

Pseudo-invasive Vasculocentric Adenomyosis- A Diagnostic Dilemma

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Abstract

Pseudo-invasive, vasculocentric adenomyosis is a rare benign lesion of the uterus, characterized by the aberrant, pathological presence of variable sized, non-neoplastic endometrial glands and surrounded by endometrial stroma deep within the myometrial blood vessels. This condition usually affects multiparous women in their fourth and fifth decades, with other risk factors being prior caesarian surgery or uterine surgery. Mostly the patients present with abnormal uterine bleeding, pelvic pain, dysmenorrhea, dyspareunia, or infertility while a third of them may be asymptomatic. Microscopic examination of hysterectomy specimens remains the gold standard to make a definitive diagnosis. Herein we present a rare case of Pseudo-invasive, vasculocentric Adenomyosis in a 47 year old multigravida female, who presented with a history of chronic pelvic pain with menorrhagia, dyspareunia and excessive menstrual bleeding for the past four months, with a history of lower uterine caesarean section a decade ago.

Keywords: Adenomyosis- Immunohistochemistry- Pseudo-invasive- Vasculocentric- Uterus

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Introduction

Pseudo-invasive, vasculocentric adenomyosis is a rare benign condition of the uterus, characterized by the pathological presence of variable sized, non-neoplastic endometrial glands surrounded by endometrial stroma deep within the myometrial blood vessels [1].

Over the past years, the disease was seen in multiparous women in their fourth and fifth decade, with other risk factors being prior caesarian surgery or uterine surgery [2,3]. Advanced imaging techniques may be a help to diagnose pseudo-invasive adenomyosis in young women [4,5]. However, microscopic examination of hysterectomy specimens still remains the gold standard to make a definitive diagnosis of pseudo-invasive vasculocentric adenomyosis [6].

The majority of patients affected present with abnormal uterine bleeding, pelvic pain, dysmenorrhea, dyspareunia, or infertility while a third of them may be asymptomatic [7,8]. Adenomyosis often coexist with other hormone dependent disease entities such as endometriosis or uterine fibroids [9].

Case Report

A 47 year old multigravida female presented with a history of chronic pelvic pain with menorrhagia, dyspareunia and excessive menstrual bleeding which had worsened over the past four months. There was limited improvement in her symptoms following pain medications. She had a history of lower uterine caesarean section a decade ago. There was no significant family history.

Pelvic examination revealed an enlarged retroverted uterus. Transabdominal pelvic ultrasound revealed an irregularly thickened endometrium. Magnetic resonance imaging (MRI) showed ill-defined endometrial borders and a poorly circumscribed, ovoid, eccentric, heterogeneous, vascular myometrial thickening, particularly in the lower uterine myometrium. An endometrial biopsy only demonstrated features of a disordered proliferative endometrium and no neoplastic elements. Given the unremitting nature of her symptoms and completed family, the patient consented to a total abdominal hysterectomy and bilateral salpingectomy.

Gross examination of the uterus showed endometrial

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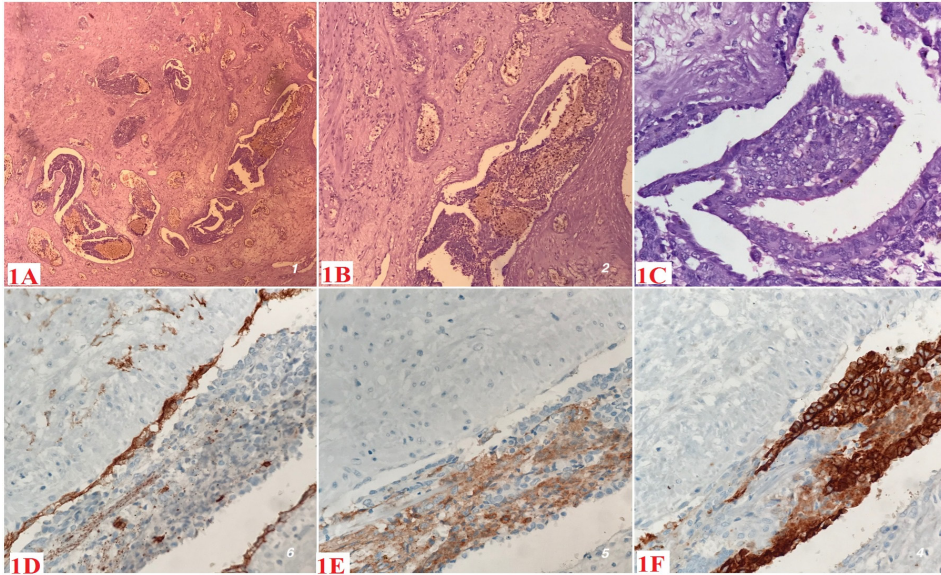


Figure 1A. Tissue Section Shows Several Vascular Spaces, with Intravascular Lesion Evident at Low Power. H & E x10; Figure 1B, Section in higher magnification shows lesional tissue within vascular spaces. H & E x20; Figure 1C, Section shows some glandular strips and condensed stromal cells. H & E x40; Figure 1D, Immunoperoxidase stain shows vascular endothelium highlighted by CD31. IHC CD 31 x40; Figure 1E, Immunoperoxidase stain shows stromal cells expressing CD10. IHC CD10 x40; Figure 1F, Immunoperoxidase stain shows glandular cells positive for CK7, stromal cells are negative. IHC CK7 x40.

thickness of 8.0 mm and multiple brownish specks with accompanying whorling and nodular myometrium, focally located deep within the uterine wall, almost upto the serosa. No other abnormality was noted in the remaining specimen.

Histopathologic evaluation showed attenuated endometrial glands and rather prominent specialised stroma, almost exclusively within thick and thin walled mural blood vessels (Figure 1A-1C). Immunohistochemistry corroborated the morphology with the vessels being highlighted by CD31 (Figure 1D); the stromal cells marking for CD10 (Figure 1E) and the endometrial glands expressing CK7 (Figure 1F). There was no cytologic atypia of either the glands or the stroma. Based on the above findings, a diagnosis of Pseudo-invasive, vasculocentric adenomyosis was given. This patient remains completely symptom free and well 6 months into follow-up.

Discussion

Adenomyosis is described as a distinct entity in the PALM-COEIN FIGO (polyp; adenomyosis; leiomyoma; malignancy and hyperplasia; coagulopathy; ovulatory dysfunction; endometrial; iatrogenic; and not yet classified – International Federation of Gynecology and Obstetrics) classification [10].

The junctional zone is the site of origin of cycle dependent contractions seen at various phases of the menstrual cycle [11]. Disturbances in these contractions leads to repeated trauma to the junctional zone, resulting in invasion of myometrium by the basal layer of the endometrium through this altered hormone responsive zone [12]. This theory explains the risk factors (multiparity, prior uterine surgery) related to the development of adenomyosis [11,12]. Another hypothesis suggests

metaplastic changes in the myometrial stem cells to play a role in disease development [13].

Adenomyosis presents with non-specific symptoms, also seen in other gynaecological disorders. Chronic pelvic pain is common which may be mediated by prostaglandin [14]. Various studies have demonstrated a significant correlation between the depth of adenomyosis and the severity of abnormal uterine bleeding (AUB) [15,16].

The clinical differential diagnosis for intra-myometrial lesions includes leiomyoma, adenomyoma/adenomyosis, hematoma, atypical adenomyoma, sarcomas and endometrial carcinoma [17,18]. The histologic findings of benign endometrial glands and stroma misplaced within smooth muscles are typical of adenomyosis [19]. The present case is morphologically exceptional, in having a pseudo-invasive, predominantly vasculocentric growth pattern of adenomyosis.

Whether adenomyosis occurs secondary to de novo, non-neoplastic intrusion of endometrium via caesarean surgery lacunae, or it reflects a developmental anomaly, or is of stem cell origin – is debatable, but the vasculocentric predilection displayed by the present case is even more intriguing. Given its rarity, it is well worth remembering this entity, when countenanced with uterine intra-myometrial infiltrative appearing lesions, that do not ‘quite fit’, clinically or morphologically.

Author Contributions

RT wrote the manuscript, KA edited the manuscript and is the article guarantor, AA edited the manuscript and approved the final manuscript.

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