

Mullerian Adenosarcoma Of The Uterus With Sarcomatous Overgrowth

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Citation

V Geetha, S Rupashree, S Bhat. *Mullerian Adenosarcoma Of The Uterus With Sarcomatous Overgrowth*. The Internet Journal of Pathology. 2008 Volume 8 Number 1.

Abstract

Mullerian adenosarcoma is an uncommon variant of mixed mesodermal tumour of the uterus. This is a case report of a 47-year-old lady who initially presented with amenorrhea followed by abnormal vaginal bleeding and a mass protruding through the os which was biopsied. Histopathological examination revealed a tumour comprising of an admixture of benign endometrial glandular component and a sarcomatous stromal component characteristic of adenosarcoma. Patient was lost to follow-up for 2 years, and returned subsequently with relapse of symptoms following which abdominal hysterectomy with polypectomy and bilateral salphingo-oophorectomy was done. The polyp contained an adenosarcoma with sarcomatous overgrowth. The distinctive morphological features of this rare tumour are emphasized here.

INTRODUCTION

Mullerian adenosarcoma is a rare mixed biphasic neoplasm generally presenting with abnormal vaginal bleeding and polypoidal masses characterized by benign glandular elements and malignant stromal component. Because of their generally low grade of malignancy, these should be distinguished from the more aggressive forms like carcinosarcoma and sarcoma of endometrium and from benign adenofibroma. This may pose a problem in diagnosis due to its rarity, and hence its morphological features merit attention.

CLINICAL SUMMARY

A 47-year-old lady presented with amenorrhea of 6 months duration followed by vaginal bleeding for which a biopsy was done and diagnosed as adenosarcoma. She was lost to follow-up for 2 years after which she returned with relapse of vaginal bleeding, loss of weight and appetite. On examination a polypoidal mass measuring 4 x 1 cms with foul smelling discharge, enlarged uterus with restricted mobility and indurated parametrium was noted following which cervical polypectomy with total abdominal hysterectomy, bilateral salphingo-oophorectomy and partial omentectomy was done and specimen sent for histopathology.

PATHOLOGICAL FINDINGS

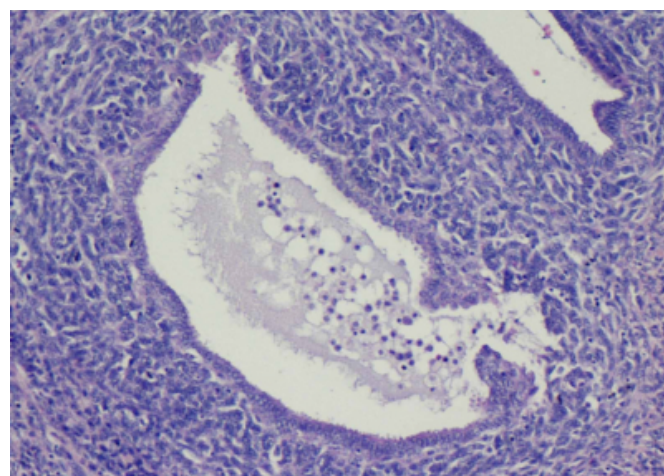
Grossly endometrial cavity was distorted with irregularly

thickened myometrium. Separately sent polyp with attached stalk measured 6x5x2 cms and showed grey white-haemorrhagic areas with occasional cystic spaces

For light microscopy conventional Haematoxylin and Eosin (H&E) stained slides were examined. The stalk of the polyp showed a tumour with varied morphological pattern comprising of numerous cystically dilated endometrial glands with periglandular overgrowth of neoplastic stromal cells (fig-1)

Figure 1

Figure 1: Periglandular cuffing by neoplastic cells (H&E X 100)



having plump hyperchromatic nuclei and patchy foci of

stromal hyalinization, while rest of the polyp showed extensive areas of sarcomatous overgrowth characterized by sweeping fascicles of spindle shaped cells with numerous mitoses ($>4/10\text{hpf}$) admixed with bizarre and multinucleated forms.

Figure 2

Figure 2: Pure sarcomatous component of the tumour (H&E X200)

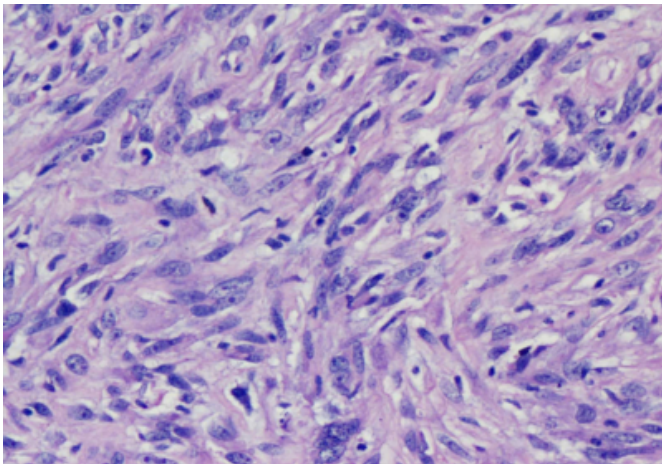
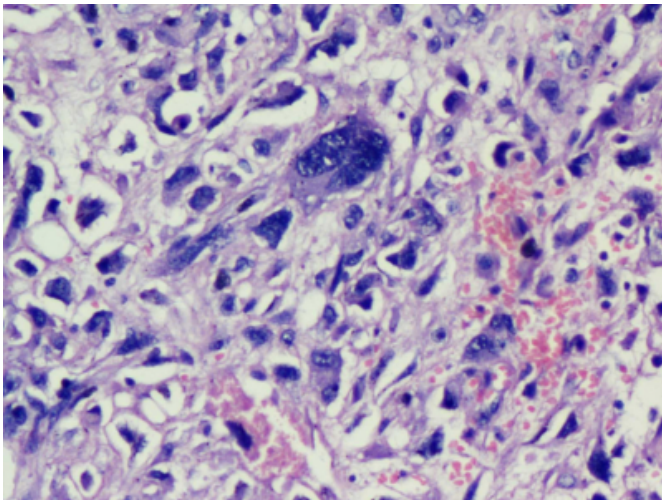


Figure 3

Figure 3: Bizarre tumour cells in the sarcomatous component (H&E X400)



interspersed thin walled vascular channels and few entrapped glands showing squamous and oncocytic metaplasia. The sarcomatous component comprised more than 10% of the polyp. The endometrium showed a small focus of adenosarcoma. Myometrium, isthmic endometrium, cervix, both ovaries and omentum were all free of tumour.

DISCUSSION

Mullerian adenosarcoma is a rare malignant tumour of the

uterus that was described by Clement and Scully¹. It is characterized by a benign, but occasionally atypical glandular component and a sarcomatous, usually low grade, stromal component. Majority of adenosarcomas, including the current case, occur in post menopausal women who presented with history of post menopausal bleeding. These tumours are usually of low malignant potential. Few of these tumours are clinically malignant, since many of them pursue an indolent course. Zaloudek et.al² noted that the only morphological feature associated with increased risk of recurrence or metastasis is the presence of deep myometrial invasion.

Mullerian adenosarcoma is characterized by an intimate admixture of benign endometrial glands and a sarcomatous stroma. These glands may be lined by proliferative endometrioid, endocervical, squamous, serous or secretory endometrioid cells and are surrounded by the sarcomatous component. The latter had a characteristic relationship with the glands i.e stromal cellularity is more around the glands resulting in the formation of “periglandular cuffs”. The stroma can be endometrial stromal sarcoma alone, fibrosarcoma alone or a mixture of the two. Further, stromal secondary changes including fibrosis and variable hyalinization may also be seen. The presence of extensive hyalinization may result in an appearance similar to a benign endometrial polyp³. In addition, foci of edema, haemorrhage, inflammation and myxoid change may be present.

The term “Mullerian adenosarcoma with sarcomatous overgrowth (MASO)” was first used by Clemant and Scully⁴ in 1989 for those tumours in which a pure sarcoma, similar to or of a higher grade than that of the underlying adenosarcoma accounts for atleast one-quarter of the tumour after thorough histological sampling. Microscopically, these tumours are composed of an intimate admixture of a bland or atypical glandular component and a sarcomatous stromal component along with a pure sarcoma comprising more than 25% of the tumour, the latter may be more poorly differentiated and of higher grade or of the same grade as the sarcomatous stroma of the associated adenosarcoma.

Seidman et al⁵, in their study have noted that tumours with sarcomatous overgrowth occurred in younger women, were larger and were more likely to have heterologous elements as compared with those without sarcomatous overgrowth.

The main differential diagnosis of MASO is typical AS with stromal predominant areas. Hence, Clement recommends that the diagnosis of MASO be rendered only when a pure

sarcoma accounts for atleast one fourth of the tumour after thorough histological sampling of the hysterectomy specimen. Correct classification of mullerian adenosarcoma with sarcomatous overgrowth is important because the limited available data suggest that the prognosis is notably worse than that for adenosarcomas without sarcomatous overgrowth⁵. Further population based studies comparing MASO with other uterine sarcomas may be of value to quantify differences in prognosis if any. The only feature associated with an increased risk of recurrence or metastasis in these tumours is the presence of myometrial invasion³. However, this feature was not seen in the present case.

The management of patient with MASO is similar to that of a highly malignant uterine sarcoma. Patients with MASO should therefore receive long-term post-operative

surveillance in view of the likelihood of late recurrence.

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