Stromal Adenomyosis, a mimic of Low-grade Endometrial Stromal Sarcoma: A Case Report and a Literature Review

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Abstract

Stromal adenomyosis is an exceptionally rare condition with only a few reported cases. It involves invasion of endometrial tissue into the myometrium and is characterized by the predominance of stroma with sparse glands in the affected area. This unique feature makes diagnosis challenging and often resembles low-grade endometrial stromal sarcoma (LGESS). Additional tissue sections must be obtained to facilitate differentiation between them. Typically, stromal adenomyosis is incidentally discovered during uterine removal performed for unrelated reasons. In this report, we present a case of stromal adenomyosis in a patient of reproductive age, a departure from previously documented cases primarily observed in postmenopausal women. Moreover, stromal adenomyosis can mimic LGESS, posing a diagnostic challenge.

Keywords: Adenomyosis; Stromal adenomyosis/ Adenomyosis with sparse glands; Low grade endometrial stromal sarcoma; Case report.

Introduction

Adenomyosis is a benign chronic gynecological condition in which the endometrium invades the myometrium and is characterized by the presence of endometrial glands and stroma.[1,2] In some cases, a variant known as stromal adenomyosis, also referred to as adenomyosis with sparse glands, may occur.[3,4] Although rare, this benign disease has been reported in only a few cases.[3,4] It is distinguished by the predominance of stroma over glands in the affected area. Notably, stromal adenomyosis can mimic low-grade endometrial stromal sarcoma (LGESS), highlighting the importance of accurately differentiating between the two conditions.[3,4] In this context, we present a case of stromal adenomyosis in which the initial clinical and radiological diagnosis was a fibroid. Furthermore, careful microscopic examination and immunohistochemistry successfully distinguished stromal adenomyosis from LGESS in this study.

Case Report

A 43-year-old female presented to our institution's emergency department with severe vaginal (PV) bleeding. She is para 4 with one previous lower segment caesarean section. She is diabetics on oral therapy. She has a known history of menorrhagia due to uterine fibroids since 2016 and previously underwent a myomectomy abroad. The patient has experienced a recurring pattern of multiple emergency department visits for heavy PV bleeding and hospital admissions for low hemoglobin levels, which required blood transfusions. She did not improve with oral medical therapy. Her last menstrual period was one month prior to the current presentation.

The pelvis ultrasound revealed the following findings:

1. The patient was actively bleeding during the imaging

2. Uterus: Anteverted, bulky, and heterogeneous in echo texture (volume = 1294.1cc).

3. The previously identified posterior fibroid is still present, measuring 6.7 * 7.7 cm in size and pushing the cavity upwards.

4. Endometrial thickness measures 10mm, with a focal lesion of 4.0 * 2.6 cm within the cavity, raising suspicion of a polyp or growth.

5. Both ovaries appear normal in shape and echo texture.

6. No obvious adnexal pathology or free fluid is observed.

The medical recommendation for the patient was a total hysterectomy; however, she reluctant to undergo this procedure as she was keen to get pregnant. Consequently, myomectomy was performed, and the intraoperative findings were as follows.

" The entire myometrium displayed a doughy to firm consistency with no definite fibroids identified, likely indicating adenomyoma. A firm mass-like fibroid, measuring 6 * 5 cm was excised. The posterior endometrium was opened and a mass was removed. Additionally, a polypoid mass measuring approximately 3 * 2 cm, displaying discoloration, was noted and removed".

Grossly, the specimen exhibited an irregular, pale, firm mass weighing 129 grams and measuring 100 * 60 * 80 mm. On one surface, an ellipse of smooth mucosal tissue measuring 40 * 20 mm. Serial slicing revealed a whorled, pale, and haemorrhagic cut surface. (Figure 1)

Microscopically, the sections revealed uterine smooth muscle containing multiple foci and aggregates of benign endometrial stromal cells, devoid of endometrial glands. These foci exhibited a distinctive concentric zonal organization, characterized by a central basal staining area of less cellular, loosely aggregated stromal cells surrounded by a more cellular thin rim of stromal or smooth muscle cells. The nets of gland-poor stromal cells were encircled by a thicker but less well-defined layer of hypertrophic myometrial smooth muscles. Individual stromal cells within these aggregates had a monotonous appearance with small, uniform nuclei. No hyaline sclerosis, foam cells, sex cord-like structures, richly vascular network, or hemangiopericytoma-like areas were observed. One focus showed features typical of adenomyosis, consisting of intramural aggregates of endometrial stromal cells with few scattered endometrial glands. No mitosis, atypia, or necrosis was appreciated. There was no evidence of leiomyoma, and the surrounding myometrium appeared hypertrophic. (Figure 2)

Immunohistochemistry was performed to rule out malignancy. The stromal cells were strongly and diffusely positive for CD10 and progesterone receptor (PR). ER showed faint and focal positivity. They tested negative for CK7, desmin, and SMA. Ki67 demonstrated significantly low (~1%). (Figure 3)

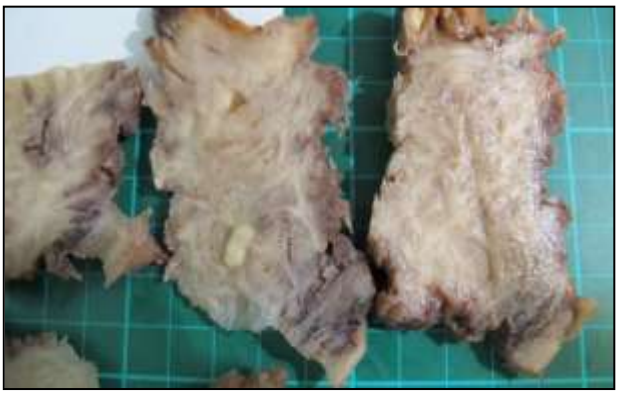

Figure 1: Gross images: irregular pale mass with whorled haemorrhagic cut surface.

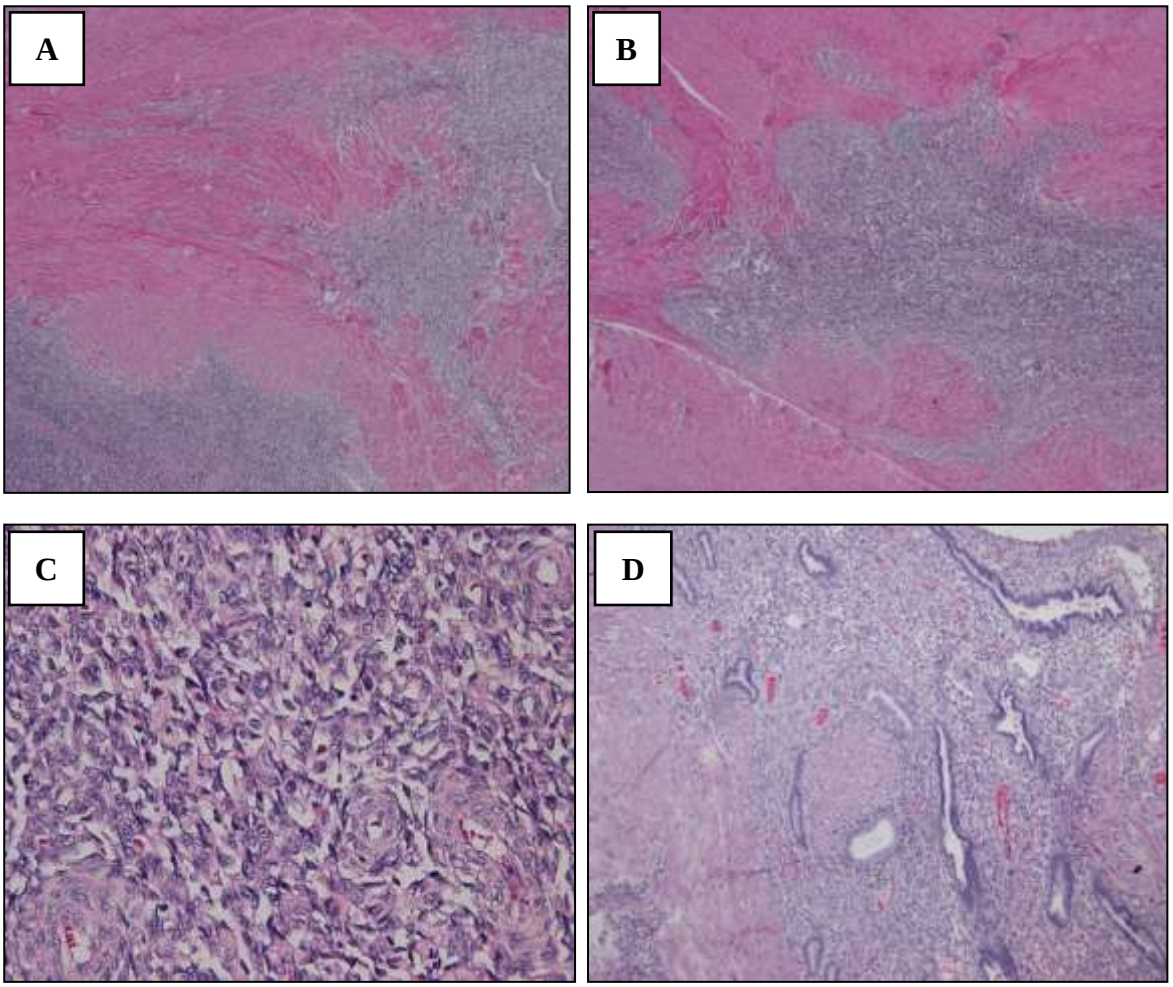

Figure 2: Microscopic images. **A, B**: Low power view shows endometrial stromal cell aggregates infiltrating uterine smooth muscle, devoid of glands, **C**: High power view illustrates bland looking endometrial cells, **D**: Only one focus reveals sparse glands.

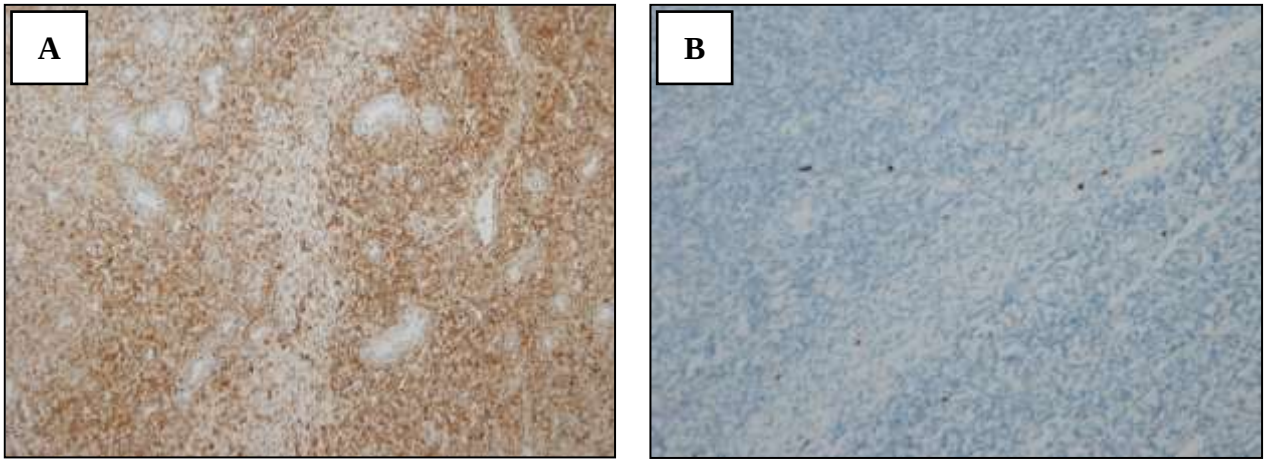

Figure 3: Immunohistochemistry. **A**: CD10 is positive, **B**: Ki67 is very low.

Considering the microscopical findings with immunohistochemistry results, Pathological diagnosis concludes:

- Stromal adenomyosis/ adenomyosis with sparse endometrial glands
- No evidence of malignancy
- No features of leiomyoma are seen.

Follow-up: The patient is still being followed up by the obstetrics and gynaecology department at our institution for recurrent fibroids. She underwent a third myomectomy abroad but continues to present to our hospital's emergency department with similar picture of PV bleeding and low hemoglobin, requiring blood transfusion. She and her partner continue to refuse hysterectomy despite a thorough explanation of all risks. Another myomectomy is planned.

Discussion

Uterine adenomyosis is one of the most common benign and chronic gynecological condition.[5] Earlier theories linked adenomyosis to uterine trauma, hormones, and dysperistalsis, but its occurrence without surgery or pregnancy suggests that endometrial-myometrial dysfunction may plays a role.[6] Although adenomyosis is typically easy to recognize, stromal adenomyosis with sparse glands is a relatively uncommon and under-discussed form of adenomyosis, characterized primarily by the presence of endometrial stroma with fewer glands compared to classical adenomyosis. The diagnosis of stromal adenomyosis can be challenging as it may resemble low-grade endometrial stromal sarcoma (LGESS).[3,4,7] LGESS is a sarcoma with low malignancy that often exhibit morphological similarities to normal proliferative-type endometrial stromal cells, accompanied by a distinctive network of small arteriole-like vascular channels. Distinguishing features include infiltrative edges, vascular invasion, and 'tongue-like' pattern of infiltration into the myometrium.[8] The overlapping histopathological and imaging features between the two conditions are where the diagnostic challenge lies. While adenomyosis is a benign condition often managed conservatively, LGESS requires timely surgical intervention to prevent disease progression. Moreover, late diagnosis of LGESS potentially increases the risk of metastasis which can significantly impact prognosis.[6] To ensure accurate diagnosis thorough sampling and careful pathological evaluation needed for appropriate management plan and improve patient outcomes.

In our case, the initial imaging suggested fibroids, and histologically, it resembled LGESS. However, further examination revealed no polymorphisms, mitotic activity, infiltration, or lympho-vascular invasion. Immunohistochemistry examination was performed using CD10, PR, and ER to highlight the stroma. Desmin and SMA were used to exclude leiomyoma, as the specimen was initially suspected to be fibroids. CK7 was utilized to highlight the endometrial glands and rule out sarcomatoid carcinoma. Ki67 was assessed to determine the proliferative index and exclude sarcoma. The findings confirmed the presence of a non-malignant tumor, demonstrating a low proliferation index of approximately 1%. Both LGESS and stromal adenomyosis typically show positivity for PR, CD10, and ER. However, LGESS can also express smooth muscle markers (SMA, desmin, caldesmon), particularly in areas of smooth muscle differentiation.[9] Despite this overlap, a detailed microscopic examination of additional tissue sections revealed sparse glands, confirming the diagnosis of stromal adenomyosis/adenomyosis with sparse glands.

A literature review identified key features distinguishing stromal adenomyosis from LGESS, including incidental discovery during uterine removal for other reasons, microscopic size without observable tumor nodules, concentric zonal organization of gland-poor stromal aggregates, atrophic stromal cells without nuclear atypia, and the absence of sclerotic areas, foam cells, sex cord-like structures, prominent vascular invasion, and extrauterine extension.[3] As opposed to previously published cases of stromal adenomyosis, our patient, although in her reproductive age, exhibited all these features, suggesting that stromal adenomyosis can occur not only in postmenopausal women but also in women of reproductive age.

Conclusion

In this report, we described a rare case of stromal adenomyosis, highlighting a novel aspect of its occurrence in women of reproductive age. Additionally, stromal adenomyosis is typically discovered incidentally, indicate its potential manifestation at any age. However, previous cases have predominantly involved postmenopausal women, likely due to their higher frequency of undergoing gynecological procedures than younger females. Furthermore, stromal adenomyosis can closely mimic adenomyosis requiring thorough sampling and careful pathological evaluation to ensure an accurate diagnosis. We suggest that the diagnosis of stromal adenomyosis should not be influenced by the patient's reproductive status. It is important to obtain additional samples and conduct a thorough examination, both grossly and microscopically, as stromal adenomyosis may resemble fibroids or low-grade endometrial stromal sarcoma (LGESS). Immunohistochemistry plays a crucial role in differentiating these conditions, and consulting a colleague, as well as reviewing previous cases, can further support an accurate diagnosis.

Disclosure

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