

Adenosarcoma of the Uterus: A Case Report

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ABSTRACT: Adenosarcoma is a biphasic tumor with benign epithelial elements and a sarcomatous stroma. Typically it presents as a solitary large polypoid mass arising from the uterine fundus. In this case report we presented a 64 year-old woman with postmenopausal bleeding and endometrial polypoid lesion in sonographic report. Pathologic examination of the curetting specimen revealed a mixed epithelial-mesenchymal tumor that it was diagnosed as adenosarcoma.

Keywords: adenosarcoma, postmenopausal bleeding.

INTRODUCTION

Adenosarcoma is a biphasic tumor with benign epithelial elements and a sarcomatous stroma(1). This entity was originally described by Clement and Scully in 1974 as Müllerian adenosarcoma(2). Although it usually arises in the endometrium, it can arise in the cervix, the myometrium, fallopian tubes, and ovaries. (1). Adenosarcoma is a rarely observed mullerian tumor of the uterus and represents 8% of uterine sarcomatous tumors(3). Occurs in women of all ages. The median age is 50 years, with a range of 13-83years (4). Although adenosarcoma is typically low grade tumor, recurrences have been reported in up to 30–40% of patients while 20–25% of women die from their tumors (1).

Typically it presents as a solitary large polypoid mass arising from the uterine fundus and fills the endometrial cavity and protrudes from the uterine cervix(1). The most common presenting symptom is abnormal vaginal bleeding. The diagnosis can be made with endometrial curettage(5).

The aim of our experience is to show the difficulty of the diagnosis with endometrial biopsies and difficulty of the diagnosis from other Mixed epithelial-mesenchymal tumors such as adenofibromas or carcinosarcomas.

Case presentation:

A 64-years-old woman in our department in November 2015, presented with postmenopausal bleeding. The gynecologic examination revealed that the uterus and ovaries were of normal size and that the parametrical tissues were free. Cytological tests of uterine cervix were negative. The ultra-sonographic evaluation showed a hyperechogenic 3 cm mass in the endometrial cavity. The ovaries were of normal size and appearance. Endometrial curetting was done. Pathologic examination of the specimen revealed a mixed epithelial-mesenchymal tumor composed of broad papillary and polypoid stromal fronds covered by epithelium project from the surface of the neoplasm or extend into cystic space within it. Tubular glands and cleft like spaces distributed throughout the tumor which occasionally resulting in phyllodes tumor-like appearance(fig 1). Prominent squamous epithelial component is also present(fig 2). The mesenchymal component consisting of mixtures of endometrial stromal cells and fibroblast with mild hypercellular appearance especially in periglandular area(fig 4,5). Mesenchymal cells were showed mild nuclear atypia and mitotic feature in about 1-2/10HPF(fig 5). The differential diagnosis including adenosarcoma and adenofibroma were discussed.

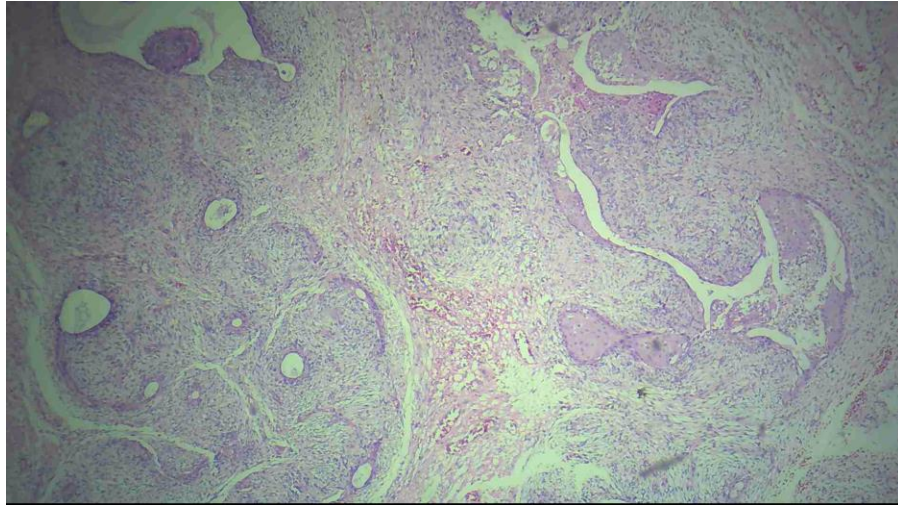


Fig 1. mixed epithelial-mesenchymal tumor with cleft like spaces(phyllodes tumor-like)

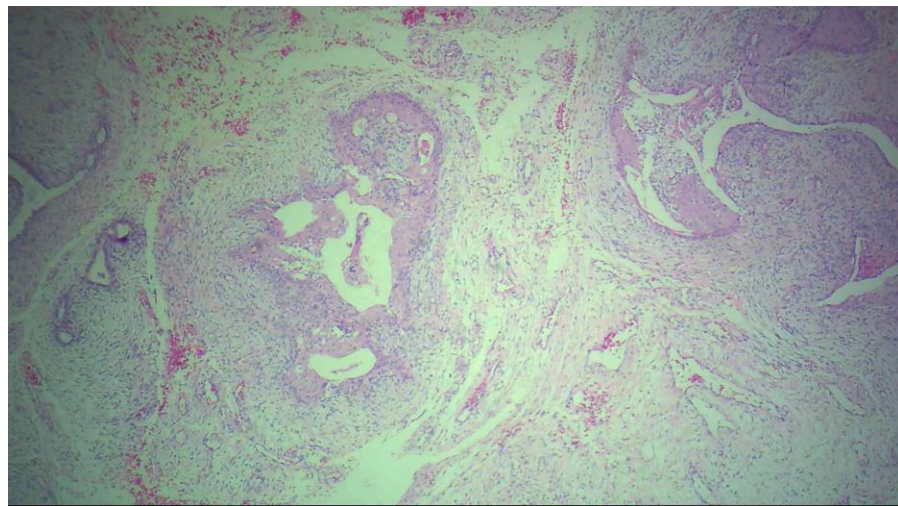


Fig 2. mixed epithelial-mesenchymal tumor with prominent squamous epithelial component

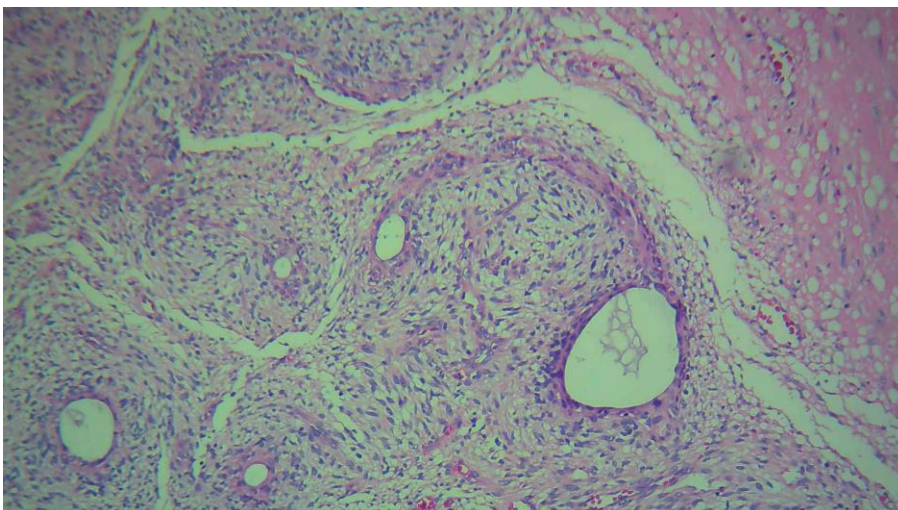


Fig 3. Increased cellularity of stroma around the epithelial elements forming a cambium layer

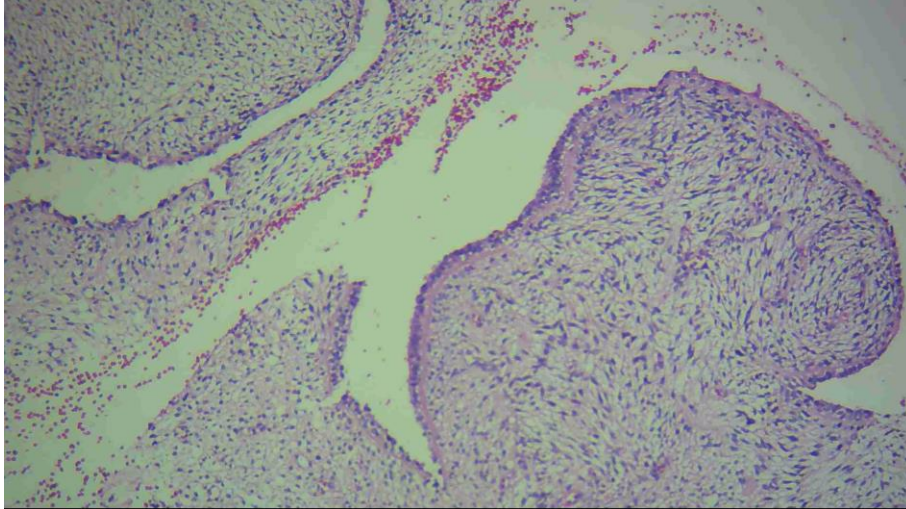


Fig 4. Increased cellularity of stroma around the epithelial elements forming a cambium layer.

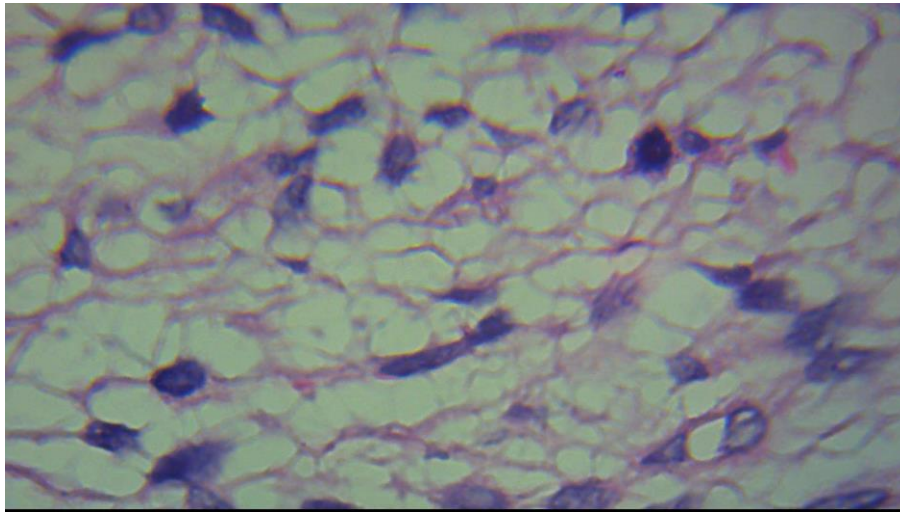


Fig 5. Mesenchymal cells were showed mild nuclear atypia and mitotic feature.

Laparotomy was planned on the basis of this diagnosis. After the peritoneal washing, a total abdominal hysterectomy-bilateral salpingo-oophorectomy was performed along with extirpation of the parametrium and pelvic-para-aortic lymph node sampling. At laparotomy, the uterus was fist size. On opening in lower segment of uterus a small polypoid lesion measuring 2x0.5x0.5 cm was seen. On microscopic examination the tumor showed glandular epithelium with little atypia and proliferation of atypical mesenchymal cells. Mitosis exceeded 2 per 10 high power fields. No myometrial invasion or lymph node metastasis was seen. The lesion was confined to the uterus. no serosal invasion was observed. Both ovaries were intact. Peritoneal cytology revealed no malignant cells. Histopathological final diagnosis was adenosarcoma.

Discussion:

Mixed epithelial-mesenchymal tumors of the uterus include adenofibroma, adenosarcoma, and carcinosarcoma. Adenofibroma has benign glandular epithelial element and benign mesenchymal stroma, where as carcinosarcoma has both malignant epithelial and mesenchymal stroma. Adenosarcoma is one of the rare diseases consisting of benign glandular epithelial element and malignant mesenchymal component(1) .It has been mostly reported in post-menopausal women but few cases were observed in women between the ages of 19 and 40 years (6,7). Clinical symptoms vary from vaginal bleeding to pelvic pain and to protrusion of the tumor from the cervix or vagina (5) .In most cases, adenosarcoma presents as a yellow to grey endometrial polyp(8).

Microscopically, the glands were lined by benign or atypical glandular epithelium, together with sarcomatous stromal cells which showed characteristic structures of 'periglandular cuff' of increased cellularity and 'intraglandular polypoid projections. (9)

The epithelium typically is cytologically bland, but hyperplastic and even atypical hyperplastic epithelium is occasionally noted (10). Increased cellularity of stroma around the epithelial elements forming a cambium layer could be a useful clue suggesting in diagnosis of adenosarcoma (8). Using the World Health Organization definition, stromal mitotic activity of 2 or more per 10 high-power fields is required for a diagnosis of adenosarcoma but in practice the diagnosis is made with stromal mitotic activity less than this if the characteristic architecture and cambium layer is present (11).

The diagnosis could be sometimes only made or confirmed after multiple pathologic examinations. In some cases, many endometrial samples are needed in order to find uterine adenosarcoma (12). In fact, in our case, the diagnosis of uterine adenosarcoma was difficult and only suspected after multiple histologic analysis. Because of suspected lesion, hysterectomy was performed and permitted to confirm histologically the diagnosis of uterine adenosarcoma. Adenosarcomas are characterized by weaker malignancy potential, late-onset local recurrence (25%), and rare metastasis. recurrence is observed more frequently in the vagina and pelvis (60%) (5). Unfavourable prognostic factors are sarcomatous overgrowth, deep myometrial invasion, presence of heterologous elements and extrauterine spread (13).

The epithelial component of adenosarcoma is keratin positive, and it usually stains for estrogen and progesterone receptors.

The mesenchymal cells in adenosarcoma typically show cytoplasmic staining for CD10, and there is nuclear staining for estrogen and progesterone receptors and for WT-1 (14).

Adenofibroma is a rare benign tumor of the cervix or endometrium. Over time the diagnostic criteria for adenosarcoma have been broadened to the point that some authors now doubt the existence of adenofibromas (4). The overall architecture is similar to adenosarcoma, but the stroma is predominantly fibrous and less cellular than that of an adenosarcoma, with no condensation of stromal cells around the epithelial elements. The stromal cells are bland, and MF are absent or difficult to find. Some adenosarcomas contain bland areas similar in appearance to an adenofibroma, so the diagnosis of an adenofibroma generally requires hysterectomy so that the entire tumor can be evaluated microscopically (15).

Adenosarcoma is usually treated by hysterectomy and bilateral salpingo-oophorectomy. Metastasis to lymph nodes is rare, although occasional patients whose tumors show sarcomatous overgrowth have pelvic lymph node metastases (16). Adenosarcomas without sarcomatous overgrowth do not require adjuvant therapy of any type. Hormonal therapy may be an option in rare cases with advanced or recurrent disease. adenosarcomas with sarcomatous overgrowth should be treated according to recommendations for high-grade sarcomas. (17)

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