



Ichthyosis Uteri with Primary Endometrial Squamous Cell Carcinoma in Situ- An Incidental Finding

Rashmi Monteiro, Shashi Sujanani, Preeti Agarwal*

Department of Pathology, Pacific Medical College and Hospital
Udaipur, Rajasthan, India

***Corresponding Author:**

Preeti Agarwal

Department of Pathology, Pacific Medical College and Hospital
Udaipur, Rajasthan, India

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ABSTRACT

Primary squamous cell carcinoma in situ of the endometrium is an extremely rare condition. It may be associated with pyometra. We report a case of a 71 year old, postmenopausal woman who presented with burning micturition, increased urine frequency and lower abdominal distention. Radiological findings were suggestive of pyometra or hematometra of the uterus. Hysterectomy was performed and histopathological examination was done, which was suggestive of primary squamous cell carcinoma in situ of the endometrium

Keywords: ichthyosis uteri, endometrium, squamous cell carcinoma, pyometra

INTRODUCTION

Pyometra if occurring in elderly women has a great risk of mortality and morbidity. Pyometra is a condition where there is collection of a lot of purulent material in the uterine cavity.¹⁻⁵ Primary squamous cell carcinoma of the endometrium is a rare condition and squamous cell carcinoma in situ of the endometrium is more uncommon condition and both can be associated with pyometra. There are very few cases of primary squamous cell carcinoma of the endometrium with ichthyosis uteri and to the best of our knowledge only six cases have been reported in literature.⁶

We herein report a case of a 71 year old, post menopausal woman diagnosed to have pyometra and on histopathological examination was diagnosed with squamous cell carcinoma in situ of endometrium.

CASE REPORT

A 71 year old woman, para 3, came to a tertiary care hospital, to the Obstetric OPD, with complains of

burning micturition and increased frequency in urination since 20 days. She also complained of lower abdominal distention since 1 month. The patient attained menopause 30 years back and had a known history of hypertension since 3 years, for which she was on medications. The patient had no history of vaginal bleeding or fever and her total white blood cell count was normal (5800/cumm).

Abdominal examination showed a round non tender lump which corresponded to 14-15 weeks pregnant uterus. Per speculum examination revealed cervical stenosis. Transabdominal ultrasonography showed a ballooned and distended uterine cavity showing fluid collection with internal low level echoes in endometrial cavity and cervical canal, possibly due to cervical stenosis, suggestive of pyometra or hematometra. Pap smear of the cervix, taken from the patient was negative for intraepithelial lesions or malignancy. Patient was admitted and total abdominal hysterectomy with bilateral salphingo-

oophorectomy was performed. The post operative period was uneventful. The specimen was sent for histopathological examination.

On gross examination, the specimen consisted of an uterus with an ill-defined cervix and bilateral adenexa. Uterus with cervix measured 10 x 8 x 7cm. External surface of the uterus was flabby. On cutting open 800ml of pus was drained and the endometrial cavity was dilated and thinned out. Endomyometrial

thickness was 0.9cm. The endometrial mucosa was irregular shaggy and yellowish-white. Right tube measured 5cm in length and on cut surface lumen was patent, while the right ovary measured 2 x 0.5 x 0.5cm, cut surface was solid and grey-white. Left tube measured 7cm in length, cut surface lumen was patent and left ovary measured 2 x 0.5 x 0.5cm, cut surface was solid and grey white (Figure1)



Figure1: Hysterectomy specimen, external surface was flabby with an ill-defined cervix and cut surface, the endometrial cavity was dilated, thinned out, shaggy and yellowish white.

Microscopically, the cervical tissue (figure 2) showed ulceration, chronic cervicitis, multiple nabothian cysts and squamous metaplasia. No evidence of dysplasia was noted in the cervix. Sections studied from the endometrium showed that the entire mucosa was replaced by stratified squamous epithelium with extensive areas of carcinoma in situ changes (figure 3). Occasional atrophic endometrial glands were present with chronic inflammatory infiltrate and hemosiderin laden macrophages. No evidence of invasive carcinoma even on extensive sampling was

found. Myometrium showed monckeberg's sclerosis of vessels. Right and left tube showed squamous carcinoma in situ in the lumen and fallopian lining epithelium was displaced to the side (figure 4). No pathology was noted in right and left ovaries. Based on the histopathological examination the impression given was 'Ichthyosis Uteri with Diffuse Primary Endometrial Squamous Cell Carcinoma in Situ Extending into the Lumen of Fallopian Tubes Bilaterally'.

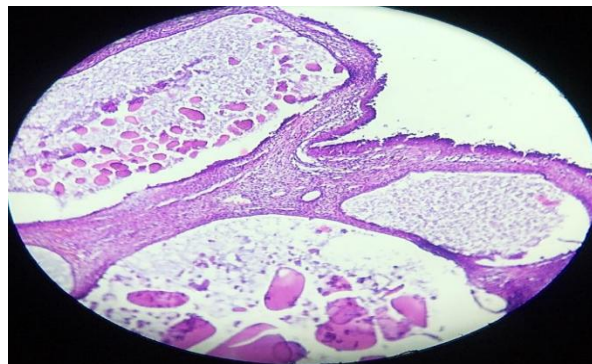


Figure 2: Cervix lined by stratified squamous epithelium with underlying tissue showing multiple Nabothian cysts.

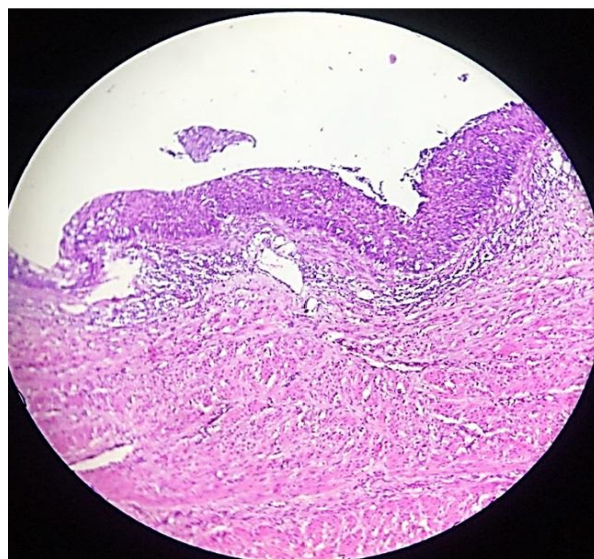


Figure 3: Endometrium showing squamous cell carcinoma in situ

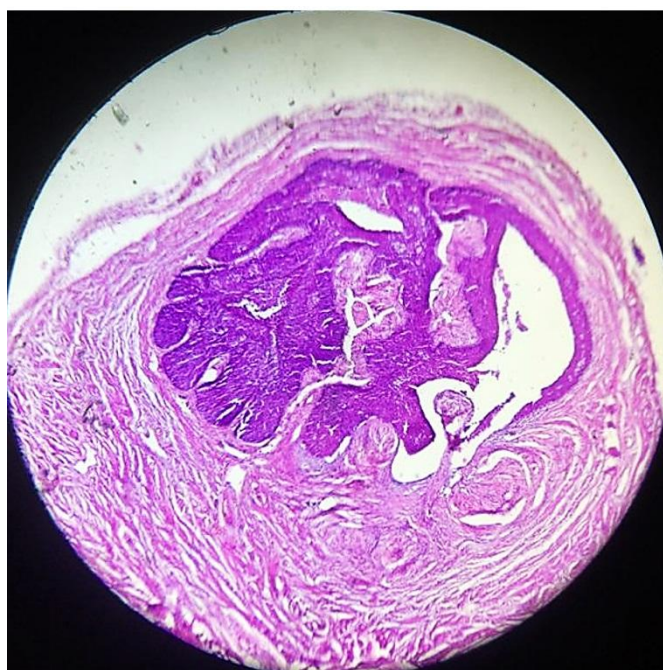


Figure 4: Fallopian tube showing squamous cell carcinoma in situ

DISCUSSION

Ichthyosis uteri is a rare condition in which the entire endometrial lining is replaced by stratified squamous epithelium.⁷ Primary endometrial squamous cell carcinoma is associated with pyometra, cervical stenosis, chronic inflammation, multiparity and ichthyosis uteri in post-menopausal women.⁶ Squamous cell carcinoma of the endometrium may arise from the stem cells of the endometrium, squamous metaplasia of the endometrial lining or from heterotopic cervical tissue.⁸ Long standing

pyometra may cause chronic irritation and may lead to malignant changes in ichthyosis uteri.⁶

Presenting symptoms of post menopausal women with pyometra are as follows- vaginal bleeding, vaginal discharge, abdominal pain and fever.¹⁻³ Absence of typical clinical symptoms were seen in our patient. Pyometra is a characterized by the accumulation of purulent material in the uterine cavity due to the alteration in the natural drainage. Diagnosis and prompt management of this condition are essential since there is significant risk of morbidity and mortality.¹ Factors that increase the

risk of pyometra are viral or bacterial cervicitis, radiation exposure, foreign bodies in the uterine cavity (e.g., intrauterine devices), exposure to chemical agents, estrogen treatment, vitamin A deficiency, and intracavitary tumors.¹ It is important to diagnose pyometra, since it can be the first sign of an underlying neoplasm. Underlying malignancies are the cause of this condition in 3.5-45% of all cases.²

Before the diagnosis of primary squamous cell carcinoma of endometrium is made, it is important to rule out squamous cell carcinoma of the cervix and squamous differentiation of endometrioid adenocarcinoma. Primary endometrial squamous cell carcinoma can be differentiated according to the criteria stated by Fluhmann- (1) there should be no evidence of coexisting endometrial adenocarcinoma or squamous cell carcinoma of the cervix, (2) there should be no connection between endometrial tumour and squamous lining of the cervix & (3) there should be no connection between in-situ carcinoma of cervix and endometrial neoplasm.⁹ The present case fulfilled all the Fluhmann's criterias without any evidence of tumor invasion.

Aggressive and malignant nature of the lesion also depends on the high Ki-67 labelling index. Primary endometrial squamous cell carcinoma has a poor prognosis as compared to endometrioid carcinoma. Prognosis is based on the stage of the carcinoma. Patient is usually treated surgically by performing hysterectomy with bilateral salphingo-oophorectomy and radiotherapy.⁹

CONCLUSION

Primary endometrial squamous cell carcinoma in situ even though is a rare condition should be considered as a differential diagnosis when a postmenopausal women presents with pyometra. Early diagnosis of this condition is important for a better prognosis and for increasing the survival rate of the patient.

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