CASE REPORT

Bilateral ovarian malignant mixed Mullerian tumour: a rare entity

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SUMMARY

A 52-year-old postmenopausal woman presented with abdominal distension for 2 months. She had no vaginal bleeding and no relevant medical history. A physical examination showed a large palpable pelvic mass. A CT scan showed an ill-defined, heterogeneously enhancing, soft tissue attenuation lesion in the lower abdomen and pelvis involving the right adnexal region with massive ascites, and the left ovary did not reveal any evidence of disease. The omentum was thickened and heterogeneous. The patient underwent a total hysterectomy with infracolic omentectomy. The diagnosis was bilateral ovarian malignant mixed Mullerian tumour (MMMT) with omental deposits. Aggressive treatment includes surgery and chemotherapy. Women with this tumour have a significant increased risk of death compared to women with epithelial ovarian cancer and very poor prognosis. We report a case of MMMT of the ovary with clinicopathological correlation and diagnostic difficulties as malignant epithelial tumour resemble ovarian stromal sarcoma in postmenopausal female.

BACKGROUND

Malignant mixed Mullerian tumour (MMMT) of ovary is a rare biphasic malignant neoplasm. The tumour is commonly seen in uterine corpus followed by the cervix. MMMT of the ovary is extremely rare and accounts for less than 1% of all ovarian tumours. We have presented a case because of its rarity and unusual presentation.

CASE PRESENTATION

A 52-year-old postmenopausal woman presented with abdominal distension, pain, decreased appetite and was started to lose her weight about 2 months prior to presentation. She had no relevant medical history, irradiation and previous surgery for any other tumour, chemotherapy and exogenous oestrogen. Gynaecological examination revealed no vaginal bleeding. Physical examination showed a palpable large pelvic mass. The patient underwent a total hysterectomy with infracolic omentectomy.

We received an entire uterus with cervix, left ovary and right ovary in multiple fragmented pieces and omentum. The uterus with both fallopian tubes and a cervix was grossly normal. The endometrial cavity showed an attached fibroid measuring 2×1 cm in size. The left ovary measured 3×2.5×1 cm, cut surface showed a dirty white area with necrosis (figure 1A). The right ovary was collectively 13×10×6 cm in size. The cut surface showed a homogeneous solid grey white to grey brown areas with necrosis (figure 1B). Omentum

showed fibrofatty tissue with solid white areas (figure 1C). The tissue was routinely processed, with the sections being cut and stained with H&E.

On microscopic examination, the uterus showed leiomyoma and the cervix showed chronic cervicitis. Sections from the right ovary showed a biphasic malignant neoplasm intermixed with epithelial (carcinomatous) and mesenchymal (sarcomatous) components (figure 2A). Carcinomatous components showed glands, sheets, and nest and complex papillae with fibrovascular core. Individual tumour cells were pleomorphic, high nuclear-cytoplasmic ratio, round to oval vesicular nuclei with prominent nucleoli and scant-to-moderate cytoplasm (figure 2C). The sarcomatous component showed storiform and fascicular pattern. Individual tumour cells were pleomorphic, spindle-shaped, polygonal to multinucleated cells, high nuclear-cytoplasmic ratio with scant cytoplasm and 3–5 mitotic figures per high power field (figure 2D). Large areas of haemorrhage and necrosis were also seen. Carcinomatous differentiation was also noted in the sarcomatous component of the left ovary (figure 2B). The section from the omentum showed a similar morphology of tumour cells.

An immunohistochemical examination was conducted. The cytokeratin was positive in carcinomatous element and vimentin in sarcomatous elements (figures 3A, B). Desmin, oestrogen and progesterone were negative. The final diagnosis was bilateral ovarian MMMT with omental deposits.

INVESTIGATION

The serum CA-125 level (576 U/mL) was elevated. An ultrasonography and a CT scan showed an ill-defined, heterogeneously enhancing, soft tissue attenuation lesion measuring 15×10×6 cm in size, seen in the lower abdomen and pelvis involving the right adnexal region with massive ascites, and the left ovary did not reveal any evidence of disease. The omentum was thickened and heterogeneous.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis includes MMMT, poorly differentiated adenocarcinoma and stromal sarcoma.

TREATMENT

The patient underwent a total abdominal hysterectomy with infracolic omentectomy. Presently she is receiving chemotherapy.

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Figure 1 (A) Gross specimen showing uterus with leiomyoma and cervix along with the left ovary (arrow head). (B) Right ovary with variegated appearance showing solid grey white to grey brown areas with necrotic areas. (C) Omentum with multiple solid white areas (arrow head).

OUTCOME AND FOLLOW-UP

After surgery, the patient was responding well with chemotherapy. There was no recurrence in the 6-month follow-up.

DISCUSSION

Carcinosarcoma is an MMMT originating from the mesenchyme of the embryonic mesoderm. The disease is manifested mostly

Figure 2 (A) Right side ovary showing biphasic malignant tumour with carcinomatous and sarcomatous components (×100, H&E stain). (B) Left side ovary showing glandular differentiation in the sarcomatous component (×100, H&E stain). (C) Carcinomatous component with pleomorphic cells, arranged in glandular pattern (×400, H&E stain). (D) Sarcomatous component showing pleomorphic spindle cells arranged in sheets, interlacing fascicles with high mitosis (×400, H&E stain).

Figure 3 (A) IHC cytokeratin positive in carcinomatous elements, $\times 400$. (B) IHC vimentin positive in sarcomatous elements, $\times 400$.

with per vaginal bleeding and abdominal mass. In our case no vaginal bleeding was present. The risk factors for MMMT include obesity, nulliparity, exogenous oestrogen and long-term tamoxifen use.¹ A rare exception aside this is a lesion of postmenopausal women.² Ovarian carcinosarcoma, also called MMMT, is a rare variant of ovarian cancer, accounting for less than 1% of all ovarian tumours, with fewer than 400 cases reported in the literature.³ MMMT is a biphasic malignant neoplasm intermixed with epithelial and mesenchymal components. The epithelial components in carcinoma are mucinous, serous, squamous, endometrioid, clear cell or mixture of these types and sarcomatous components, which may be either homogeneous (composed of malignant stromal elements native to the ovary) where as heterogeneous (composed of sarcomatous tissue not normally found in the ovary such as bone, cartilage, skeletal muscle and adipose tissue). They are aggressive and have a poor overall survival rate.⁴ Research in MMMTs of the uterus have suggested that the sarcomatous and carcinomatous components both arise from a single malignant epithelial precursor which has undergone metaplastic change to a sarcomatous form in some areas of the malignant tissue which contributed to the presence of both histological types.⁵ MMMT are usually diagnosed at an advanced stage of disease and survival after diagnosis varies by stage of disease. Despite aggressive treatment that includes surgery and chemotherapy, women with this tumour have a significant increased risk of death compared to women with epithelial ovarian cancer and very poor prognosis.⁶ According to Singh,⁷ owing to the common occurrence of mixed tumour in the female genital tract, these need careful evaluation, as tumours of apparently different histologies may still represent metastases with a minor component being missed at the primary site.

Contributors MK and MMG diagnosed the patient and wrote the manuscript. VD was the operating surgeon.

Competing interests None.

Patient consent Obtained.

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Learning points

- It is important to be aware of the unusual occurrence and to differentiate it from primary malignancy.
- Mostly the tumours are seen in postmenopausal women and have a poor prognosis. Improved survival times would probably be obtained if accurate diagnosis and aggressive treatment are applied early.
- Immunohistochemistry provides confident and confirmatory diagnosis.

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