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Case Report

Primary retroperitoneal malignant mixed Mullerian tumour: A rare case report

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ABSTRACT

Malignant mixed Mullerian tumour commonly known as carcinosarcoma is a rare and aggressive biphasic tumour with epithelial and mesenchymal components. In this reported case, a 68 year old post menopausal, nulliparous female, presented with complaints of difficulty in breathing and abdominal discomfort. Clinical evaluation and radiological findings were consistent with malignancy of left ovarian origin. Patient was taken up for total abdominal hysterectomy with bilateral salpingo-oophorectomy. But intraoperatively, the tumour was found as a single large retroperitoneal mass with no connection to ovaries or other pelvic organs. Histopathological examination of the specimen revealed features of malignant mixed Mullerian tumour. Immunohistochemical markers showed positivity for CK 20, Pan CK and EMA in carcinomatous areas and vimentin in sarcomatous areas. ER and PR were found to be negative. Patient was on regular follow-up and responded well to chemo and radiotherapy. Primaries in retroperitoneum also have poor prognosis similar to carcinosarcomas of female genital tract.

1. Introduction

Malignant mixed Mullerian tumour(MMMT) is also known as carcinosarcoma. Being a rare and aggressive biphasic tumour composed of epithelial and mesenchymal components, MMMTs account for <1% of female genital malignancies and are known to occur in postmenopausal women.[1,2]Extragenital carcinosarcomas are even rarer. Only 29 cases have been reported in literature so far to the best of our knowledge.[3] Usual extragenital locations include pelvic wall, peritoneum, omentum, ureters and kidneys.[4]

Primaryretroperitoneal carcinosarcomas are extremely rare which makes this case novel.

2. Case History

A 68 year old post menopausal nulliparous female presented with complaints of breathlessness and pressure symptoms of three months duration. Per – abdominal examination showed a large ill-defined, non- tender pelvic – abdominal mass. Per – speculum and bimanual examination were within normal limits. Routine blood investigations and cervical smear were normal. Ultrasound of abdomen showed a complex lesion probably arising from left ovary suggestive of malignant ovarian lesion. Bilaterals kidneys and adrenals were normal. FNAC was not performed. Following this contrast enhanced CT abdomen was done which showed a large heterogenous lesion with predominantly cystic and solid components likely to represent malignant left ovarian mass (Figure 1). However, renal function tests and CA-125 were within normal limits (18.1 U/ml).

Patient was taken up for total abdominal hysterectomy with simultaneous bilateral salpingo-oophorectomy. But

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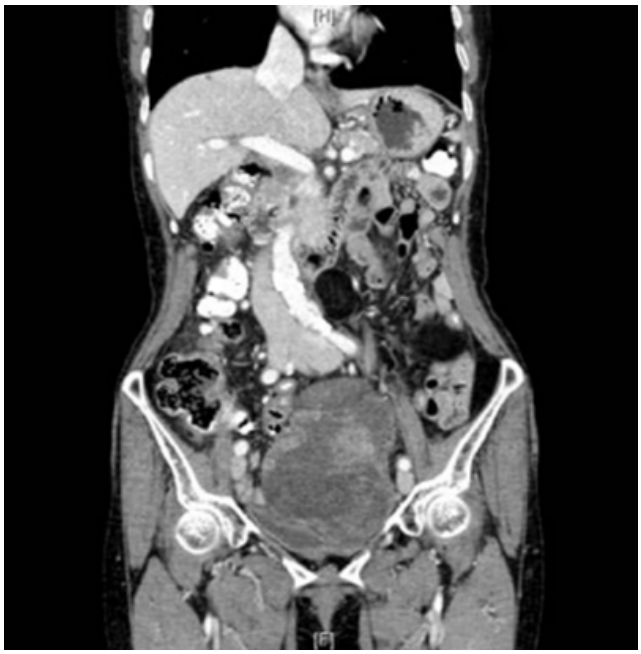

Fig. 1: Coronal reformatted image of contrast enhanced CT scan

per-operatively, uterus and bilateral ovaries were normal. They found a large retroperitoneal mass which had no connection with the pelvis. The mass was removed and sent for histopathological examination.

Macroscopically it was a single grey- white to grey-brown soft tissue mass measuring 30x18x9cm (Figure 2) which was partly encapsulated, lobulated and congested. Cut surface of the tumour showed predominantly solid with partly cystic areas with adjacent areas showing extensive necrosis and haemorrhage. Microscopic examination revealed tumour showing both epithelial and sarcomatous elements (Figures 3 and 4).

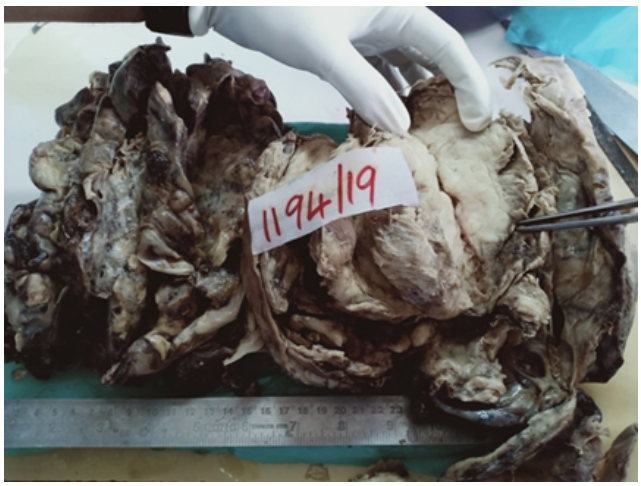

Fig. 2: Gross picture showing tumour which is partly encapsulated, with areas of necrosis and hemorrhage

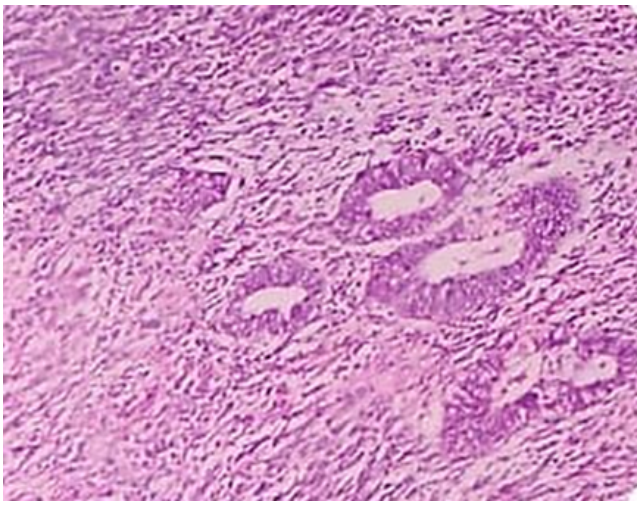

Fig. 3: Microscopy showing epithelial glandular and stromalspindle cell components, 10x

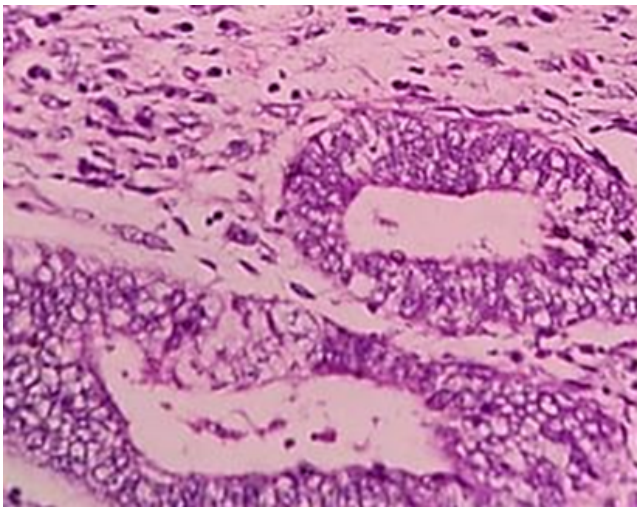

Fig. 4: Microscopy showing malignant glandular component withnuclear atypia, 40x

Epithelial elements were composed of papillary and insular patterns whereas sarcomatous elements showed spindle cell component with brisk atypical mitosis. Also, there were extensive areas of necrosis and haemorrhage. Immunohistochemical markers were done which showed positivity for Cytokeratin 20, Pan cytokeratin, Epithelial Membrane Antigen in carcinomatous areas. (Figure 5) and positivity for vimentin in sarcomatous areas(Figure 6). ER and PR were found to be negative.

These morphological and immunohistochemical features lead to the diagnosis of malignant mixed Mullerian tumour. Post operative period was uneventful. Patient was given a cycle of whole abdominal radiation by external beam at a dose of 45-50. 4Gy in 25-28 fractions for five days in a week over five and a half weeks in combination with carboplatin

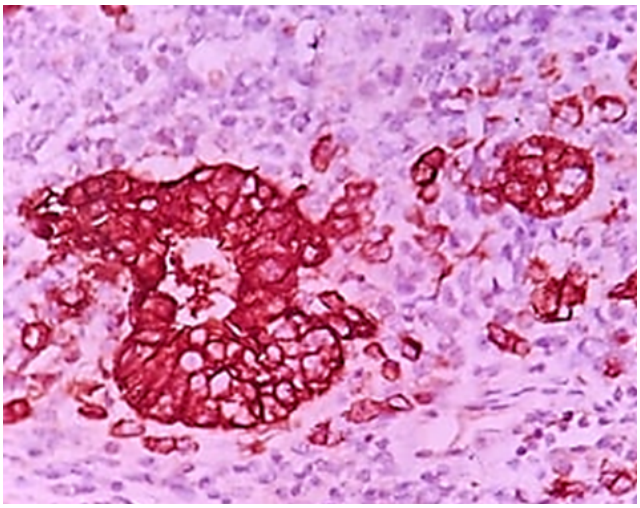

Fig. 5: Epithelial component showing strong cytoplasmic positivity for cytokeratin in malignant glandular area

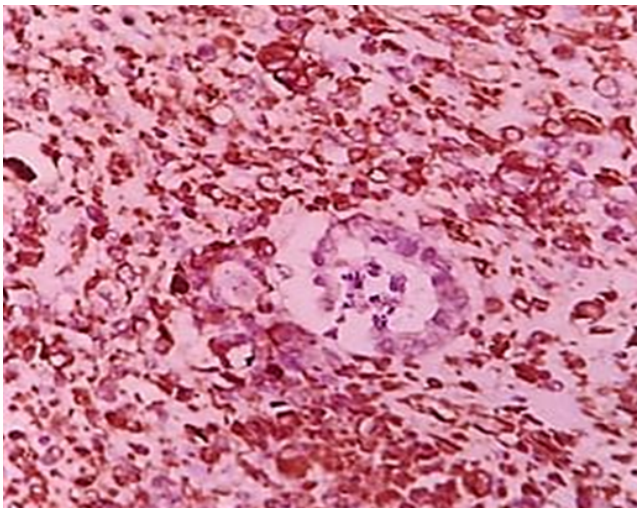

Fig. 6: Sarcomatous element showing strong cytoplasmic positivityfor vimentin and negative staining in glandular area

(AUC 5) and ifosfamide (3g/m^2.) every three weekly for 6cycles. Last follow up was done six months back and was found to be responding well to chemo and radiotherapy.

3. Discussion

Carcinosarcoma usually arise in tissues of the female genital tract and rarely occurs in retroperitoneal location. Our scenario is of a retroperitoneal mass presenting as carcinosarcoma. Common risk factors for developing carcinomasarcoma includes history of pelvic irradiation, obesity, nulliparity, oestrogen exposure, endometriosis and synchronous or metachronous gynaecologic or non gynaecologic malignancies.[5,6]In the present case, even though patient had few of the known associated risk factors, uterus, ovaries and other adnexal areas were free of tumour even after meticulous examination. Due to the anatomical complexity of the retroperitoneal space proper diagnostic strategy using clinical, radiological and histopathological examination has to be followed to reach a proper diagnosis.[7]Three main theories have been proposed to explain the histogenesis of MMMT : a) Collision theory – which states that carcinomas and sarcomas are independent neoplasms. b) Combination theory – which suggests that the two components originate from a single stem cell which then later on undergoes differentiation. c) Conversion theory – according to which the sarcomatous component is believed to originate from the carcinoma component as the tumour evolves.[8]The biphasic nature of the tumour is explained based on origin from “secondary Mullerian system” or from residual hormone independent ducts present in the retroperitoneum.[9,10]According to a study done by Sparks D[3] et al these tumours are believed to arise from an endometriotic foci. Another mechanism which they suggested is the possibility of secondary Mullerian system which states that retroperitoneal mesenchyme has a potential for Mullerian differentiation which they attribute to the close embryonic relationship to the Mullerian ducts.[3] Recent studies suggest monoclonal origin of these tumours which is explained by conversion and combination theories stated above. In the present case no endemetriotic focus was identified suggesting the possibility of secondary Mullerian system origin. Immunohistochemical panel done showed results as expected for a carcinosarcoma.[11]Closest differentials includes mesothelioma, retroperitoneal soft tissue sarcomas, subserosal leiomyomas, malignancies of kidney, adrenal glands and germ cell tumours.[12] The main mode of treatment is surgery followed by chemo and radiotherapy. Role of radiotherapy still remains uncertain. Combination chemotherapy is found to have better role than single drug regimen out of which cisplastin and ifosfamide are found to have better effect.[13]Carcinosarcomas are prone to recur locally. Local recurrence is the main cause of death unlike metastasis.[13] According to a study done by Rajshekar S K et al concurrent chemo and radiotherapy is shown to have less local recurrence and thereby better survival.[14]Similar to carcinosarcomas of female genital tract, primaries of retroperitoneum also have poor prognosis.[3]

4. Conclusion

Primary retroperitoneal carcinosarcoma can occur in postmenopausal age group. It is a rare and aggressive tumour with poor prognosis. Eventhough extremely rare, it has to be considered in the differential diagnosis of retroperitoneal mass in postmenopausal female.

5. Source of funding

None.

6. Conflict of interest

None.

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