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Endometrial Serous Adenocarcinoma and Perineal Leiomyosarcoma: A rare case report of Synchronous Primaries

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ABSTRACT

Background : The simultaneous presentation of endometrial carcinoma and perineal leiomyosarcoma in a patient as a synchronous tumor is very rare.

Case presentation : We describe a case of a 70-year-old postmenopausal lady who presented with post menopausal bleeding. On MR imaging, the patient had multiple irregular nodular polypoidal lesions arising from the endometrium along with another well-defined lobulated and encapsulated lesion in the perineum. Patient then underwent hysterectomy followed by histopathological examination which revealed coexisting serous type endometrial carcinoma and perineal leiomyosarcoma.

Conclusions : This case report describes this uncommon and interesting association of two rare entities with imaging features and histopathology. Despite its rarity, synchronous primary tumors in the female genital tract exist, and diagnosis and staging of both tumors preoperatively are largely beneficial

KEYWORDS : *Endometrial carcinoma, postmenopausal, perineal leiomyosarcoma, synchronous, Case report*

INTRODUCTION

Endometrial carcinoma is the most common gynecological malignancy worldwide and the second most common in developing countries, including India, with 4.3 per 100000 lakh Indian women [1]. Type I endometrial cancer is the most common (90%) and is associated with a goo prognosis, whereas type II is less common and has a poor prognosis [2]. Type II endometrial cancer, also known as non-endometrioid carcinoma, includes clear cell carcinoma, serous papillary carcinoma, and carcinosarcoma. They occur in older postmenopausal women, are non- estrogen-

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dependent, usually higher grade, and carry poor prognoses than type I cancers. On the other hand, leiomyosarcoma is a malignant tumor of a smooth muscle cell that presents as a painless, non-tender mass, usually in the 5th or sixth decades. It usually arises from the gastrointestinal tract, uterus, and retroperitoneum; however, it can rarely involve the rectum, vulva, vagina, and perineum. Perineal location as a primary site is infrequent, with only a few cases reported in the literature. Perineal leiomyosarcomas are extremely rare and aggressive cancer with high metastatic potential and no defined standard treatment [3]. Furthermore, the coexistence of endometrial carcinoma and perineal leiomyosarcoma is even rarer. However, endometrial carcinoma can sometimes coexist with leiomyoma. Here we report a unique and challenging case about the unusual simultaneous presentation of serous carcinoma of endometrium and perineal leiomyosarcoma in a postmenopausal patient, which is not reported to the best of our knowledge in the literature

CASE REPORTS

A 70-year-old postmenopausal lady with complaints of postmenopausal bleeding and slow- growing painless mass in the perineum for one year presented to the gynecology department of our hospital. The patient was neither overweight nor had a history of intake of any hormone replacement therapy. However, she had intramural fibroids. In addition, her serum tumor biomarkers were unremarkable, including CA125, CA199, AFP (alpha-fetoprotein), and CEA (carcinoembryonic antigen). The patient was referred to the radiology department for contrast-enhanced magnetic resonance imaging (CE-MRI) pelvis, which revealed a bulky, anteverted uterus, measuring 11.9 cm (CC) x 12.9 cm (AP) x 13.7 cm (Tr). There was the presence of two large well-encapsulated lesions seen in both anterior and posterior myometrium measuring 5.7 cm (CC) x 5.2 cm (Tr) x 3 cm (AP) and 8.2 cm (CC) x 5.8 cm (Tr) x 6cm (AP) respectively and compressing the surrounding myometrium with the formation of a pseudo capsule. The lesions appeared predominantly hypointense on both T1 and T2 weighted images with few areas of altered signal in the stroma, appearing intermediate on T1 and T2 weighted imaging with mild enhancement on postcontrast images suggestive of myxoid change. These findings were s/o intramural leiomyomas with areas of myxoid degeneration. The endometrial cavity was markedly distended with fluid intensity contents within, with evidence of multiple irregular nodular polypoidal lesions arising from the endometrium and projecting into the distended endometrial cavity. The largest lesion measuring ~ 3.5 cm (Tr) x 3 cm (CC) x 2.8 cm (AP) was seen arising from the anterior endometrial lining. These nodular lesions appeared isointense on both T1 and T2WI showing restricted diffusion on diffusion-weighted imaging (DWI) and marked enhancement on postcontrast images. Some of them extended into the adjacent myometrium, maximum for a depth of ~ 5.2 mm. No evidence of pelvic and para-aortic lymphadenopathy was seen (Figure 1). There was another well-defined lobulated and encapsulated lesion in the perineum. It appeared intermediate on T1WI and heterogeneously hyperintense on T2WI and showed diffusion restriction on DWI and heterogeneous contrast enhancement. The lesion involved the vaginal vault on the left side anterolaterally, abutting the rectum and anal canal posteriorly and the external sphincter on the left side without any apparent invasion and inferiorly reaching up to the skin surface without any skin ulceration(Figure 2). Based on these findings, a presumptive diagnosis of endometrial stromal sarcoma with perineal metastases was given, and a histopathological correlation was advised. The patient underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy. Resected specimens and biopsy from the perineal lesion were sent for histopathological analysis (Figure 3). The pathological assessment revealed two intramural leiomyomas. Sections from the endometrial lesions revealed highly atypical cells with high mitotic index and arranged as glands with lymphovascular invasion. It showed positivity for CK (cytokeratin), EMA (epithelial membrane antigen), and P53. No evidence of cervical invasion was seen. No abnormalities were detected in the bilateral adnexa. These findings were suggestive of high-grade adenocarcinoma of serous type (Figure 4). Biopsy from the perineal lesion revealed the tissue composed of bundles and sheets of spindle cells with eosinophilic fibrillary cytoplasm with high mitotic index and positivity for SMA (smooth muscle actin). It was negative for CK, EMA, and Desmin. Tumour was also seen to infiltrate into the adjacent adipose tissue. Findings were suggestive of perineal leiomyosarcoma (Figure 5).

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Figure 1(A to F) : T1W axial (A), T2w axial (B) and T2w sagittal (C) MRI pelvis images reveal two large intramural fibroids with myxoid degeneration(white Asterix). There is also presence of multiple irregular nodular polypoidal lesions (white arrows)seen arising from the endometrium and projecting into the distended endometrial cavity, largest measuring ~ 3.5 cm (Tr) x 3 cm (CC) x 2.8 cm (AP) seen arising from anterior endometrial lining. These nodular lesions appear isointense on both T1 and T2WI showing restricted diffusion (D and E) and enhancementon post contrast images (F)

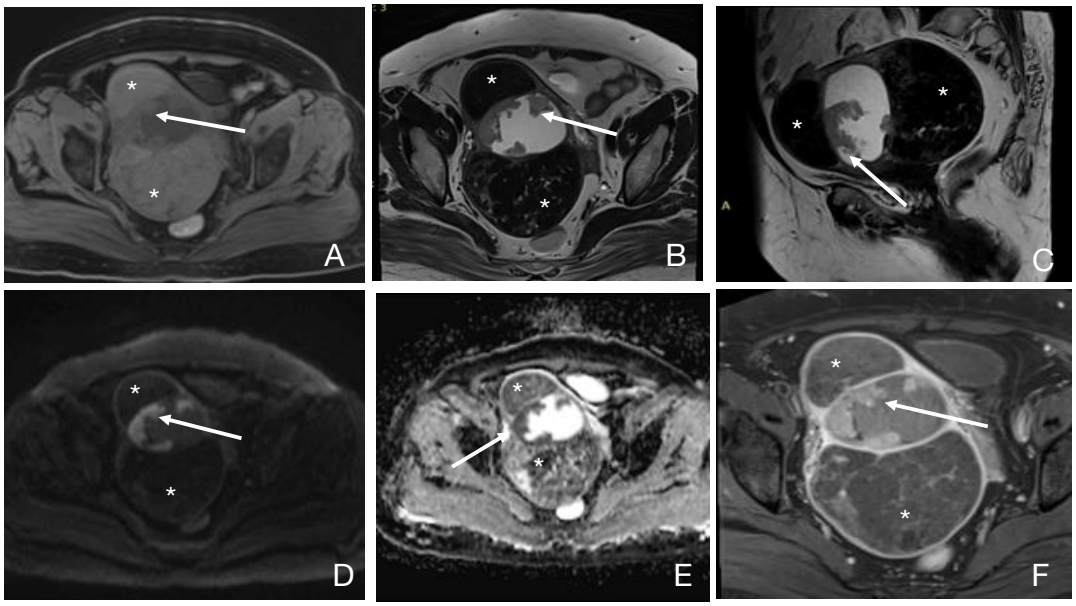

Figure 2 (A to F) : T1w axial (A), T2w axial (B) and T2w sagittal (C) MRI pelvis images further reveal well defined lobulated and encapsulated lesion seen in the perineum appearing intermediate intensity on T1w and heterogeneously hyperintense on T2w (white arrows) which shows restricted diffusion (D and E) and marked contrast enhancement (F). Anterolaterally, the lesion is involving the vaginal vault on the left side, posteriorly abutting the rectum, anal canal and external sphincter on the left side without any obvious invasion and inferiorly reaching up to the skin surface without any skin ulceration.

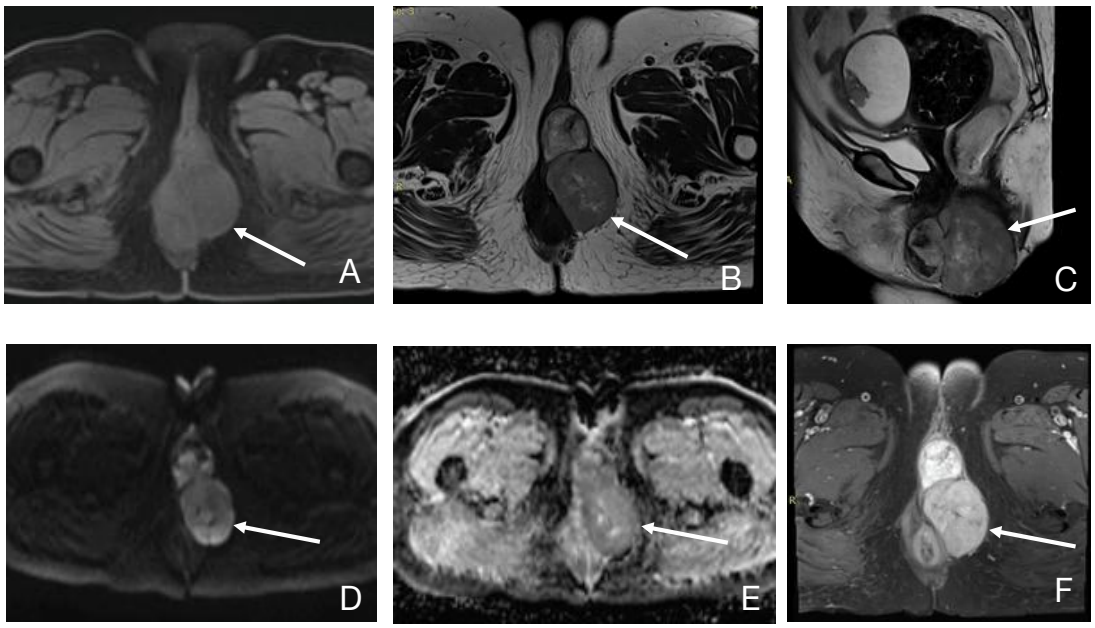

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Figure 3 (A to D) : Post Hysterectomy gross specimen : Post hysterectomy gross specimen of the uterus with bilateral adnexa shows two large intramural fibroids (white asterix in A and black arrows in B and C). On cut open of the endometrial cavity, there is presence of whitish friable growth measuring ~ 5 x 2.5 x 1 cm in the endometrial cavity.

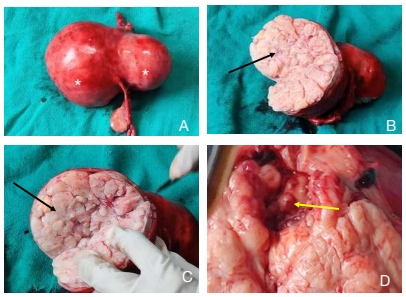


Figure 4 : Photomicrograph from the histopathological examination of growth in endometrial cavity shows the tumour to be composed of highly atypical cells having clear to pale eosinophilic cytoplasm, large nucleoli and coarse chromatin.The tumour show papillary architecture with tumour cells arranged as tubules and glands with foci lymphovascular invasion. Findings s/o serous adenocarcinoma (high grade).

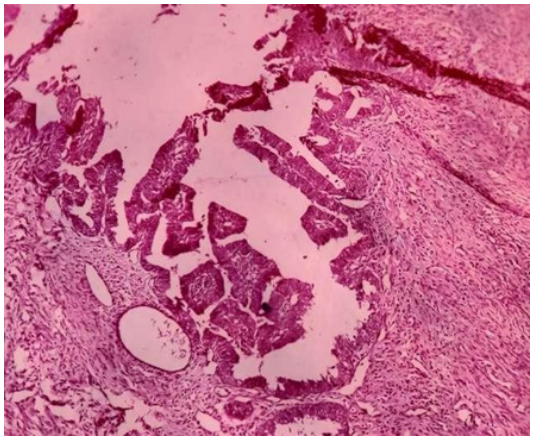
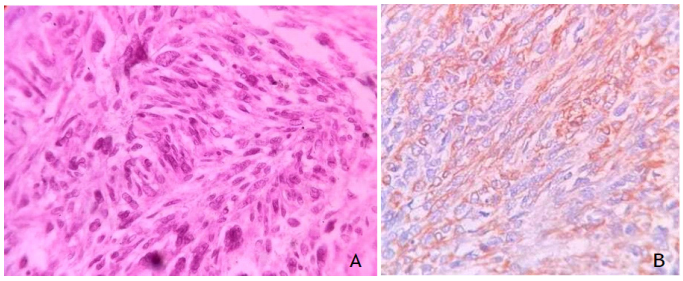


Figure 5 : Photomicrograph of histopathological section from the perineal lesion show tumour to be arranged in fascicles. Individual tumour cells are oval to spindle shaped showing pleomorphism, fibrillary cytoplasm, nucleus is cigar shaped and shows atypia(A). On IHC, tumour cells are positive for and show positivity for SMA (B).



DISCUSSION

A comprehensive literature search for associations of other primary tumors with endometrial carcinoma revealed that such associations had been described in multiple population-based studies. [4] According to one such study, the incidence of other primary malignancies in endometrial carcinoma is around 22%, with the most frequent associations being colorectal, ovarian, and breast cancers [5]. However, to the best of our knowledge and as per the extensive literature search, this is the first case report about the coexistence of serous endometrial adenocarcinoma (ESC) with perineal leiomyosarcoma, which is an extremely rare entity with no case reported in the literature. In the year 1983, Bokhman [6] first described a histological classification of endometrial carcinoma based on clinical and histological features into two types: type 1(endometrioid adenocarcinoma), which are estrogen-dependent, lower grade, and has a good prognosis, and type 2 (non-endometrioid adenocarcinoma) are non-estrogen dependent, occur in older females, poorly differentiated and has a poor prognosis. Type II cancers include clear cell carcinoma, serous papillary carcinoma, and carcinosarcoma. Type II cancers are characterized by p53, HER- 2/neu, p16, and E-cadherin [7]. Endometrial serous carcinomas, comprise approximately 10% of endometrial carcinomas. This tumor type constitutes 40% of all deaths and recurrence associated with endometrial cancer [2]. However, this differentiation cannot be made on imaging modalities, and imaging mainly plays a role in evaluating disease extent and staging. [8] Our patient was nonobese and had no history of estrogen exposure. The serous endometrial

adenocarcinoma is a clinically aggressive tumor. It typically occurs in the atrophic endometrium of postmenopausal women. It has extra-uterine spread and a high risk for recurrence and metastasis. Its association with leiomyomas has been described in the literature [1].

On the other hand, perineal leiomyosarcoma is rare, with only a few cases reported in the literature. It is an extremely rare and aggressive cancer with high metastatic potential and no defined standard treatment. It also tends to affect women more commonly than men in the 5th and sixth decades of life and presents as a slow-growing, non-tender mass. [3,9] Most of the time, their diagnosis is delayed as they are unexpected. Computed tomography (CT) and magnetic resonance imaging (MRI) shows the lesion's extent and help in planning management. Histopathological examination confirms the final diagnosis. Pathologically, it shows positivity for vimentin, desmin, and smooth muscle actin (SMA) [3]. The mainstay of treatment for perineal leiomyosarcomas is aggressive surgical resection. Our patient had a similar presentation as a painless perineal mass, which had a nonspecific appearance on imaging. In the presence of suspected primary endometrial malignancy, a provisional diagnosis of perineal metastases was given. In the case of primary endometrial malignancy, the differentiation between metastases and a synchronous primary tumor is not easy. Furthermore, the imaging appearances are often nonspecific in metastases and other perineal primary malignancies, including leiomyosarcoma. Moreover, such an association of these two rare tumors simultaneously was not previously described. Histopathology with immunological markers revealed the final diagnoses of high-grade serous endometrial carcinoma and perineal leiomyosarcoma.

CONCLUSION

Imaging features help in the detection of such rare lesions, show their extent, and help in planning management. Histopathology with immunological markers confirms the diagnosis in the case of two synchronous primary tumors. Our case reports add helpful information to the literature so that a possibility of such association can be considered while reporting so that appropriate referral and management can be done. Despite its rarity, synchronous primary tumors in the female genital tract exist, and diagnosis and staging of both tumors preoperatively are largely beneficial.

LIST OF ABBREVIATIONS

AFP - Alpha-fetoprotein
CEA - Carcinoembryonic antigen
CE-MRI- Contrast-enhanced magnetic resonance imaging
Tr- Transverse
CC- Craniocaudal
AP- Anterposterior
T1WI- T1 weighted imaging
T2WI- T2 weighted imaging
DWI- diffusion-weighted imaging
CK- Cytokeratin
EMA- Epithelial membrane antigen
SMA - Smooth muscle actin
CT- Computed tomography
MRI- Magnetic resonance imaging
ESC- Serous endometrial adenocarcinoma

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