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Uterine Mullerian Adenosarcoma With Sarcomatous Overgrowth Showing Rhabdo Myoblastic Differentiation In A Post Menopausal Patient - A Rare Interesting Case Report.

Anuradha S*a Bharathi Ka

^aDepartment Of Pathology, Sri Satya Sai Medical College and Research Institute, Tamilnadu, India

A 65 year old lady presented with postmenopausal bleeding of 1 month duration and abdominal pain. Ultrasonogram showed a bulky uterus. The patient underwent Total Abdominal Hysterectomy with Bilateral Salphingo Oophorectomy. We received the specimen of uterus with a polypoidal fleshy growth occupying the endometrial cavity. On examining the tissue sections microscopically it was diagnosed as " Uterine Mullerian Adenosarcoma With Sarcomatous Overgrowth Showing Rhabdomyoblast Like **Differentiation.**" This is an uncommon uterine tumour of higher grade with an admixture of benign epithelial and malignant stromal elements. It is regarded as a variant of Malignant Mixed Mullerian Tumour(MMMT). Presence of sarcomatous overgrowth predicts its poor prognosis because of the association with deep myometrial invasion and distant metastasis. Presence of heterologous elements like rhabdomyoblast like differentiation does not have any influence on its prognosis. After the surgical treatment, close follow up is needed to look for post operative recurrence.

KEY WORDS: Postmenopausal Bleeding, Mullerian Adeno Sarcoma, Sarcomatous Overgrowth, Rhabdomyoblastic Differentiation.

ABBREVIATIONS: MMMT - Malignant Mixed Mullerian Tumor; MIB-1 - An antibody for Ki- 67 nuclear antigen for estimating the Mitotic Index; ER- Estrogen Receptor; PR- Progesterone Receptor; CD - Cluster Differentiation; H&E- Hematoxylin And Eosin Staining.

INTRODUCTION

A bulky polypoidal growth filling the endometrial cavity in a post menopausal elderly woman is generally thought as endometrial carcinoma but here we are presenting an uncommon case of 'Uterine Mullerian Adenosarcoma with Sarcomatous Overgrowth showing Rhabdomyoblast like differentiation' a high grade uterine tumour with an admixture of benign epithelial & malignant stromal elements[1]. Presence of stromal overgrowth, deep myometrial invasion and distant metastasis indicates the aggressive nature of the tumour as will be described and discussed.

CASE REPORT

A 65 year old lady came with complaints of abdominal pain & low backache and occasional post menopausal vaginal bleeding. Ultrasound showed a bulky uterus of size 10x8x5 cm. Total Abdominal Hysterectomy with bilateral Salphingo Oophorectomy was done.

*Corresponding author: Dr. S. Anuradha, Assoc. Prof. Pathology, Sri Satya Sai Medical College and Institute, E-mail: Research Tamil nadu. sanudoc50@vahoo.co.in

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Fig 1: Shows the uterus with an irregular polypoid friable growth of size 6x5x4 cm in the endometrial cavity with areas of hemorrhage and necrosis.

We received the specimen in Department Of Pathology, Shri Satya Sai Medical College & Research Institute. Grossly uterus showed an irregular, bulky, spongy, polypoidal growth occupying the endometrial cavity measuring 6x5x4 cm [fig 1] with cut surface showing cystic spaces and areas of hemorrhage and necrosis. Multiple Sections studied microscopically showed a neoplasm with fascicles of spindle to plump atypical endometrial stromal cells [fig 4 & 5] with mitosis > 4 per 10 high power field along with many benign glands filled with secretions [fig 2].

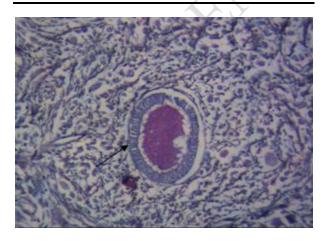


Fig 2: Shows the neoplasm with benign endometrioid gland surrounded by neoplastic mesenchymal cells.[H&E stain, 10x10 X]

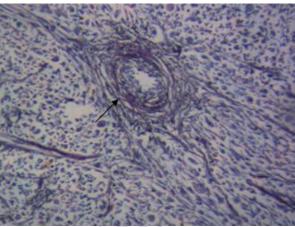


Fig 3: Shows the perivascular lobular arrangement of Rhabdomyoblast like mesenchymal cells. [H&E stain , $10x10\ X$]

Neoplasm also showed extensive areas (>80% of the tumour) of Rhabdomyoblast like cells which are large cells with abundant eosinophilic cytoplasm [fig 3 & 6]. The neoplasm showed deep myometrial and vascular invasion.

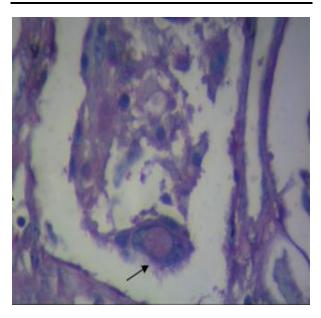


Fig 4: Shows a high power view of a binucleated bizarre stromal cell. [H&E stain, 40x10 X]

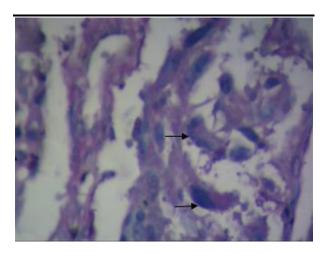
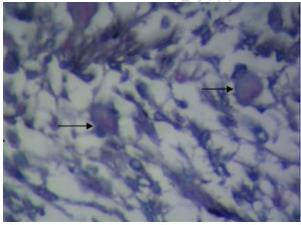


Fig 5: Shows the high power view of the atypical stromal cells. [H&E stain, 40x10 X]

DISCUSSION

Uterine Mullerian Adenosarcoma is a rare biphasic tumour with benign glands regularly distributed in a cellular mesenchyme.[2] It is a low grade malignant tumour unless it has a stromal overgrowth. It usually arises in the uterine corpus but it can also occur in the endocervix, vagina, ovary and pelvis. They generally occur in the post menopausal age group.[3] The tumour has a polypoidal friable growth pattern with a microscopic appearance of a phyllodes tumour.[9] adenosarcomas contain homologous Most mesenchymal elements composed of tissue types like fibroblasts and endometrial stromal cells which are the



Shows the high power view Rhabdomyoblasts like cells with abundant eosinophilic cytoplasm. [H&E stain, 40x10 X]

normal component of uterine tissue. The special feature of Mullerian adenosarcoma is that the stromal component very rarely shows heterologous elements skeletal muscle (ie) Rhabdomyoblastic differentiation in 20% of cases [4] The stroma can also differentiate into sex cord elements, smooth and angio sarcomatous muscle differentiation differentiation.[4][5][6] These heterologous elements does not have any influence on the overall prognosis of the patients. Occasionally there can be a marked decidualisation of the stroma due to hormonal usage.[9]

Adenosarcoma in which more than 25% of the tumour shows pure sarcomatous change is termed as "Uterine Mullerian Adenosarcoma With Sarcomatous Overgrowth" [9] It is very aggressive in nature associated with myometrial invasion and metastasis. In such cases recurrence can be seen in the vagina and pelvis in 25% of cases.

Eminent pathologists say that Adeno Sarcoma is not a type of Malignant Mixed Mullerian Tumour(MMMT) but rather as a variant of Endometrial Stromal Sarcoma having the capacity to differentiate into glands. Uterine Mullerian Adenosarcoma is known to be associated with Tamoxifen therapy in breast cancer[7], Stein Leventhal Syndrome and pelvic irradiation. In cases of sarcomatous overgrowth, Immunohistochemical reactions show high MIB-1 index and the loss of ER,PR & CD 10 + ve.[8] Adenosarcoma of ovaries are more aggressive than uterus due to lack of anatomic barrier to prevent the peritoneal dissemination.[9] The closest differential diagnosis for this tumour are Adeno Fibroma, Atypical Polypoid Adenomyoma, Malignant Mixed Mullerian Tumour and High Grade Endometrial Stromal Sarcoma.[3]

CONCLUSION

This case is presented for its rarity and to create an awareness that a case of post menopausal bleeding and a mass in the endometrial cavity can also be due to Uterine Mullerian Adenosarcoma. Presence Sarcomatous Overgrowth is associated with high rate of post operative recurrence and it is important to recognize it for a close follow up after surgical treatment.

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