but can also be found in the uterine cervix, the tubes and the ovaries. They develop from mesenchymal (Mullerian) cells that can be differentiated into epithelial and stromal elements⁴ The tumor consists of homologous or heterogeneous epithelial (carcinomatoid) and mesenchymal (sarcomatoid) components of cells in

different mutual relationships. The heterologous sarcomatous component arises from nonnative elements such as rhabdomyoblastic, osteogenic, chondroblastic, or lipoblastic elements. The epithelial component can be endometrioid, undifferentiated, clear cell, or serous consisting of one or more types of carcinomas, the most common being adenocarcinoma (serous, mucous, papillary, endometrial or the light-cell type), or anaplastic carcinoma. On the other hand, within the group of malignant mesenchymal component, the most common are the homologous sarcomas (fibrosarcoma, angiosarcoma and leiomyosarcoma), although some cases of heterologous sarcoma were also described.^{4,5}

The histogenesis is not clear yet. Some authors are of the opinion that there is transformation of the epithelial cells into sarcomatoid ones (metaplastic theory), while the others by the usage of the immunohistochemical analysis and the cell culture, point out the epithelial like characteristics of both kinds of tumor cells. The cellular heterogeneity of tumor by the co-expression of some of the epithelial (Cytokeratin, CEA, EMA) and mesenchymal antigens (Vimentin, Desmin) was proved. Such co-expression of the antigens supports the hypothesis that the epithelial and mesenchymal elements, which create the MMT of the ovaries, descend from a common cellular precursor - the stem cell.⁶

MMTs usually occur in postmenopausal women, but occasionally occur in relatively younger patients. Some ovarian germ cell tumours can sometimes be quite challenging in histological diagnosis. Mixed GCTs can mimic malignant mixed Mullerian tumors. Yolk sac tumours can display multiple morphological patterns and can mimic different types of carcinoma such as clear cell carcinoma or endometrioid adenocarcinoma. Cytoreduction has proven to have an impact on survival. Chemotherapy does not appear to be beneficial.⁵

To summarize, malignant mixed mullerian tumors or carcinosarcomas of the ovaries are very aggressive tumors with a very poor prognosis. They are diagnosed at an older age of about 5-7th decade in post menopausal women. As in this case, MMT usually have reached an advanced stage at the time of diagnosis, and survival varies with stage and histological type. Despite aggressive treatment which includes surgery and chemotherapy, patients have an

increased risk of death compared to women with epithelial ovarian cancer.^{7,8}

CONCLUSION

In conclusion, we have described a rare case of carcinosarcoma (homologous type of malignant mullerian tumor) of bilateral ovaries that presented with abdominal distension for two months. These tumors are usually seen in the 5th to 7th decade and are usually asymptomatic. Only about 10 % of them are bilateral and are frequently encountered in nulliparous women. More than 80% of the patients had an extra ovarian abdominal spread at the time of diagnosis^{3,8}.

We wanted to throw light that although it is more frequently unilateral and seen among the postmenopausal nullipara women, malignant mixed müllerian tumor can also be bilateral and seen among multiparas in the reproductive period as with this case owing to its rarity.

Conflict of Interests: Nil

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