Case Report

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A Case Report of High-Grade Endometrial Stromal Sarcoma in a 32-Year-Old Woman

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ABSTRACT

High-grade endometrial stromal sarcomas (HG-ESS) represent infrequent neoplasms originating from the endometrial stromal layer, predominantly afflicting perimenopausal women. Their incidence in adolescents and young adults is exceedingly rare. Given the scarcity of occurrences of this particular tumor, the available literature is notably deficient in comprehensive documentation concerning its clinical manifestations and diagnostic imaging. Herein, we present an instance of HG-ESS in a 32-year-old nulligravida woman, whose initial presentation involved abnormal uterine bleeding and hypermenorrhea. Magnetic resonance imaging of the abdominal region for our patient revealed a conspicuously irregular tumor measuring 10cm x 10cm x 9cm, inducing an expansion of the endometrial cavity, accompanied by the identification of multiple nodules in the cul-de-sac. Subsequently, a comprehensive intervention was undertaken, involving total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy, and elective lymphadenectomy. The ascertained diagnosis pertained to stage IIIC HG-ESS, prompting subsequent administration of adjuvant radiotherapy and chemotherapy. The exceptionally low prevalence of HG-ESS underscores its status as a highly unusual affliction. The unfavorable prognosis associated with this disease necessitates maintaining a heightened level of suspicion for HG-ESS when evaluating cases of abnormal uterine bleeding and abdominal pain.

Abbreviations: HG-ESS: High-Grade Endometrial Stromal Sarcomas; ESS: Endometrial Stromal Sarcoma; MRI: Magnetic Resonance Imaging; CT: Computed Tomography

Introduction

Endometrial stromal sarcoma (ESS) represents a rare neoplasm of endometrial stromal lineage, constituting a mere 0.2% of all uterine malignancies and comprising 10% of uterine sarcomas [1,2]. It predominantly manifests in the premenopausal demographic. High-grade endometrial stromal sarcoma (HG-ESS) is prevalent among individuals of advanced age, with an average age approximating 60 years, displaying a proclivity for heightened aggressiveness [3,4]. Common presenting manifestations of HG-ESS encompass irregular vaginal bleeding, discernible masses, and pelvic discomfort. The occurrence of HG-ESS in adolescents and young women is exceedingly uncommon, with scant documented cases in the extant literature [5]. Therefore, an amalgamation of documented instances of this exceptional clinical entity is imperative for a more comprehensive understand of its inherent trajectory. This study delineates a singular case of HG-ESS in a 32-year-old woman, whose presentation featured hypermenorrhea and the presence of a pelvic mass.

Case Report

A 32-year-old nulligravida woman presented with a six-month history of hypermenorrhea, accompanied by a palpable mass in the lower abdomen exhibiting rapid enlargement. Her medical history was devoid of noteworthy events. Transvaginal ultrasonography revealed an enlarged uterus, surpassing 10 cm in overall size, hosting a heterogenous mass within the endometrial cavity. Magnetic resonance imaging (MRI) disclosed an enlarged and well-defined uterine border with heterogeneous thickening in the endometrial cavity, encompassing the entire uterine corpus and cervix (Figure 1). A biopsy from the endometrial cavity was procured, and its histopathological

examination indicated the potential presence of undifferentiated carcinoma with a heterogeneous component. The clinical suspicion pointed towards an advanced-stage malignancy of the cervix. At surgery, the uterus exhibited enlargement, and multiple nodules were identified in the cul-de-sac. The uterine serosa displayed diffuse involvement, harboring numerous tumor plaques. Neither the adnexae nor the omentum manifested any remarkable features. The surgical interventions encompassed total abdominal hysterectomy, bilateral salpingo-oophorectomy, partial omentectomy, and resections of enlarged pelvic lymph nodes.

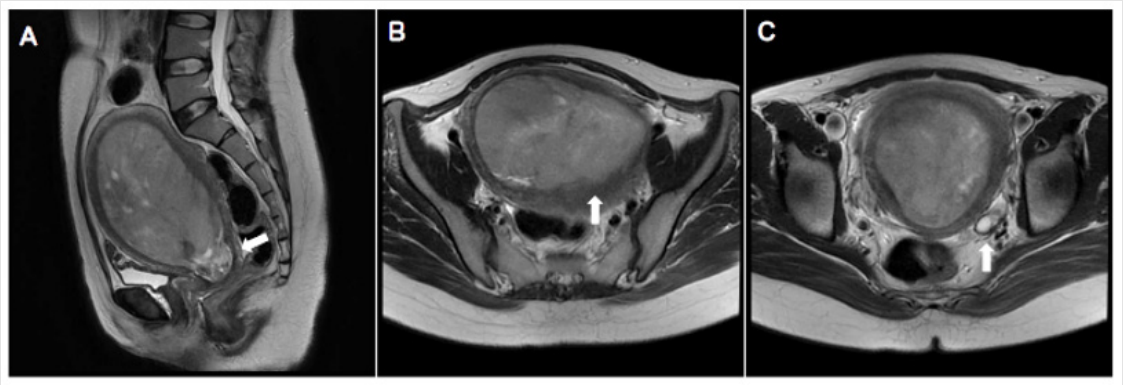

Figure 1:
A. Sagittal T2-weighted MRI shows an enlarged over 10cm heterogenous mass in the endometrial cavity and endocervical canal (arrow).
B. Axial finding shows an enlarged well-defined border of endometrial cavity with involving the posterior myometrium (arrow).
C. Normal appearance of left ovary was visible (arrow).

The postoperative phase transpired without incidents, and the final histopathological report delineated high-grade endometrial stromal sarcoma with a heterologous element, characterized by more than 10 mitoses per 10 high-power fields (Figure 2). Additional immunohistochemistry studies unveiled positive diffuse staining for CD34 and vimentin. The tumor exhibited negative staining for estrogen receptor, and progesterone receptor, melan A, c-kit, and EMA. A diagnosis of stage IIIC high-grade endometrial stromal sarcoma was established based on these investigative outcomes. During a two-week deliberation concerning the diagnosis of high-grade ESS and the life-threatening nature of the condition, the patient encountered rapid disease progression, accompanied by vaginal bloody discharge emanating from the exophytic polypoid mass (1 x 1 cm) on the vaginal stump while undergoing adjuvant radiotherapy. Despite being slated to initiate adjuvant chemotherapy involving adriamycin, her employer and social supporters were diligently working towards facilitating her further treatment in her home country.

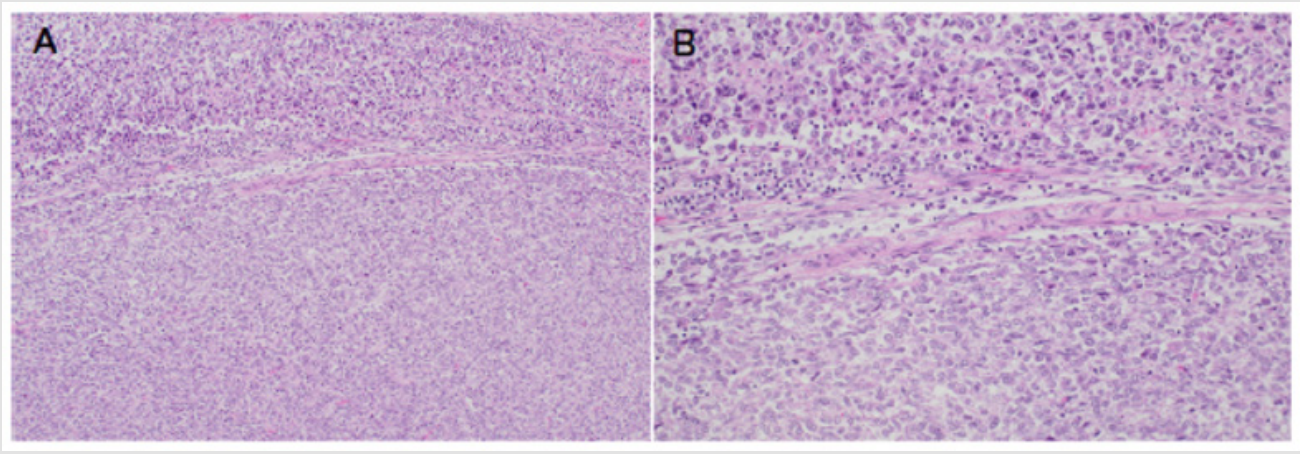

Figure 2: Histopathological images of the resected tumor.
A. Hematoxyline-eosin staining (x100).
B. Hematoxyline-eosin staining (x200), consistent with high-grade stromal sarcoma.

Comment

Due to the low incidence of HG-ESS, a dearth of distinct clinical manifestations and imaging features exists. A comprehensive literature review revealed eight singular case reports detailing high-grade endometrial stromal sarcoma occurrences in adolescents and young adults [5]. These instances often manifest as abnormal uterine bleeding, acute abdominal pain, and the presence of a pelvic mass, and hypermenorrhea in the current case. All previously documented cases were diagnosed at advanced stages (III and IV) during the initial assessment. Notably, three cases were reported in teenage girls [6-8], with one of them experiencing menarche at the age of 14 and commencing oral contraception at 15 [6]. HG-ESS exhibits significantly distinct biological characteristics from its low-grade counterpart: while low-grade ESS may manifest as estrogen-dependent, the investigation into HG-ESS occurrences in adolescents is ongoing [9]. Transvaginal ultrasonography and endometrial curettage, although beneficial in diagnosing endometrial polyps, uterine leiomyomas, or adenomyosis, prove unpredictable in diagnosing ESS. Magnetic Resonance Imaging (MRI) may identify ESS through the presence of heterogeneous areas within the lesion caused by aggressive tumor permeations into the myometrium. When coupled with contrast-enhanced Computed Tomography (CT), these modalities contribute to the preoperative diagnosis of potential extents, as seen in the current case. Treatment protocols encompass debulking surgery, followed by total hysterectomy with bilateral salpingo-oophorectomy and omentectomy [1,10,11].

The contentious nature of pelvic and para-aortic lymphadenectomy stems from its uncertain role [12]. Given the heightened risk of recurrence and metastasis associated with HG-ESS, postoperative adjuvant therapy assumes a pivotal role. To date, chemotherapy and pelvic external radiotherapy have been widely employed as adjuvant measures for HG-ESS. Notably, anthracyclines exhibit specific efficacy against sarcomas. Amant et al. [13] chronicled a case of transperitoneal HG-ESS complicating a pregnancy at 15 weeks, exploring diverse avenues to sustain the pregnancy, including anthracycline-based chemotherapy and cytoreductive surgery, even during gestation. Unfortunately, the patient succumbed to the disease six weeks post-diagnosis. In summary, we elucidated the clinical and imaging findings of a rare case involving a 32-year-old woman afflicted by HG-ESS. Despite the ongoing intensive adjuvant chemotherapy, the bleak prognosis associated with this disease underscores the imperative to maintain a high degree of suspicion for HG-ESS in the evaluation of any case presenting with abnormal uterine bleeding and the presence of a rapid growth of a pelvic mass.

Conflict of Interest

The authors declare that they have no conflict of interest.

References

1. Chan JK, Kawar NM, Shin JY, Osann K, Chen LM, et al. (2008) Endometrial stromal sarcoma: A population-based analysis. Br J Cancer 99(8): 1210-1215.
2. Rauh Hain JA, del Carmen MG (2013) Endometrial stromal sarcoma: A systematic review. Obstet Gynecol 122(3): 676-683.
3. Puliyath G, Nair MK (2012) Endometrial stromal sarcoma: A review of the literature. Indian J Med Paediatr Oncol 33(1): 1-6.
4. Adiga CP, Gyanchandani M, Goolahally LN, Itagi RM, Kalenahalli KV (2016) Endometrial stromal sarcoma: An aggressive uterine malignancy. J Radiol Case Rep 10(9): 35-43.
5. Ushida S, Ichigo S, Takagi H, Matsunami K, Ito Y Imai A (2023) High-grade endometrial stromal sarcoma in adolescents and young adults: a case report and literature review. Biomed J Sci Tech Res 49(2): 40522-40525.
6. Michalas S, Creatsas G, Deligeoroglou E, Markaki S (1994) High-grade endometrial stromal sarcoma in a 16-year-old girl. Gynecol Oncol 54(1): 95-98.
7. Wang KC, Liang DC, Su TH, Hung FY, Yang (1998) High-grade endometrial stromal sarcoma in a 10-year-old girl: Case report. Changgeng Yi Xue Za Zhi 21(3): 312-317.
8. Emmert C, Dietrich F, Wecke WD, Riedel HH (1999) High-grade endometrial stromal sarcoma (HG-ESS) in an adolescent girl with benign mediastinal Schwannoma. J Obstet Gynaecol 19(2): 210–212.
9. Maccaroni E, Lunerti V, Agostinelli V, Giampieri R, Zepponi L, et al. (2022) New insights into hormonal therapies in uterine sarcomas. Cancers (Basel) 14(4): 921.
10. Zhang YY, Li Y, Qin M, Cai Y, Jin Y, et al. (2019) High-grade endometrial stromal sarcoma: A retrospective study of factors influencing prognosis. Cancer Manag Res11: 831-837.
11. Bai H, Yuan F, Liang B, Sun H, Gao Y, et al. (2022) Clinicopathological characteristics and treatment of patients with high-grade endometrial stromal sarcoma: A retrospective study of 40 cases. Medicine (Baltimore) 101(2): e28490.
12. Amant F, Coosemans A, Debiec Rychter M, Timmerman D, Vergote I (2009) Clinical management of uterine sarcomas. Lancet Oncol 10(12): 1188-1198.
13. Amant F, Van Calsteren K, Debiec Rychter M, Heyns L, De Beeck KO, et al. (2010) High-grade endometrial stromal sarcoma presenting in a 28-year-old woman during pregnancy: A case report. J Med Case Rep 4: e243.

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