

Uterine Adenosarcoma with Low Grade Overgrowth and Smooth Muscle Differentiation Mimicking Leiomyosarcoma

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Case Report

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Abstract

Background: Adenosarcoma is a rare mixed mesodermal tumor of the uterus which is composed of benign glandular epithelium and malignant mesenchymal stroma.

Case Presentation: 59-year-old postmenopausal women, presented with vaginal bleeding. Per vaginal examination showed a whitish polypoid mass protruding through the cervix which appeared to originate from the uterine body. Magnetic resonance imaging (MRI) revealed an intravaginal polypoid tumor originating from the lateral wall of the uterus. The biopsy from the polypoid mass revealed a leiomyosarcoma. Therefore, the patient underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy. The histological examination revealed a biphasic tumor composed of benign and dilated endometrial glands and malignant mesenchymal components which were composed of fascicles of spindle shaped cells with mild to moderate nuclear atypia and many mitosis. There are with areas of periglandular cuffs made by small round cells around glands. Immunohistochemical stains were performed and showed that round cells were positive for CD10, while spindle shaped cells were stained with AML and desmine. Histopathological final diagnosis was adenosarcoma with low-grade sarcomatous overgrowth and extensive smooth muscle differentiation. The patient has no evidence of disease, and she is doing well 18 months after the surgery.

Conclusion: We report a rare case of Uterine adenosarcoma with low grade overgrowth and smooth muscle differentiation mimicking leiomyosarcoma.

Keywords: Adenosarcoma; Low Grade Overgrowth; Smooth Muscle Differentiation; Mixed Mullerian Tumors

Introduction

Uterine adenosarcoma is an uncommon malignant tumor, first described by Clement and Scully in 1974. It

was defined as "...mixed tumors of the uterus, in which the stromal component has been malignant, but the epithelial elements, benign [1]. This tumor occurs most frequently in postmenopausal women [2]. However,

exceptional cases have been reported in children [3]. The pathologic diagnosis is based on characteristic morphologic features with Immunohistochemical study. We here report a case of adenosarcoma mimicking a leiomyosarcoma of uterine corpus.

Case Presentation

A 59-year-old lady postmenopausal for more than 10 years, with vaginal bleeding. She had no past medical or surgical history. Per vaginal examination showed a large polypoid and whitish mass protruding through the cervix which seemed to originate from the uterine body. Magnetic resonance imaging (MRI) revealed an

intravaginal polypoid tumor of 87× 70mm originating from the lateral wall of the uterus (Figure1). A smooth Muscle Tumor (Leiomyoma or leiomyosarcoma) was suspected. A biopsy from the polypoid mass was performed with immunohistochemical study. Histopathological diagnosis was a leiomyosarcoma. Serum levels of CA19-9, CA125 and CEA were normal. The patient underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy. Gross examination of surgical specimen revealed a polypoid whitish mass measuring 7x8 cm protruding through the cervix and arising from the lateral wall of uterine corpus. It was firm with cystic areas. There was no macroscopic invasion of the myometrium.

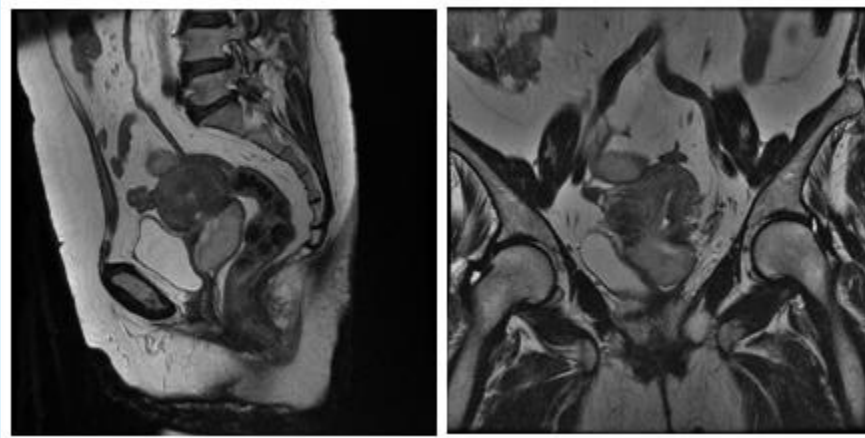


Figure 1: MRI imaging for adenosarcoma (A and B): both transverse plane and coronal plane showed an intravaginal polypoid tumor originating from the lateral wall of the uterus.

Histologically, the tumor was made by predominantly mesenchymal component (more than 25%) which was composed of fascicles of spindle shaped (Figure 2) cells with mild to moderate nuclear atypia. Mitosis exceeded 40 per 10 high power fields. Extensive sampling showed benign dilated endometrial glandular elements with periglandular cuffs made by areas small round cells (Figure3&4). No myometrial or lympho-vascular invasion was seen. Immunohistochemical stains were performed and showed that round cells were positive for CD10, while spindle shaped cells were stained with AML and desmine. Histopathological final diagnosis was adenosarcoma with extensive smooth muscle differentiation and low-grade sarcomatous Overgrowth. The patient has no evidence of disease 18 months after the surgery and she is doing well.

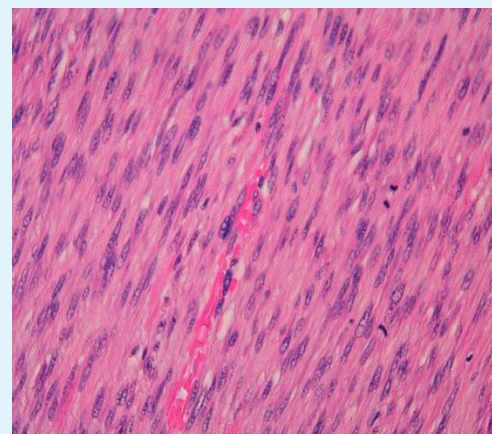


Figure 2: The tumor was made by fascicles of spindle shaped cells with mild to moderate nuclear atypia and numerous mitosis (HESx20).



Figure 3: Benign dilated endometrial glandular element with predominate mesenchymal component (HEx5).

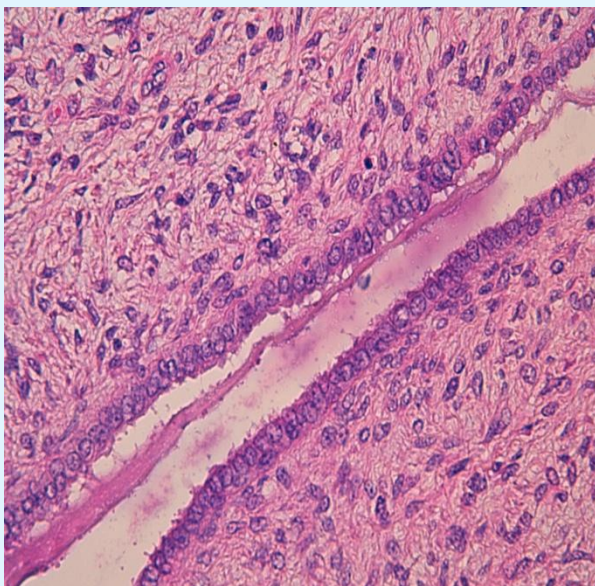


Figure 4: Areas of small, round cells that concentrate around the glandular elements forming peri-glandular cuffs (HESx10).

Discussion

Adenofibroma, adenosarcoma and carcinosarcoma are the three types of uterine mixed epithelial-mesenchymal tumors. Adenofibroma is composed of benign glandular

elements and benign mesenchymal stroma, whereas carcinosarcoma is composed of malignant epithelial elements and mesenchymal stroma. Adenosarcoma is a rare entity which has benign glandular elements and malignant stroma [4], representing 3 to 9 % of uterine cancers [5]. Some studies reported that estrogen stimulation; the use of tamoxifen, ororal contraceptive pill may increase the risk of Adenosarcoma [6-8].

In the current case, patient had no history of Long-term hormone therapy. The most common clinical symptoms are genital bleeding, pelvic pain or demonstration of a pelvic mass. MRI is the imaging modality of choice to examine the local extent of disease. It shows a large uterine mass that is hypointense on T1, with cystic appearance on T2 [5]. It typically presents as an exophytic or polypoid mass, originating from the uterine wall or fundus which protrudes from the cervix [4].

Such tumor is usually observed in the endometrium, but can arise in the cervix, ovaries, and fallopian tubes. Histologically, adenosarcoma is composed of benign glandular elements and malignant stroma that may compress the benign epithelium, and then resembling a phyllodes tumor of the breast. Condensation of small stromal cells around dilated glands is characteristic and it is called Periglandular cuffing. The epithelium may show mild to moderate atypia and is usually endometrioid. The mesenchymal component is classically a low-grade spindle cell sarcoma with mitoses (at least 2 per 10 high-powerfields). The main histological prognostic factors are sarcomatous overgrowth, and deep myometrial invasion.

Sarcomatous overgrowth is defined as the presence of pure mesenchymal component without any epithelial elements comprising at least 25% of the tumor. It typically consists of high grade sarcomatous areas with severe cytologic atypia, high mitotic activity, and necrosis; it is associated with a poor outcome. However, rare cases of low grade sarcomatous overgrowth, as in our case have been reported [9,10]. In the current case, the majority of the sampled tissue showed a low grade mesenchymal neoplasm with smooth muscle differentiation resembling a leiomyosarcoma with only a minor component showing a conventional adenosarcoma. This is why it was initially misinterpreted as a leiomyosarcoma on biopsy. Furthermore, Smooth muscle differentiation has rarely been reported in the literature [9,11-13] and consists typically in compact fascicles of smooth muscle cells as it was observed in our case. At immunohistochemical study, mesenchymal cells of

adenosarcomas is typically stained by ER, PR, CD10, and WT1. Other markers that can be positive in adenosarcomas like vimentin, smooth muscle actin, desmin, CD34, calretinin, and AE1/3 cytokeratin [14]. The Ki67 index is usually <5% but more than 20% in areas of sarcomatous overgrowth or glandular cuffing.

Lymph node metastasis rate is low, therefore staging lymphadenectomy is not recommended in patients with disease confined to the uterus and without high risk factors. No systemic or adjuvant treatment have been validated for adenosarcoma, however chemotherapy can be used in patient with adenosarcoma with sarcomatous overgrowth.

Conclusion

Mullerian Adenosarcoma should be considered in the differential diagnosis of polypoid mass protruding from the cervix. The final diagnosis is usually made on surgical specimen and it is based on characteristic morphologic features with *benign epithelial component* and *malignant mesenchymal component*.

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Competing Interest: No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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