

Primary Endometrial Squamous Cell Carcinoma in-situ with Extensive Ichthyosis Uteri: A Rare Case Report

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ABSTRACT

Primary squamous cell carcinoma of the endometrium is a rare entity with primary endometrial squamous cell carcinoma in-situ being more uncommon. We report a 60-year-old multiparous post-menopausal woman who presented with a lower abdominal swelling alongwith difficulty in urination since five months. Total abdominal hysterectomy with bilateral salpingo-oophorectomy showed an enlarged uterus with pyometra. A diagnosis of primary squamous cell carcinoma in-situ of the endometrium was made on histopathology.

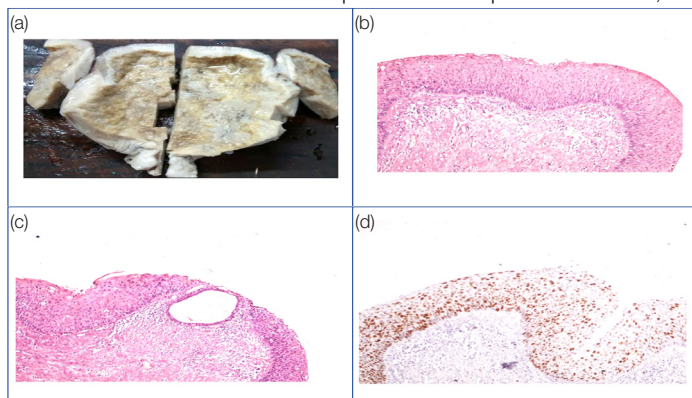
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CASE REPORT

A 60-year-old multiparous post-menopausal woman presented with lower abdominal swelling and urinary discomfort since five months. There was no history of vaginal bleeding and the patient had been post-menopausal for 15 years. Abdominal examination revealed a soft, nontender lump corresponding to the size of 24 weeks pregnant uterus. A speculum examination showed cervical stenosis. Pelvic ultrasound showed an enlarged uterus with accumulation of around one litre fluid within the endometrial cavity. A Papanicolaou smear of the cervix was negative for intraepithelial lesions or malignancy. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. One litre of pus was drained from uterus per operatively.

On gross examination, the uterus was enlarged with a thinned out cervix with focal areas of ulceration. There was no evidence of any cervical growth. Endometrial cavity was distended with an irregular, shaggy, grey yellow endometrial surface. Myometrium and bilateral adnexae were unremarkable [Table/Fig-1a]. Almost entire endometrium was replaced by stratified squamous epithelium with extensive areas of moderate dysplasia and carcinoma in-situ with presence of occasional atrophic endometrial glands covered by areas of ulceration [Table/Fig-1b,c].

Underlying myometrium was unremarkable. Cervix showed extensive areas of ulceration with focal squamous metaplasia. However, no



[Table/Fig-1]: a) Hysterectomy specimen irregular, shaggy, grey yellow endometrial surface; b) Extensive replacement of endometrial lining by sheets of squamous cells with dysplasia on left (H&E 10X); c) Endometrial squamous cell carcinoma in situ with occasional atrophic endometrial gland (H&E 10X); d) Immunohistochemistry showing high Ki-67 labelling index (IHC 10X).

evidence of dysplasia was seen on processing the entire cervix. Also, there was no evidence of malignant endometrial gland on extensive sectioning. Based on these histopathology findings and a high Ki-67 labelling index [Table/Fig-1d] on immunohistochemistry, a final diagnosis of ichthyosis uteri with extensive dysplasia and foci of primary endometrial squamous cell carcinoma in situ was given. Patient is disease free, three months after surgery with negative Papanicolaou smear of vaginal vault.

DISCUSSION

Primary Endometrial Squamous Cell Carcinomas (PESCC) have been found in association with pyometra, cervical stenosis, chronic inflammation, multiparity and ichthyosis uteri in post-menopausal women. An ichthyosis uterus is a rare condition of replacement of the entire endometrial surface by stratified squamous epithelium [1]. PESCC can arise from endometrial stem cell, squamous metaplasia of the normal endometrium or from heterotopic cervical tissue [2]. The presence of squamous epithelium in the endometrium is termed as ichthyosis uteri, leukoplakia, epidermalization psoriasis uteri, epidermoid heteroplasia, cholesterometra, acanthosis and indirect regenerative squamous metaplasia [2].

Chronic irritation due to long standing pyometra may lead to the malignant change in ichthyosis uteri as seen in the present case. Primary endometrial squamous cell carcinoma in-situ arising in ichthyosis uteri is a very rare entity with a very few case reports in the literature. To the best of our knowledge, only six cases of PESCC in situ have been reported to date [3-8]. Pailoor K et al., and Jetley S et al., had described similar rare case of PESCC in-situ with extensive ichthyosis uteri in a post-menopausal woman with pyometra [3,4].

To reach a diagnosis of PESCC, it is important to exclude cervical SCC extension into the endometrium and squamous differentiation of an endometrioid adenocarcinoma. PESCC can be differentiated from endometrial SCC involvement by pathological criteria recommended by Fluhmann: (1) no evidence of a co-existing endometrial adenocarcinoma or primary cervical SCC; (2) no connection between the endometrial tumour and squamous epithelium of the cervix; (3) no connection between any in-situ carcinoma of cervix and endometrial neoplasm [9]. Our present case fulfilled Fluhmann's criteria with presence of changes of dysplasia and carcinoma in-situ without any signs of an invasive carcinoma even on extensive sampling.

A high Ki-67 labelling index has also been reported, explaining the aggressive and malignant nature of the lesion. The prognosis for patients with endometrial SCC depends on the stage of the tumour. PESCC shows a poorer prognosis than endometrioid carcinomas and is managed by surgical hysterectomy with salpingo-oophorectomy and radiotherapy [9].

CONCLUSION

Primary squamous cell carcinoma of endometrium although rare, should be considered as a diagnosis while evaluating a post-menopausal elderly females presenting with pyometra. Early and prompt diagnosis is important to improve the survival rate.

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