

Case Report

# Epithelioid Trophoblastic Tumor (ETT) Confined to the Uterus: A Case Report and Brief Literature Review

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## Abstract

**Introduction:** ETT is a rare entity, with few reports in the literature. There is no prospective data regarding this issue and its management is based in retrospective series of cases and specialists opinion.

**Case presentation:** We present a case of a 33 years old woman, 5 pregnancies (3 para) who presented with spontaneous vaginal bleeding after an abortion. Pelvic MRI revealed uterus of 162 cm<sup>3</sup>, and a nodular lesion of 4,3x 4,1x3,3cm in isthmus, compromising uterine cavity. She underwent a hysteroscopy whose biopsy material was highly necrotic, with proliferative pattern but no malignancy identified. Next was performed a Laparoscopic total hysterectomy plus bilateral salpingo-oophorectomy with a pathological finding of ETT confined to the uterus. The hCG level was undetectable. We decided to put the patient on follow up.

**Conclusion:** Management of ETT is a challenge due to its rarity and diverse presentation, requiring efforts on grouping cases together in order to understand its evolution.

**Keywords:** Epithelioid Trophoblastic Tumor; Gestational Trophoblastic Disease

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**Consent:** Consent was taken from the patient for publication of this case report.

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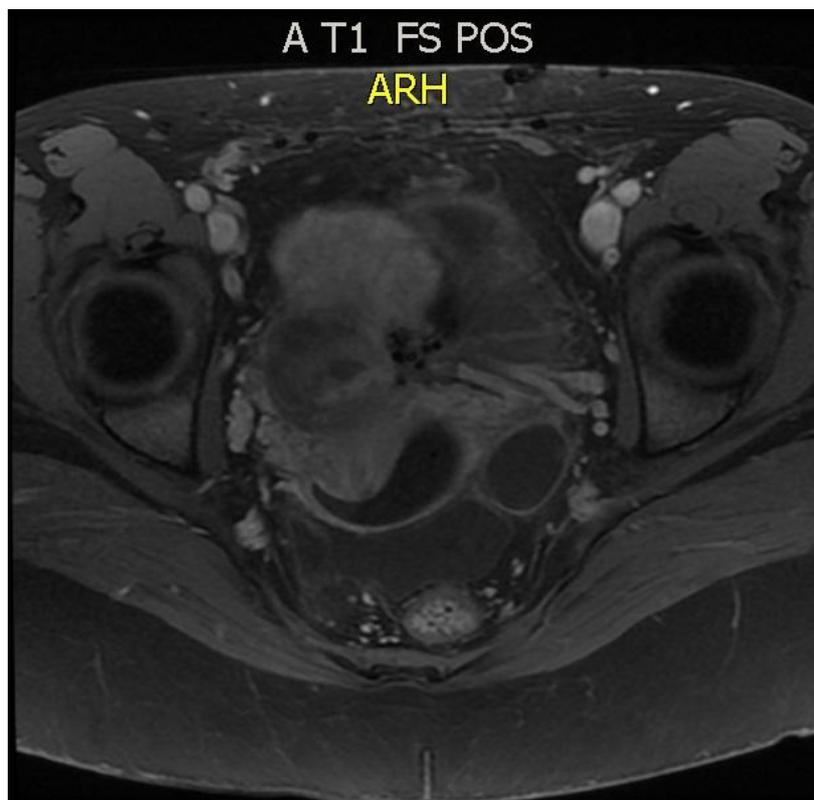
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## Introduction

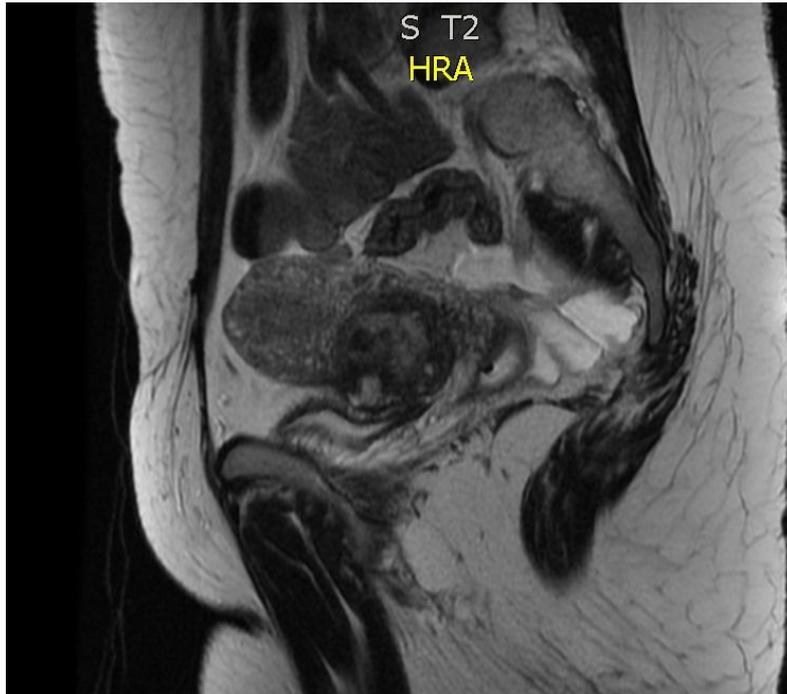
Epithelial Trophoblastic Tumor (ETT) is a rare entity, first described by Mazur and Kurman in 1994 [1] and incorporated into the classification of gestational trophoblastic disease as a distinct subtype in 1998 by Shih and Kurman [2]. Since then, about 108 cases were reported in the literature [3,4]. There is no prospective data regarding this issue and its management is currently based in the analyses of retrospective series of cases and specialists opinion.

## Case Presentation

33 years-old, female, gravida 5, para 3 (3 cesarean deliveries), 2 spontaneous abortions, presents in our department on May 2015 with the following medical history. In September/2014, after a spontaneous abortion, started a continuous vaginal bleeding. She sought medical assistance in primary care in January 2015, and a transvaginal ultrasound was performed with the finding of a hypoechoic and heterogeneous lesion, about 3,8cm in the largest diameter, evolving myometrium, serosa and submucosa. An MRI of the pelvic region revealed a heterogeneous nodular formation measuring 4,3x4,1x3,3cm in the uterus right isthmic position, communicating with endometrial and cervical cavity (Figures 1,2,3). Beta Human Chorionic Gonadotropin (hCG) dosage by that time was undetectable.



**Figure 1** Transversal view of Heterogeneous nodular formation measuring: 4,3x4,1x3,3cm in the uterus right isthmic position, communicating with endometrial and cervical cavity. T1 pos = T1 view after contrast administration



**Figure 2** Sagittal view of Heterogeneous nodular formation measuring 4,3x4,1x3,3cm in the uterus right isthmic position, communicating with endometrial and cervical cavity. T2 = T2 view



**Figure 3** Coronal view of Heterogeneous nodular formation measuring 4,3x4,1x3,3cm in the uterus right isthmic position, communicating with endometrial and cervical cavity. T2 = T2 view

Gynecological local examination had no relevant findings. In February/2015, a Hysteroscopy with biopsy was performed, but the pathological diagnosis was inconclusive – extensively necrotic material.

Next, she went through a Laparoscopic total hysterectomy plus bilateral salpingo-oophorectomy with the pathological description of a Tumor constituted by cells with intermediated trophoblastic deposit of hyalinous material in between neoplastic cells. Extensive necrosis and decidualized stromal cells could be seen in the periphery of the lesion. Tumor size = 2,5cm; IM 9/10 CGA; Immunohistochemistry - bHCG = positive in isolated cells; Ck18 = positive; p63 = positive; PLAP = positive; Inhibin = positive; CD146 = positive; hPL = positive; MUC4 = negative; Ki67 = present in 40% of the cells – Compatible with the diagnosis of Epithelioid Trophoblastic tumor.

There was no evidence of spread disease on CT scans of the chest and abdomen. hCG follows undetectable after surgery.

Because of localized disease, short period since pregnancy and issues regarding the true benefit of chemotherapy, we decided to put patient in follow up.

## Discussion

The vast majority of ETT cases have been reported in women of reproductive age and its most frequent symptom is abnormal vaginal bleeding, which is present in 69% of the cases [3]. Most patients will have an elevated hCG at the onset of the disease [4]. Regarding the primary site, ETT is most frequently localized in the uterus (40%), followed by the cervix (31%). Extra uterine disease is also described - the lungs account for 19% of the cases [3]. Time since pregnancy and disease development is very variable, ranging from 1-18 years after antecedent pregnancy, which in 67% of cases will be full term [4].

Pathological features are described as the finding of a nodular growth of nested and corded monomorphic epithelioid cells often with areas of necrosis [5]. These tumors may be confused with squamous cell carcinomas because of its similar location in the lower uterine segment and cervix, and positive immunohistochemical staining for cytokeratin and p63 [4].

The mortality associated to ETT, similarly to other GTD, is low, though the rate of metastasis has been reportedly high, ranging 25-88,9% [2,6]. Because of its rarity, the biologic behavior of ETT is not firmly established. Despite absence of strong literature data, metastatic disease at presentation, interval greater than 5 years from antecedent pregnancy, extra uterine disease and high mitotic rate appears to be poor prognostic factors [4,5].

Surgery is the cornerstone of treatment, and hysterectomy should be advised in patients with non-metastatic disease to maximize the opportunity for cure [4,5]. In general, ETT shows a poor response to multiple agent chemotherapy, in spite of that, the majority of the case reports offer multi-agent chemotherapy, and since there is a significant variability of regimens and lack of standardization of therapy, it is difficult to draw conclusions regarding its results [3,4].

Management of ETT is a challenge due to its rarity and diverse presentation, requiring efforts on grouping the case reports together, in order to understand the evolution of the disease and make better decisions regarding its treatment.

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