

Subtotal vaginal Hysterectomy for a case of Uterine Carcinosarcoma complicated by uterine inversion

Afshan Azhar,¹ Magid Gab,² Abdelmagid Gaboura,³ Ciara Nolan⁴ and Kevin Hickey⁵

1. Registrar of Ob. Gyn., University Maternity Hospital Limerick, Ennis Road, Limerick, Ireland. 2. Senior House-officer of Ob. Gyn., University Maternity Hospital Limerick, Ennis Road, Limerick, Ireland. 3. SpR of Ob. Gyn., University Hospital Waterford, Dunmore Road, Waterford, Ireland. 4. Consultant of Ob. Gyn., University Hospital Limerick, St. Nesson's Road, Limerick, Ireland.

Abstract :

Background :

Non puerperal Uterine inversion caused by a uterine carcinosarcoma is a very rare condition. Endometrial carcinoma is the 6th most commonly occurring cancer in women and carcinosarcoma accounts for only 4.3%.¹ However it accounts for 15% of the mortality rate caused by uterine malignancies.²

Case Report :

We present a rare case of uterine carcinosarcoma with uterine inversion in 91-year women.

Discussion- Non puerperal uterine inversion with Uterine carcinosarcoma is not only a diagnostic but also surgical challenge.

Keywords :

Uterine inversion, uterine carcinosarcoma (UCS), radiotherapy, chemotherapy.

Case Report :

91-year-old woman was admitted acutely with heavy vaginal bleeding and a palpable abdominal mass. She was transfused with 2 units of RCC in view of her low haemoglobin of 7 g/dl. Pelvic Ultrasound showed uterine enlargement and heterogeneous lesion measuring (3.7 x 4) cm in the body of the uterus. Ultrasound was inconclusive and dynamic MRI with contrast showed an inverted uterus sitting in the vagina with a likely neoplastic lesion measuring (3.6 x 4.7 x 1.6) cm and no pelvic lymphadenopathy. Clinically, a mass was extending to the vaginal introitus. Her case was reviewed at the oncology multi-disciplinary meeting of the University Hospital Limerick. The decision was made to perform examination under anaesthesia. A biopsy was taken from the mass and Colpocleisis procedure was performed, with a plan for palliative radiotherapy. Histopathology was consistent with uterine carcinosarcoma.

Given her advanced age, it was decided to administer palliative radiotherapy to shrink the size of the mass and make it less vascular. Patient tolerated the treatment well, and few weeks later she was readmitted with the fundal wall of the uterus outside the vagina as Colpocleisis did not work. Subtotal vaginal hysterectomy was performed leaving back the cervix. The histopathological examination of the specimen showed the uterus with polypoidal exophytic tumour measuring (65 x 50 x 15) mm in size and a separate endometrial polyp (15 x 10 x 10) mm. The histology was similar to the biopsy specimen consistent with carcinosarcoma. Tumour was confined to the endometrium and no involvement of the myometrium (FIGO Stage 1A). Patient was followed up in clinic at four and eight weeks after surgery.

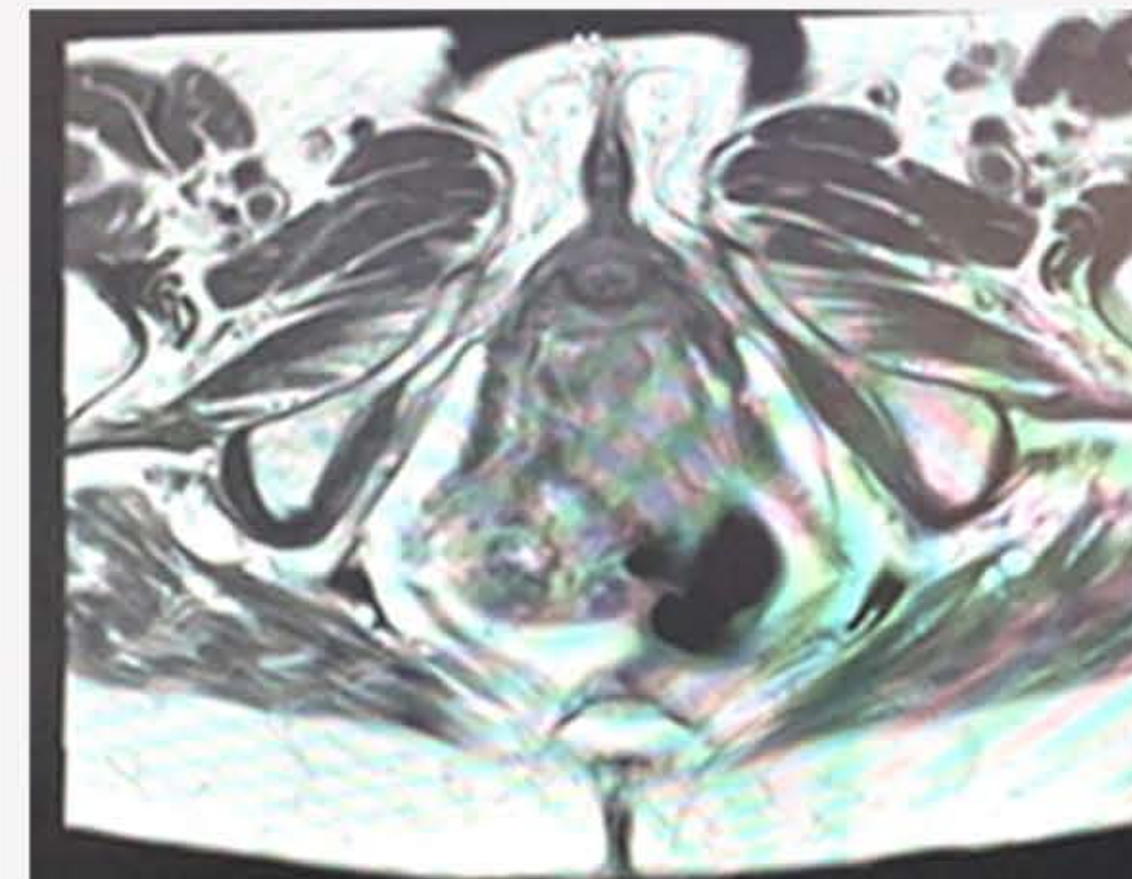


Figure (1)

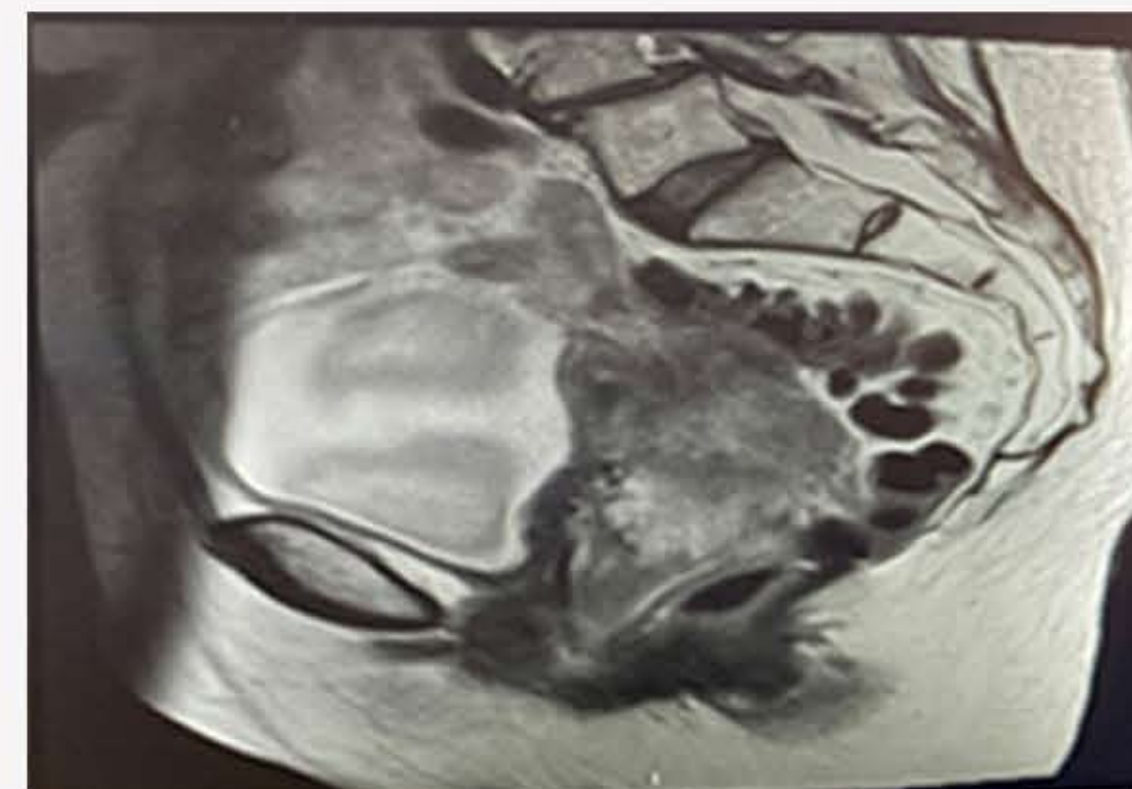


Figure (2)

Figure (1) Shows Axial and Figure (2) shows sagittal views of inverted uterus with apparent abnormal appearing tumour presenting in the vagina with (3.6 x 4.7 x 1.6) cm likely neoplastic lesion.

Discussion :

Benign masses like submucosal leiomyomas account for most of non-*puerperal* uterine inversion and very rarely malignant neoplasm are the cause. Being a very rare condition, it needs a high degree of clinical suspicion.⁷ Its unique features on Ultrasonography and MRI aids prompt diagnosis.⁷ Carcinosarcoma was previously called as Malignant mixed Mullerian tumors or Malignant mixed Mesodermal tumors as they comprise both epithelial and mesenchymal elements. The results of recent research in genetic and molecular levels concluded that carcinosarcoma are monoclonal in origin. ⁸⁻¹¹ The primary reason of its malignant potential is due to its carcinomatous component rather than its sarcomatous element.

¹¹ Recently, they are reclassified as part of differentiated malignant endometrial carcinoma instead of sarcoma.¹² According to FIGO, carcinosarcomas are graded as high-grade tumors. Multiple studies have proven Uterine Carcinosarcoma to be an aggressive tumor.¹³⁻¹⁶ It has been suggested that recurrence is high even if the disease is confined to the uterus.¹⁶ Uterine Carcinosarcomas are more common in African American and Black races.¹⁷ They have more risk of developing an advanced disease at initial presentation with bad prognosis.¹⁷ Risk factors associated were hypoestrogenism, obesity, nulliparity, tamoxifen, prior pelvic radiation. Association of smoking and tumour aggressiveness is also been reported.¹⁸⁻²⁰

The clinical presentation of Uterine Carcinosarcoma can be a triad of abnormal uterine bleeding, abdominal pain, and a rapid enlargement of uterus.²¹ However, in 40-60% of cases the first presentation can be in the form of extra uterine disease. Non *puerperal* uterine inversions resulting from UCS is extremely rare. A high degree of clinical suspicion and prompt diagnosis is critical in this rare scenario.

On ultrasound, Uterine Carcinosarcomas present as hyperechoic masses compared to endometrium with expansion of endometrial cavity.²¹ MRI plays an important role in pre-operative assessment of tumour localization and staging. Features such as intense contrast enhancement, irregular or nodular margins, necrosis, rapid growth, and restriction at diffusion weighted imaging aids in diagnosing of carcinoma. ²² MRI and fluorodeoxyglucose PET-CT scans help in detection of lymph node metastasis prior to surgery. Metastasis to distant organs such as lungs, peritoneum, bones, liver, and central nervous system has been reported in 49%, 44%, 17%, 15% and 8%, respectively.²³ Treatment options include surgery, radiotherapy, and chemotherapy according to its staging. Surgical staging of Uterine Carcinosarcomas include hysterectomy, with bilateral Salpingo-Oophrectomy, pelvic/para-aortic lymphadenectomy and cytoreduction of the neoplasm. Patients with Carcinosarcoma stage 1A have a better prognosis. Brachytherapy alone can be considered for patients with no lympho-vascular invasion.

For patients with stages more than 1B chemotherapy and external beam radiotherapy +/- vaginal vault brachytherapy should be considered. In patients with stage 3 and 4 disease, treatment decisions are based on the tumour extent, the patient's general condition and the patients' wishes.²⁴ However, there is no firm evidence to guide the treatment protocols. Most of the Gynaecological oncologists aim for optimal cytoreduction for improved survival. There are no proper prospective trials regarding optimal chemotherapy regimen. Paclitaxel and carboplatin showed around 54% overall response rate in phase 2 trials.²⁴ Studies of phase 3 Randomized trial of Paclitaxel and carboplatin versus Ifosfamide and carboplatin were done, but results are still awaited.

Results of a SEER study of 1819 women proved 21% reduction in mortality rate in women with stage 1-2 disease with pelvic radiotherapy.²⁵ Multimodal mode of treatment depending on surgical-pathological staging helps in improving the survival rate in patients with Uterine Carcinosarcoma.

Conclusion :

Uterine Carcinosarcoma cases presenting as non-*puerperal* uterine inversion is very rare. Timely detection is extremely crucial. MRI plays an important role as a diagnostic tool. Multimodal approach with surgery, chemotherapy, and radiotherapy aid to decrease the mortality and morbidity associated with Uterine Carcinosarcoma.

Citation :

Gaboura A., Hickey K. et al,

Corresponding Author :

Mr. Kevin Hickey, MRCPI MRCOG, Consultant Gynaecological Oncologist, University Hospital Limerick, St. Nesson's Road, Limerick, Ireland.

Email: kpwhickey@gmail.com

Introduction :

Uterine inversion is described as invagination of the fundus of the uterus through the vagina. Uterine carcinosarcoma (UCS) is a rare and extremely aggressive tumor. Carcinosarcoma accounts for only 5-6% of all uterine malignancies.³⁻⁵ In nearly 60% of cases the first presentation is in the form of extrauterine disease. Even with surgical and adjuvant therapy the recurrence rate of carcinosarcomas is as high as 50%.⁵ The primary surgery plays a critical role in treatment of Uterine Carcinosarcoma. The prognosis of uterine carcinosarcoma is associated with surgical-pathological stage and treatment modalities, same as all cancers.⁶