

Non-Puerperal Uterine Inversion Caused by Embryonal Rhabdomyosarcoma in an Adolescent: A Case Report

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Abstract

Case Report

Introduction: Non-puerperal uterine inversion is an uncommon gynecologic emergency that can be triggered by either malignant or, more commonly, benign tumors. Uterine sarcoma, including the rare subtype known as embryonal rhabdomyosarcoma, is among the malignant causes. The management of this condition poses a significant challenge for surgeons. **Case Report:** A 15-year-old nulliparous and virgin female arrived at the emergency department in hemorrhagic hypovolemic shock, experiencing pelvic discomfort and expelling endometrial tissue through the vagina. She underwent a total abdominal hysterectomy with bilateral oophorectomy, a pathological examination revealed that the uterine inversion was caused by embryonal rhabdomyosarcoma. Subsequently, the patient was referred to an oncologist for chemotherapy. **Conclusion:** Rapid perioperative diagnosis can be achieved through physical examination and magnetic resonance imaging (MRI), with histopathological examination serving as the definitive diagnostic confirmation tool.

Keywords: uterine inversion, gynecologic emergency, tumors, nulliparous, chemotherapy.

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INTRODUCTION

Uterine inversion, a rare occurrence, can be classified into puerperal, obstetric, non-puerperal, or gynecologic inversion. This condition involves the fundus of the uterus turning inside out and potentially being expelled through the vaginal introitus [1]. Non-puerperal inversion, which is extremely uncommon, may be caused by uterine corpus neoplasms. The physiopathology involves the tumor within the uterine cavity leading to a thin uterine wall or an enlarged myometrium, causing irritability and initiating expulsive contractions. These contractions can dilate the cervix and aid in expelling the tumor. Factors such as the tumor's weight, manual traction on the tumor, or increased intra-abdominal pressure from activities like coughing, straining, and sneezing can also contribute to non-puerperal uterine inversion [2].

CASE PRESENTATION

A 15-year-old nulliparous woman has been admitted in Obstetric emergency department.

The patient, who had undergone an exploratory laparotomy few hours prior to her admission, due to an

acute hemorrhagic hypovolemic shock associated to a pelvic discomfort and discharge of endometrial tissue through the vagina.

A rectal examination revealed a movable and painless vaginal mass.

A magnetic resonance imaging (MRI) of the pelvis showed a Uterus presenting an inverted glove-finger appearance, prolapsed into the vagina and limited on either side through the fornixes and the vaginal walls. It measures 7 cm in height and 5x5.8 in large transverse diameter, with an intermediate heterogeneous signal in T2 and isointense in T2, in hyper diffusion, containing hemorrhagic zones and others in vacuum of signal, intensely enhanced after injection of PDC, delimiting areas of loss of substance with contour irregularities, achieving a shredded cauliflower appearance at the level fundic. Relative respect for the zonal anatomy of the cervical region (Figure 1). T2 Weighted transversal section show the target sign (figure2). The Axial DWI and T1 gadolinium images show a vaginal heterogeneous hyper intense mass enhanced by gadolinium (figure 3)

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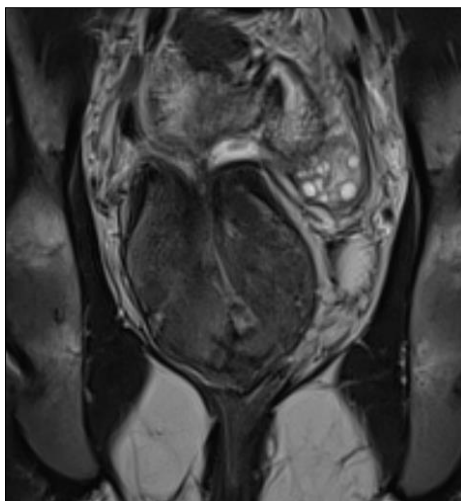


Figure1: T2- Weighted MRI of complete non-puerperal uterine inversion with the uterine corpus in a U-shape above the mass. The cervix surrounds the corpus and the vaginal fornix surrounds both the corpus and cervix.



Figure2

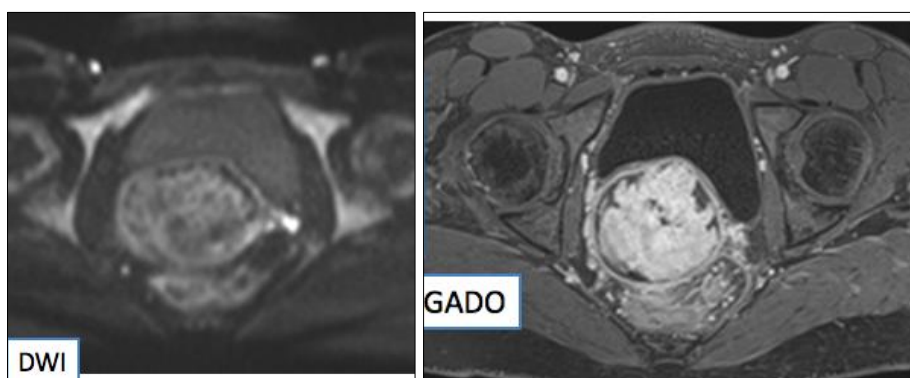


Figure 3

The first results of the pathologist's report of the uterine mass prolapsed through the cervix at the vulvar

level demonstrated a malignant tumor proliferation with sarcomatous components (figure 4).

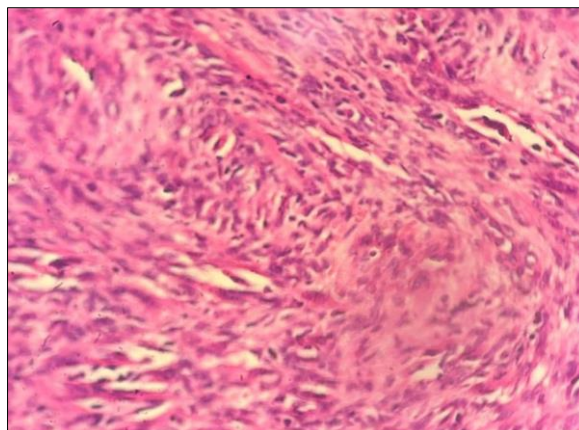


Figure 4: Uterine sarcoma showing nuclear atypia and numerous mitotic figures.

After a diagnosis of non-puerperal uterine inversion, the patient was transferred to the operating room to undergo an abdominal hysterectomy. During laparotomy, the fundus was found to be inverted into the cervix pulling with it healthy appearing ovaries and fallopian tubes (figure5).

A total hysterectomy and bilateral oophorectomy were conducted following Haultin method. The patient recovered from the surgery with no complications (figure5).

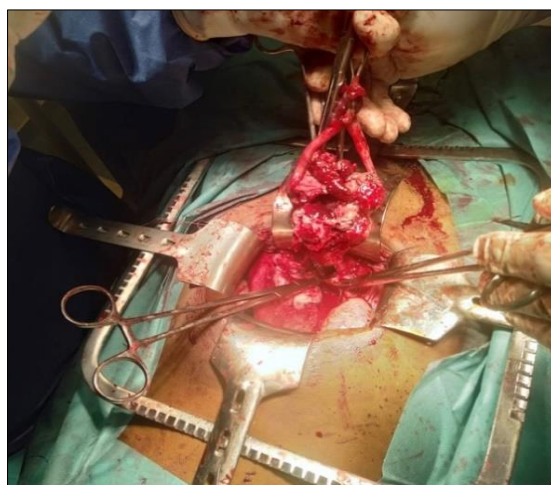


Figure 5: Per op view



Figure 6: The resected inverted uterus with tumor on surface

Histopathology revealed a malignant tumor growth of sarcomatous origin in favor of embryonal rhabdomyosarcoma. The results showed that the surgical

resection margins were free. The immunochemistry was in favor of embryonal rhabdomyosarcoma: with positive myogenin.

Post-operatively, the patient benefited from 16 sessions of chemotherapy based on actinomycin D, and ifosfamide.

The CT-scan with intravenous contrast showed an intraparenchymal nodule in the middle lobe with no malignancies. The patient has been well for two years with no signs of illness to date.

DISCUSSION

Non-puerperal uterine is caused most of the time by benign tumors, although rare cases involve malignant tumors like leiomyosarcoma, endometrial carcinoma, rhabdomyosarcoma, immature teratoma, and

others. Rhabdomyosarcoma (RMS), a mesenchymal cell-derived tumor with subtypes including embryonal, alveolar, spindle cell/sclerosing, and pleomorphic, typically affects areas like the head and neck, urogenital tract, and extremities.

Genitourinary rhabdomyosarcoma has two peak incidences: between 2 to 6 years and 15 to 19 years of age [3]. Unfortunately, RMS carries a grim prognosis, with mortality rates reaching 47%, and 20% of cases experiencing recurrence [4]. However, RMS causing uterine inversion is exceptionally uncommon, with only seven reported cases in English literature up to date, including the present instance [Table 1].

Table 1: Summary of the literature

References	Age (years)	Histology	presentation	Mass size (cm)	Treatment	Follow-up
Sharma <i>et al.</i> , [2]	18	RMS	Bleeding Vaginal mass	17×15	TAH +BSO, CT	Pelvic recurrence
Ambreen <i>et al.</i> , [3]	22	ERMS	Bleeding Vaginal mass	10×8	TAH+BSO CT, RT	Live metastasis, DOD
Peng <i>et al.</i> , [4]	19	ERMS	Venous sinus thrombosis	18×16	VH + BSO, CT	Pelvic recurrence
Case <i>et al.</i> , [5]	21	ARMS	Bleeding	10.2×8.2	NACT, TAH + BSO, CT	ANED
Ojiwang <i>et al.</i> , [6]	16	ERMS	Bleeding	10.5×9	NACT, TAH, RT	
da Silve <i>et al.</i> , [7]	15	ERMS	Bleeding Vaginal mass	11×9.7	TAH, CT, RT	Vaginal recurrence, DOD
Current case	15	ERMS	Hemorrhagic Hypovolemic shock	7×5	TAH+BO CT	ANED

ARMS, alveolar RMS; ERMS, embryonal RMS; TAH, total abdominal hysterectomy; BSO, bilateral salpingo-oophorectomy; BO, Bilateral-oophorectomy; VH, vaginal hysterectomy; CT, chemotherapy. RT, radiotherapy; NACT, neoadjuvant chemotherapy; ANED, alive with no evidence of disease; DOD, dead of disease.

The primary clinical symptom reported is vaginal bleeding, leading to potential complications such as anemia or hypovolemic shock, often accompanied by a foul-smelling vaginal discharge. Additionally, patients may experience abdominal cramps, pelvic discomfort, heaviness sensation, or dysuria. During a bimanual pelvic examination, the uterine fundus may not be palpable. Diagnosis can be facilitated by the presence of a mass in or outside the vagina [4].

Uterine inversion is classified into four stages:

Stage 1: The uterus Inversion is intrauterine or incomplete. The fundus remains within the cavity.

Stage 2: A complete inversion of the uterine fundus through the fibromuscular ring of the cervix.

Stage 3: Total inversion, whereby the fundus protrude through the vulva.

Stage 4: The vagina is also involved with complete inversion through the vulva along with the inverted uterus.

Magnetic Resonance Imaging has proven to be a very useful diagnostic tool, In T2 weighted MRI scan, a U-shaped uterine cavity with a thickened, inverted uterine fundus on a sagittal image and a “bull’s eye” configuration on an axial image are signs indicative of uterine inversion.

Treatment for uterine inversion is multidisciplinary, starting with patient stabilization. Surgical approach selection depends on factors like etiology, inversion stage, patient age, and fertility desires. Radical hysterectomy is favored for late-stage or malignant cases and for women seeking pregnancy. Alternatively, tumors may be resected with uterine fundus repositioning.

Various techniques exist for repositioning the uterus, including Huntington and Haultain for abdominal approaches, and Kustner and Spinelli for vaginal approaches. The Haultain procedure, involving posterior

incision of the constriction ring followed by uterine reversion, has shown success [8, 9].

Vincristine, actinomycin-D, and cyclophosphamide (VAC) chemotherapy, with or without radiotherapy, are standard for 1-10 cycles [10].

Repositioning's impact on fertility post-nonpuerperal uterine inversion surgery remains uncertain, with a recommended 12-month postoperative period before assessing pregnancy.

CONCLUSION

Uterine inversion is an emergency associated with serious morbidity and mortality. The prognosis depends on the rapidity of the detection, identification, and management of the disease and its histopathology.

Chemotherapy has an important role in the management of uterine sarcomas.

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