

Malignant Mixed Mesodermal Tumor of Ovary in Young Female: A Rare Case Report

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Abstract

Carcinosarcoma, also known as malignant mixed mesodermal tumor (MMMT), occurs predominantly in postmenopausal women of low parity these are very rare neoplasms associated with an aggressive clinical course and overall poor prognosis, stage is best predictor, and most patients present at advanced stage. Uterus and ovary are common sites for MMMT, though it can occur anywhere along the female genital tract and in the peritoneum. Microscopically, carcinosarcoma is a biphasic neoplasm with intermixed epithelial and mesenchymal elements. The sarcomatous component can be homologous or heterologous depending on whether it is composed of native mesenchymal elements of the organ it arises from or other non-native elements such as rhabdomyoblastic, osteogenic, chondroblastic or lipoblastic element. The epithelial component can be endometrioid, undifferentiated, clear cell, or serous. We report a case of MMMT in 22-year-old parous women who present to a gynecologic department with the complaints of the abdominal mass, pain, vaginal bleeding. Histopathological examination confirms the diagnosis of MMMT.

Key words: Carcinosarcoma, Mixed mesodermal tumor, Ovary

INTRODUCTION

Ovarian carcinosarcomas, also called as malignant mixed mesodermal tumor (MMMT), is a rare variant of ovarian cancer, accounting for <1% of all ovarian tumors.¹

MMMTs of the ovary are included in the endometrioid category because they resemble the tumors with those designations that are encountered most commonly in the endometrium.^{2,3} Although this tumor contains epithelial and mesenchymal elements (both of which are malignant), immunohistochemistry, and genetic studies have supported a clonal origin of both components.

The most common symptoms are pelvic or abdominal pain, abdominal distention, bowel symptoms, and weight loss. The serum concentration of CA-125 is usually elevated.

Most patients have a palpable adnexal mass and many have ascites. More than 70% of carcinosarcomas have spread beyond the ovaries at the time of diagnosis.^{1,4}

The pattern of spread, as in ovarian carcinoma, is primarily to the peritoneum, omentum, and regional lymph nodes. Treatment is by hysterectomy, bilateral salpingo-oophorectomy, and excision of as much extraovarian tumor as possible, followed by chemotherapy.³

Carcinosarcomas tend to be large, with an average diameter of 15 cm. They are either cystic and solid or entirely solid tumors. The solid portions are gray or tan, and areas of hemorrhage and necrosis are usually prominent. Microscopically, carcinosarcoma is a biphasic neoplasm with intermixed epithelial and mesenchymal elements.

CASE REPORT

A 22-year-old Hindu married female visited gynecologic outpatient department with the chief complaints of irregular, excessive vaginal bleeding since 1 year and heaviness in the lower abdomen since 6 months. She also complains headache, weakness, fatigue, abdominal pain and weight loss.

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On physical examination, a mass was palpated in right pelvic area. The mass was about 15 cm × 12 cm in size, firm, and immobile on palpation.

Per vaginal examination, reveal blood clots and a firm growth. Percussion of abdomen reveals fluid in the peritoneal cavity. On auscultation, bowel sound are normal. A provisional diagnosis of ovarian tumor was made.

Her laboratory investigation reveal - Microcytic hypochromic anemia (hemoglobin 9 g/dl), leukocytosis (13,500/ μ l). Other hematological parameter was normal. Her bleeding time, clotting time were normal. Liver and kidney functions were normal. Enzyme-linked immunosorbent assay for HIV and hepatitis B antigen was non-reactive. Urine analysis revealed no pathology. The serum CA-125 level was elevated (276 U/ml).

Pelvic ultrasound and computed tomography revealed a huge and heterogeneous pelvic mass containing solid parts with ascites. At laparotomy, the surgeon found 17 cm × 11.5 cm × 3 cm unilateral ovarian tumor. The patient underwent unilateral salpingo-oophorectomy. Gross specimen was sent for histopathological examination.

Gross

Ovarian mass measuring 17 cm × 11.5 cm × 3.5 cm in size. Outer surface is glistening white and slightly nodular. Cut surface shows homogenous greyish white solid areas along with hemorrhagic areas seen at periphery (Figure 1).

Microscopy

Section shows malignant epithelial and mesenchymal component (Figure 2). Epithelial component shows endometrioid carcinoma. Epithelial cells are arranged in diffuse as well as glandular pattern (Figure 3). Glands are well formed. In some gland secretion present. Cells



Figure 1: Cut surface shows homogenous greyish white solid areas along with cystic and hemorrhagic areas

in diffuse or sheet are round to oval pleomorphic with moderate to abundant cytoplasm. Nucleus is vesicular. In some cells, nucleoli are visible. A few mitotic figure also present. Hyaline globules are present at places. Mesenchymal component shows features of undifferentiated sarcoma (Figure 4). Spindle cells are arranged in fascicles and show minimal pleomorphism (Figures 5 and 6). At places, blood vessels hemorrhage and cysts present (Figure 7).

Histologically, the major part of the tumor consists of the carcinomatous which is adenocarcinoma with undifferentiated sarcomatous element. Hence, on the basis of clinical finding, gross and microscopic finding a diagnosis of MMMT was offered.

DISCUSSION

Microscopically, carcinosarcoma is a biphasic neoplasm with intermixed epithelial and mesenchymal elements. The epithelial component can be any type of surface epithelial carcinoma, but serous, endometrioid, and undifferentiated carcinoma are most common.⁵ The mesenchymal component is a pure sarcoma or a mixture of various types of sarcoma. Homologous sarcomatous elements include fibrosarcoma, leiomyosarcoma, and undifferentiated sarcoma. Heterologous elements, such as rhabdomyosarcoma, chondrosarcoma, or osteosarcoma, are present in a majority of ovarian carcinosarcomas.⁶ Eosinophilic hyaline globules are often present in carcinosarcomas, scattered among either the epithelial or mesenchymal cells. These globules are positive on periodic acid-Schiff staining, and a minority is immunoreactive for α 1-antitrypsin.

Singular cases of mixed mesodermal tumor have been reported to show trophoblastic or neuroectodermal differentiation or to express alpha-fetoprotein (AFP).⁷

Gross Features

These tumors are composed of soft to firm, yellow to brown, solid tissue, often exhibiting hemorrhage, necrosis, and cystic degeneration. They may be predominantly cystic. Occasionally, bone or cartilage is evident on palpation.

Microscopic Features

The epithelial component is usually of high grade and most often resembles serous or endometrioid carcinoma, but malignant mucinous, squamous, or clear-cell elements or undifferentiated carcinoma, including small-cell carcinoma of the pulmonary type, may be encountered as well.

Nonmesodermal types of tissue in rare tumors have included glial and neuronal differentiation (including

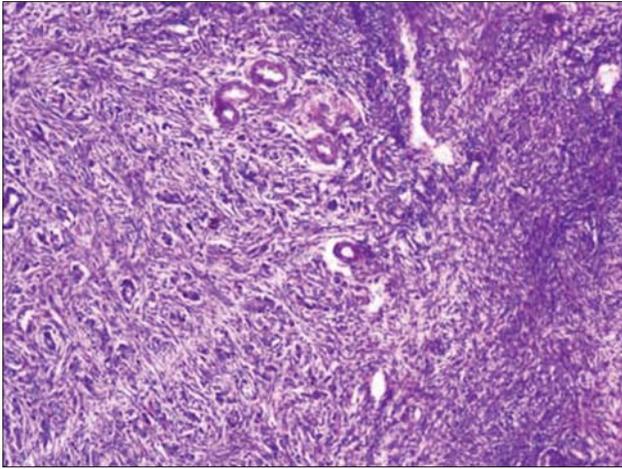


Figure 2: Epithelial (glands) and mesenchymal component (spindle cells), (H and E, 10x)

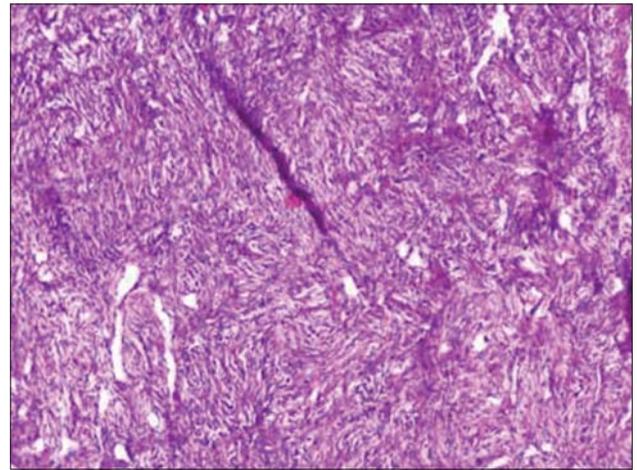


Figure 5: Undifferentiated sarcomatous component (H and E, 10x)

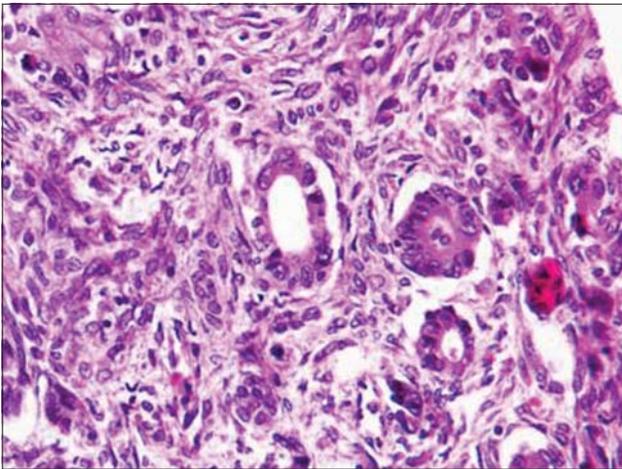


Figure 3: Well-formed gland with round to oval cells representing carcinomatous component (H and E, 40x)

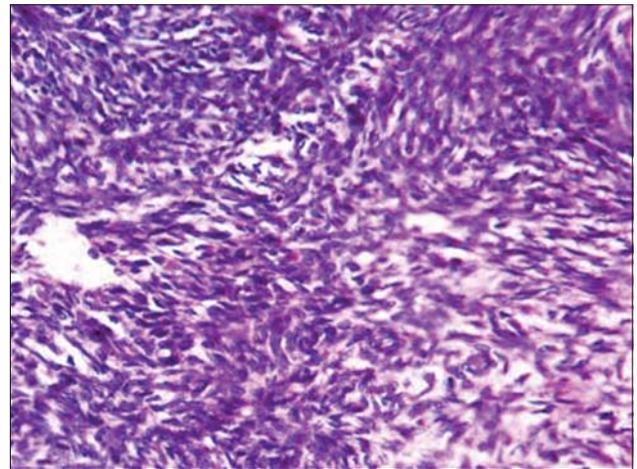


Figure 6: Spindle cells arranged in fascicles with minimum pleomorphism representing undifferentiated sarcomatous component (H and E, 40x)

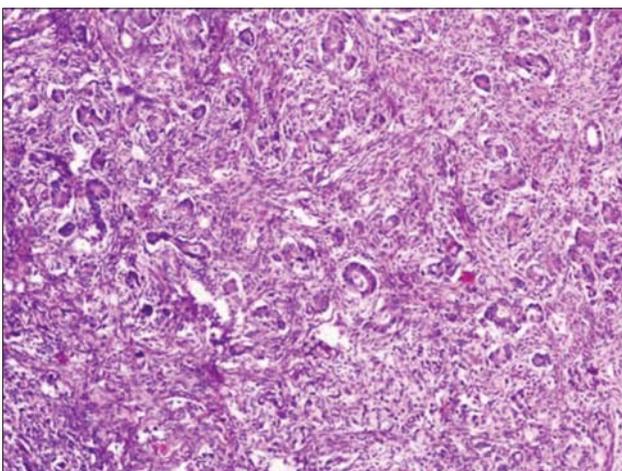


Figure 4: Intermixed epithelial and mesenchymal component. Hyaline globules are present at places (H and E, 10x)

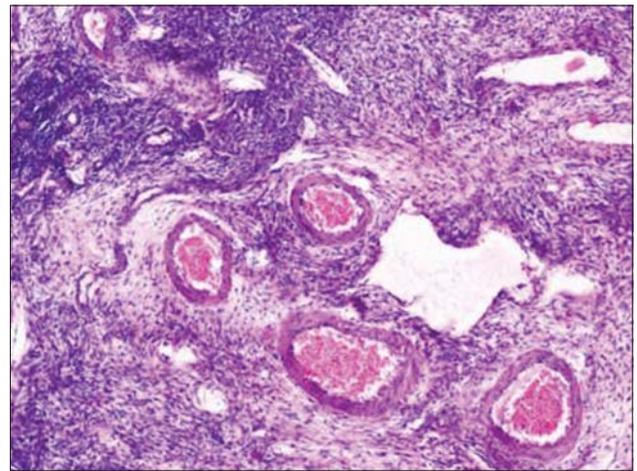


Figure 7: Blood vessels, cyst formation with tumor component (H and E, 40x)

foci resembling peripheral neuroectodermal tumor), well-differentiated hepatic-type cells, and trophoblastic

differentiation. Rare tumors have been associated with an increased serum level of AFP.

Differential Diagnosis

The neoplasm that is most often confused with the heterologous MMMT is the immature teratoma. In contrast to the former tumor, the latter is almost never found in women older than 50 years. It contains elements derived from all three germ layers, lacks a malignant müllerian-type component, and almost always contains a prominent primitive neuroectodermal component. However, as noted above, rare MMMTs contain tissue of neuroectodermal derivation (both glial and neuroepithelial), and occasional tumors contain neuroendocrine cells. Finally, the cartilage in immature teratomas has a fetal appearance, whereas in MMMTs it usually resembles the cartilage of a poorly differentiated chondrosarcoma, with marked nuclear pleomorphism.⁸

Heterologous Sertoli-Leydig cell tumors (SLCTs) with islands of cartilage or rhabdomyoblasts may cause diagnostic difficulty, but the finding of Leydig cells, sex cord formations, tubules, or gastrointestinal-type mucinous glands or cysts should facilitate the diagnosis. Finally, SLCTs often lead to virilization. In difficult cases, staining for epithelial membrane antigen (EMA) and α -inhibin and/or calretinin is useful because MMMTs are usually EMA+, whereas only occasional SLCTs are EMA+ and then in only a few Sertoli cells; α -inhibin and/or calretinin staining identifies the tumor as belonging in the Sertoli-Leydig cell category.⁸

CONCLUSION

MMMT are rare and aggressive tumor of ovary in reproductive age group. MMMT should always keep in

mind as a differential diagnosis in ovarian mass and should be differentiated from mixed germ cell tumor.

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