Mullerian Adenosarcoma of Uterus with Sarcomatous Overgrowth and Heterologous Component Associated with Stromal Deposit in Omentum

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Abstract:

Mullerian adenosarcoma with sarcomatous overgrowth (MASO) is a very rare variant of uterine sarcomas described by Clement as early as 1974. The presence of heterologous sarcomatous components is associated with aggressive biological behavior. Case presentation. This is a case report of a 62-year female (P) presenting with postmenopausal vaginal bleeding. Her preoperative USG revealed subserosal fibroid with adherent omentum. She underwent abdominal hysterectomy with bilateral oophorectomy. Histopathological diagnosis of resected specimen was Mullerian adenosarcoma with sarcomatous overgrowth and presence of heterologous elements involving body of the uterus. The whole thickness of the myometrium was involved along with the presence of serosal nodules and omental deposits of sarcomatous component. Conclusion. MA is considered as a low-grade malignant tumor, but MASO is a high-grade tumor frequently associated with invasion and metastasis with poor treatment outcome. Because of its rarity, correct identification of these tumors and distinction from other uterine sarcomas are a challenging job and hence its morphological features merits attention.

Keyword: Mullerian adenosarcoma, sarcomatous overgrowth, sarcomatous cells

Introduction:

Malignant stromal tumors account for 1–3% of all female genital tract tumors and Mullerian adenosarcomas (MAs) constitute 8–10% of these malignancies. Typically, MA involves endometrium and presents as a polyoid mass. But infrequently this malignancy is reported from ovaries, cervix, vagina, peritoneum, and even pouch of Douglas. MA is a tumor of postmenopausal women and the commonest presentation is vaginal bleeding. According to classical description by Clement and Scully, MA is a mixed stromal and epithelial neoplasm. Stromal component shows malignancy but glandular tissue, though at focal areas may present with atypical proliferation and is devoid of malignant features. Further classification of MA is done depending upon mesenchymal elements; homologous tumors are composed of nonspecific spindle-shaped sarcomatous cells whereas special differentiation of mesenchymal components into cartilage, osteoid, or striated muscles is associated with heterologous neoplasms Mullerian adenosarcoma with sarcomatous overgrowth (MASO) is a rare variant of MA, characterized by growth of a second pure high-grade sarcoma occupying at least one-fourth of the tumor mass. In contrast to MA, MASO runs a biologically aggressive course.

Case Report:

This 62-year female came to gynecology OPD with complains of postmenopausal vaginal bleeding. Preoperative ultrasonography of pelvis revealed presence of a subserosal fibroid with omental nodules. She underwent abdominal hysterectomy with bilateral salpingoophorectomy along with excision of omental nodules.

Figure 1 (a) Gross photograph of omental tissue showing haemorrhagic, necrotic fleshy mass. (b) Gross photograph of cut section of uterus showing fleshy mass distorting uterine cavity with invasion of myometrium with areas of haemorrhage and necrosis (Red arrow), bony tissue (black arrow), and subserosal nodule (blue arrow).

Figure 2 Photomicrograph showing phyllodes-like architecture on low power (H&E stain, ×100).
Sarcomatous overgrowth in mullerian adenosarcoma is a distinctive type of mullerian mixed tumor. It is challenging to differentiate from MA because stromal atypia and focal condensation are often present. Extensive squamous metaplasia and fibromyxomatous stroma favor diagnosis of APA though close follow up is mandatory. Adenosarcomas are considered as a low-grade malignant neoplasm with reported local recurrence in 25% cases. The most important histologic poor prognostic indicators are sarcomatous overgrowth and deep myometrial invasion. Other unfavorable prognostic factors include presence of heterologous elements, necrosis, high mitotic rate, and extra uterine spread. Vascular invasion, rarely seen, represents a poorer prognosis. Overall metastasis is reported in 5% of cases with the metastatic component solely represented by the sarcomatous element, consistent with findings of the present case. Median survival time of patients with uterine MASO is 13 months. In this paper, we are presenting a rare uterine tumor with uncommon histological features. Involvement of whole thickness of myometrium with formation of serosal nodules and omental deposits is a rare presentation of MASO. Treatment outcome in this particular case was poor because of association of multiple unfavorable prognostic factors like sarcomatous overgrowth, deep myometrial invasion, and extra uterine spread. However, MASO should be taken into differential diagnosis of all high-grade uterine sarcomas and multiple sections must be examined to rule out presence of benign glandular element with periglandular cuffing of stromal

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