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RESEARCH ARTICLE

A RARE CASE OF PRIMARY OVARIAN CARCINOSARCOMA - A CASE REPORT WITH REVIEW OF LITERATURE

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ABSTRACT

Carcinosarcoma of the ovary is also known as Malignant Mixed Mullerian Tumour. It is an ovarian neoplasm with an incidence of 1-3% of all ovarian neoplasms. Usually, patients are postmenopausal women with a history of low parity. We report a rare case of 44 years old woman who presented with complaints of amenorrhea and pain in the abdomen of three months duration. Per speculum and per vaginum examination showed a mass of size 10x 8 cm in the right iliac fossa. Ultrasonogram and CT scan revealed a neoplastic mass in the right adnexa. CA-125 levels were 714U/ml. The patient underwent staging laparotomy and was found to be in stage 4a. Microscopic examination showed the presence of both carcinomatous and sarcomatous components with rhabdomyosarcomatous and chondrosarcomatous differentiation. The immunohistochemistry confirmed rhabdomyosarcomatous differentiation. The final diagnosis of carcinosarcoma or ovarian malignant mixed mullerian tumour (OMMMT) was rendered.

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INTRODUCTION

Ovarian carcinosarcoma is a rare neoplasm with an incidence of 1-3%. It is also referred as ovarian malignant mixed mullerian tumour (OMMMT) (Rashmi patnayak, 2015). Most commonly it occurs in postmenopausal women but cases have also been reported in the premenopausal age group (Uçar, 2016). It is a biphasic neoplasm with both carcinomatous and sarcomatous components. Ovarian carcinosarcoma is very aggressive tumour with poor prognosis (Torres, 2017).

CASE REPORT

A 44-year-old female P1L1 presented with chief complaints of pain in abdomen and amenorrhea since 3 months. Per speculum and per vaginum examination showed a mass of size 10x 8 cm in right iliac fossa which was mobile with firm to hard consistency. Cervix and vagina were healthy. Per rectal examination revealed a hard mass at the anterior rectal wall. Ultrasonogram showed a large heterogeneous mass of size 17x 10cm seen in the right adnexa suggestive of ovarian neoplasm. Further, a CT scan of the abdomen and pelvis revealed a hypodense lobulated solid mass measuring 10x9x10 cm in the pelvis and right adnexa, which was not seen separately from ovaries. The lesion was abutting the urinary bladder, iliac vessels, sigmoid colon, and rectal wall. The final impression on CT scan was a neoplastic mass in pelvis likely to be arising from the ovary with omental secondaries and ascitis. As the patient had ascites, cytological examination of ascitic fluid was done. In view of the papillary structure and clusters of atypical cells, the possibility of ovarian neoplasm was suggested. The CA-125 levels were significantly raised (714U/ml). Other baseline investigations were within normal limits. CBC was normal except the hemoglobin concentration which was 7.7gm%. LFT and KFT were within normal limits. Considering all investigations treating surgeons planned for staging laparotomy. Intraoperatively a mass of size 20x14x10 cm was seen arising from the pelvis. Mass was fragile and adherent to surrounding structures involving the uterus, the wall of the bladder and omentum. Overall abdominal cavity was visualized and all excised mass sent for histopathological examination.On staging laparotomy patient was found to be in stage 4a. Grossly specimem showed multiple grey white irregular nodular masses altogether measuring 15×10×5 cm.External surface was nodular and bosselated. Cut surface showed few necrotic, hemorrhagic and cystic areas. Representative bits from mass were submitted for histopathological examination.

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Figure 1. Multiple irregular nodular masses, external surface is bosselated and cut surface shows few necrotic and haemmorhagic areas

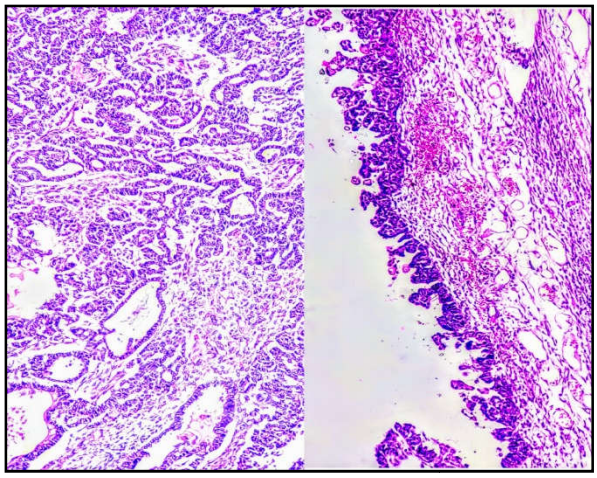

Figure 2. Carcinomatous component showing neoplastic proliferation of complex papillae and glandular structures.

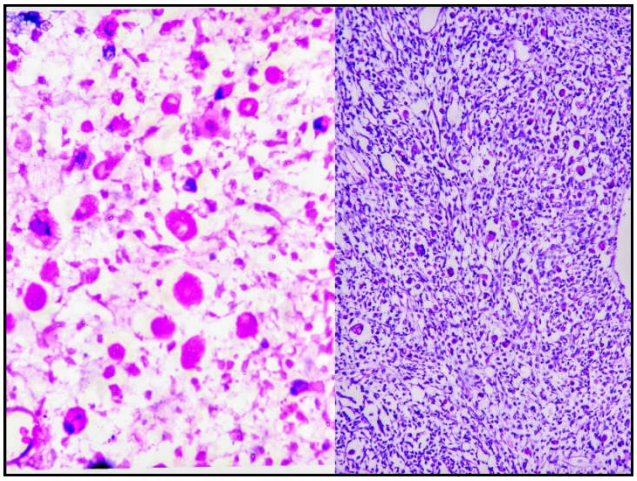

Figure 3. Highlighting extra cellular hyaline globules in stroma and Proliferation of spindle to oval pleomorphic sarcomatous cells.

Microscopically sections studied showed a tumour having biphasic pattern with an intermingling of epithelial (carcinomatous) and mesenchymal (Sarcomatous) components. Epithelial component showed neoplastic proliferation of complex papillae and glandular structures. Sarcomatous component composed of a proliferation of oval to spindle cells with pleomorphic hyperchromatic nuclei and moderate to scanty cytoplasm.

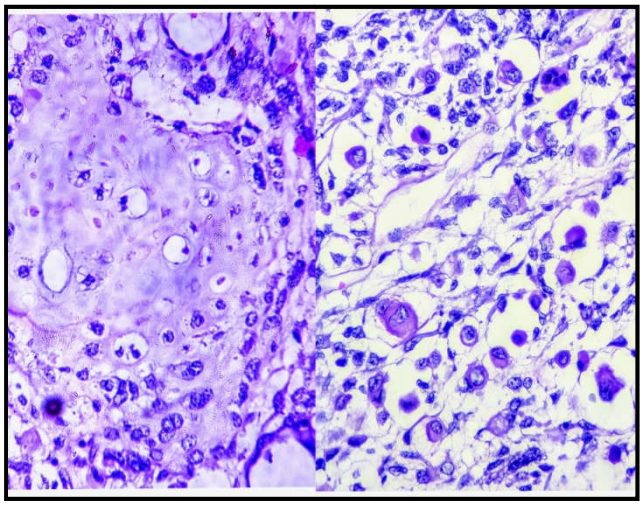

Figure 4. Chondrosarcomatous and Rhabdomyosarcomatous differentiation.

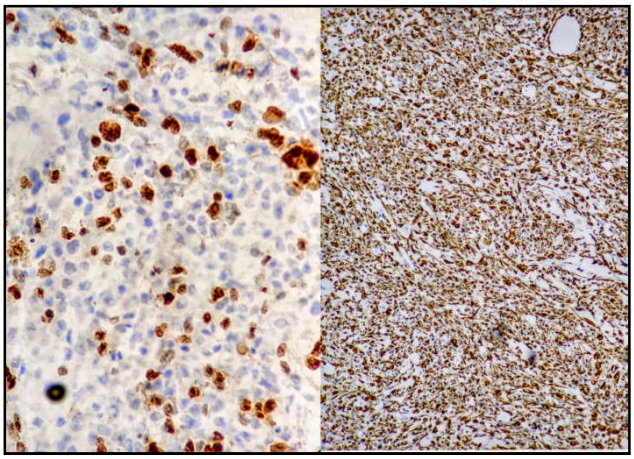

Figure 5. Focal and strong immunoreactivity for Myogenin and strong, diffuse immunoreactivity for vimentin.

The foci of cells showing rhabdomyosarcomatous differentiation in the form of large cells with eosinophilic cytoplasm were seen. The sarcomatous component also showed an element of chondroid differentiation. Numerous extracellular and intracellular hyaline globules along with areas of hemorrhagic necrosis were also seen. Immunohistochemistry showed diffuse and strong immunoreactivity for vimentin, focal and strong immunoreactivity for myogenin. Finally the diagnosis of ovarian malignant mixed mullerian tumour (Carcinosarcoma) with rhabdomyosarcomatous and chondrosarcomatous differentiation was offered.

DISCUSSION

Carcinosarcoma of the ovary is a rare neoplasm. The term Carcinosarcoma was first used by Virchow in 1864 [4]. It is also known as Ovarian Malignant Mixed Mullerian Tumour(OMMMT).OMMMT most commonly occurs in uterus followed by fallopian tubes, ovaries, and vagina. The incidence of carcinosarcoma of the ovary is 1-3 % of all ovarian neoplasms (Rashmi patnayak, 2015). Usually, the patients are postmenopausal women having low parity but cases have also been reported in the premenopausal age group (Uçar, 2016). The median age at the time of diagnosis by various series ranged from 51 to 56 years (Zheng, 2017).

Clinical features are ascites, abdominal discomfort or pain and a lump in the abdomen. The present case had ascites, abdominal pain and amenorrhea. Diagnosis of OMMMT Or Carcinosarcoma is quite challenging as the clinical, radiological features and raised CA 125 levels simulate other surface epithelial tumors (Zheng, 2017; Siddharth Tewari, 2017; Maheshwari, 2013). The present case had similar features, hence OMMMT was the last consideration. Preoperative diagnosis is essential as this tumour has a worse prognosis than other surface epithelial tumours. Many studies have proposed the following risk factors in the development of Carcinosarcoma such as obesity, nulliparity, exogenous estrogen and long term use of tamoxifen (Uçar, 2016).

Carcinosarcoma of the ovary is a biphasic malignant neoplasm. Many theories have been proposed for the evolution of Carcinosarcoma 1) collision theory which states that both components occur independently from two different stem cells.2) combination theory says that a single stem cell differentiates into an epithelial and sarcomatous component (Siddharth, 2017). Recent studies suggest the monoclonal theory of histogenesis which explains the transformation of the epithelial component into a sarcomatous component akin to metaplastic carcinoma. Courtney Fox in his article quoted that during morphogenesis of embryo transdifferentiation of epithelial to mesenchymal components ensue. A similar phenomenon reemerges in tissue neoplasm. But the difference in morphogenesis and tumourogenesis is that the various cytokines and growth factors which regulate the morphogenesis are dysregulated in malignancy and it's progression (Fox, 2019).

Pathological examination:--A single epithelial component or a mixture of many components may be seen. Santosh Menon *et al* in his series of 27 cases reported two or more epithelial components in 10 cases. The commonest epithelial components reported by various authors are endometroid adenocarcinoma followed by serous adenocarcinoma (Maheshwari *et al*., 2013). Other epithelial components like squamous cell carcinoma, clear cell carcinoma, undifferentiated carcinoma may be seen (Daimon, 2019; Dasgupta Senjuti, 2015; Kothari, 2015). Heterologous sarcomatous components are commoner than homologous components. Of which rhabdomyosarcomatous differentiation is frequently seen (Maheshwari, 2013). Other heterologous components seen are chondrosarcoma, osteosarcoma, angiosarcoma (Dasgupta Senjuti, 2015). The teratoid OMMMT is extremely rare. They are so labelled when foci of germ cell tumours or neuroectodermal elements as seen in immature teratoma are identified (Maheshwari, 2013). The present case had serous adenocarcinoma as an epithelial component. The tumour had sarcomatous predominance over carcinomatous elements and showed rhabdomyosarcomatous and chondrosarcomatous heterologous elements. Tumour showed diffuse strong positivity and focal strong positivity for Vimentin and Myogenin respectively. The extracellular and few intracellular hyaline globules were also seen in the rhabdomyosarcomatous area. Santosh Menon in his study of 27 cases reported hyaline globules in 15 cases. This may be reactive to alpha 1antitrypsin.In a biopsy, it forms a clue to search for OMMMT (Maheshwari, 2013).

The prognosis of this tumour is very poor. The prognosis is multifactorial. It depends upon the stage of the disease at the time of diagnosis, necrosis, mitotic count, type of sarcomatous component, homologous or heterologous, type of sarcoma, evidence of sarcomatous component outside the ovary and residual disease after primary surgery. Studies done for individual parameters as prognostic indicators are negligible (Zheng, 2017; Maheshwari, 2013). Most of the studies are in agreement that the stage of the disease at the time of diagnosis is an important prognostic factor. The presence of a sarcomatous component outside the ovary indicates the worst prognosis (Maheshwari, 2013). Quantum of residual disease after cytoreduction surgery has a great impact on patient survival. Suboptimal or non-optimal cytoreduction surgery has poor disease free survival (Makris, 2015; Zheng *et al*., 2017).The three-year survival rate with optimal cytoreduction surgery is 64.8% as against 33.3%with nonoptimal surgery[5].So cytoreduction surgery should be aimed at achieving macroscopic disease-free peritoneal cavity. The present case was subjected to staging laparotomy which made it sure that the tumour is inoperable. However, the debulking of the tumour was done and resected specimen was subjected for histopathological examination. The postoperative period was uneventful. The patient was advised complementary adjuvant chemotherapy but she lost the follow-up. Exact etiological factors and carcinogenesis of this tumour are not known. According to the monoclonal theory where epithelial to mesenchymal transdifferentiation generates various sarcomatous heterologous elements in the tumour. In the process, various pathways are involved, which are difficult to target and eradicate the cancer cells. So the effective chemotherapeutic regime is yet to be reached out. More prospective studies are needed to find out the effective drug treatment for improved survival of the patient.

Conclusion

It is an extremely rare tumour with a dismal prognosis. Preoperative workup for early diagnosis and optimal cytoreduction surgery with recommended complementary adjuvant chemotherapy is of great value in improving disease-free survival. However such cases need to be registered to aid in research for developing effective drug treatment. The present case study had limitations as the patient failed to register for follow up.

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