



Primary squamous cell carcinoma of the endometrium with ichthyosis uteri - a case report.

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ABSTRACT

Ichthyosis uteri is a rare condition in which the entire surface of endometrium is replaced by stratified squamous epithelium. Primary squamous cell carcinoma of the endometrium (PSCCE) is still rare and its pathogenesis is unclear. Identification of PSCCE and its differentiation from endometrial involvement by squamous cell carcinoma of cervix & endometrioid carcinoma with massive squamous differentiation is essential for correct patient management and it is based on fluhmann criteria. We report a case of a 65-year-old postmenopausal woman satisfying the diagnostic criteria for PSCCE.

INTRODUCTION

The incidence of primary squamous cell carcinoma of the body of the uterus is extremely rare. In 1892, Gebhard reported the first case of PSCCE.[1,2] About 70 cases have been reported by World Health Organization till 2003.[3,4] Pubmed search revealed another 9 case reports of PSCCE in various journals from 2004 to 2011. Fluhmann (1928) formulated three criteria, which must be met before the diagnosis can be considered and are as follows.1) There should be no co-existing adenocarcinoma of the endometrium 2) There should be no demonstrable connection between the tumor in the endometrium and the squamous epithelium of the cervix and 3) There should be no co-existing primary squamous cell carcinoma of the cervix. A more recent criterion included by the World Health Organization is that the tumor must exhibit keratinisation and/or intercellular bridges. [2] The

present case satisfies both Fluhmann criteria & WHO criterion for primary endometrial squamous cell carcinoma.

CASE REPORT

A 65 year female presented with history of distention of abdomen & backache of 15 days duration. She was para 3, live 3 and had attained menopause 15 years back. Per speculum examination showed an atrophic cervix. Per vaginal examination revealed an enlarged uterus of 20 weeks size. Cervix was completely flushed with vagina. Ultrasonography revealed enlarged uterus with pyometra & thinning of myometrial wall. Cervix & adnexae were normal. The clinical diagnosis of pyometra secondary to cervical stenosis was made.

Endometrial cavity was drained by cervical dilatation. Curettage of uterine cavity with punch biopsy of cervix were sent for the histopathological

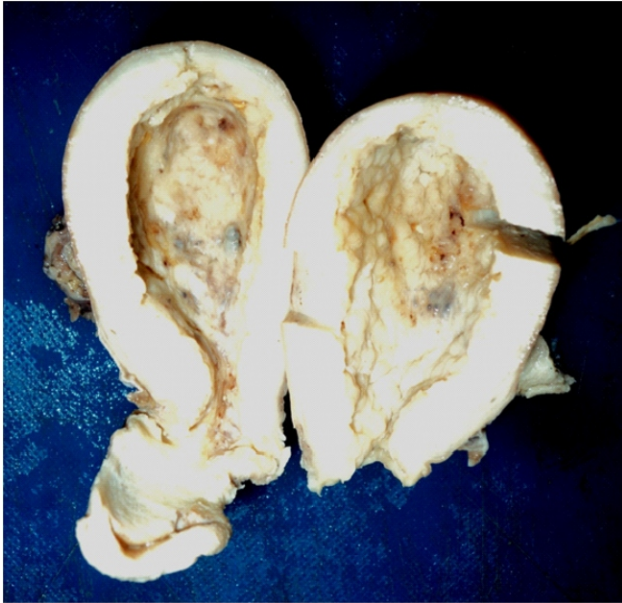


Figure 1: Dilated uterine cavity showing irregularly folded & thickened mucosal surface having small gray white nodular growth.

examination. Sections studied show only few fragments of stratified squamous epithelium showing carcinoma in situ. Cervical biopsy revealed a single tiny tumor deposit with stromal reaction. Origin of the tumor could not be precisely identified, as neither endometrial glands or stroma nor endocervical glands or epithelium were identified.

Surgery was carried out & specimen of radical hysterectomy with bilateral salphingo-oophorectomy including right & left pelvic lymphnodes were sent for the histopathological examination.

Pathology

Macroscopy:

Uterus, cervix with vaginal cuff measured 9x6x2 cm. with bilateral adnexae. On cut section uterine cavity was dilated showing irregularly folded & thickened mucosal surface having small gray white nodular growth which was friable involving the whole of the endometrial cavity & extending almost down up to the endocervical canal. Cervix appeared atrophied. No growth was seen in the cervix & vaginal cuff. (Fig 1) Bilateral ovaries, tubes and pelvic lymph nodes were unremarkable.

Microscopy:

Multiple serial sections studied from cervix showed only a focus of atrophied lining epithelium of stratified squamous epithelial cells. No endocervical lining & endo-cervical glands seen. Stroma was infiltrated with chronic inflammatory cell infiltration.

Multiple sections from endomyometrium showed shallow endometrium with replacement of the entire endometrium by metaplastic stratified squamous epithelium which showed transition from dysplasia through carcinoma in situ and area of micro invasion. Except a single endometrial gland seen in one section there were neither endometrial glands nor normal endometrial lining in other sections. Endometrial stroma was seen at focal areas and was unremarkable (Fig 2 & 3). There was no evidence of adenocarcinoma in the sections studied.

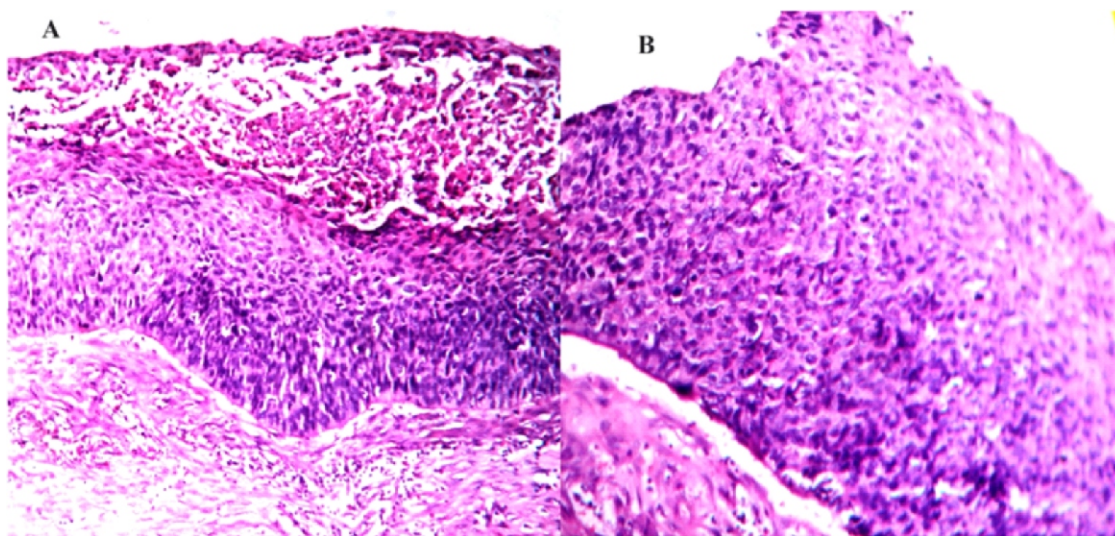


Figure 2: Photomicrographs of section from endo myometrium shows A) dysplasia (H& E X 200)with B) Carcinoma in situ (H& E X 400).

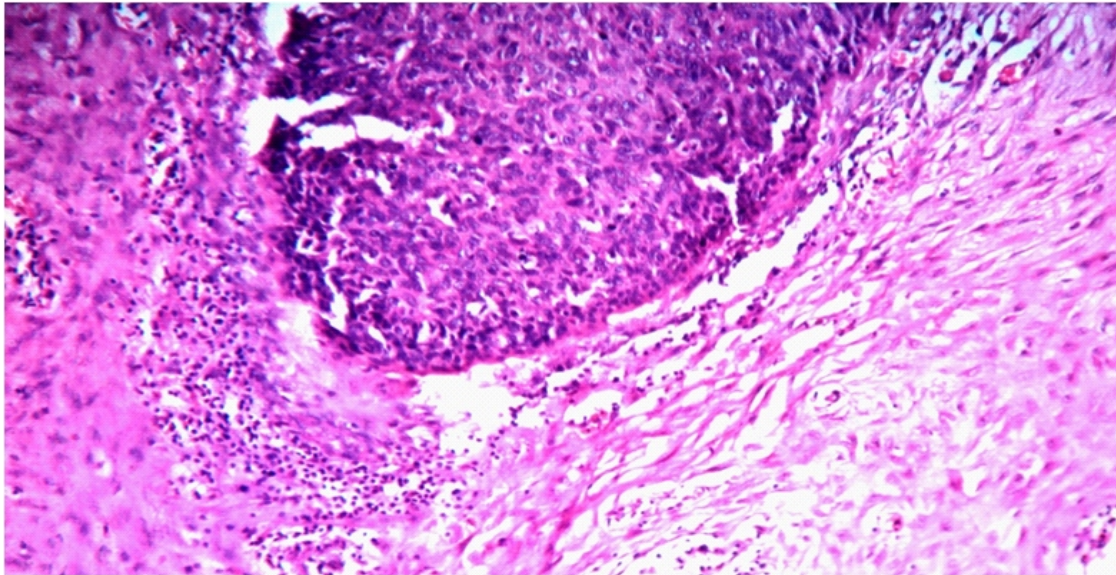


Figure 3: Photomicrograph of section from endo myometrium shows foci of microinvasion. (H & E X 400)

Histopathological examination of myometrium, vaginal cuff, bilateral fallopian tubes & ovaries were unremarkable.

Histopathological examination of pelvic lymph nodes showed features of follicular hyperplasia.

Hence, the diagnosis was given as **Micro invasive squamous cell carcinoma of endometrium with ichthyosis uteri.**

DISCUSSION

The term "ichthyosis uteri" was initially coined in 1885 by Zeller.[1] According to some authors ichthyosis lack malignant potential. However, dysplastic & anaplastic changes have been reported with few exceptional case reports describing the occurrence of PSCCE in the background of ichthyosis uteri.[5]

Primary squamous cell carcinoma of the endometrium usually occurs in postmenopausal women and is often associated with cervical stenosis and pyometra.[6] In a population-based study from Norway, the prevalence of PSCCE was found to be 0.1%.[7] After extensive sampling, this case was diagnosed as primary squamous cell carcinoma of the endometrium as the Fluhmann's (1928) criteria are satisfied. Our case also had ichthyosis uteri, so appeared as a sequel to such a generalized metaplastic process progressing to dysplasia, carcinoma in situ with microinvasion.

CONCLUSION

Although rare, diagnosis of primary squamous cell

carcinoma of endometrium should be considered in a postmenopausal elderly female presenting with pyometra and cervical stenosis. These needs to be correctly identified & should be differentiated from endometrioid carcinoma with massive squamous differentiation & secondary spread from cervical squamous cell carcinoma, because PSCCE are associated with an adverse prognosis and patients will require adjuvant treatment to reduce the risk of pelvic relapse and metastasis.[8]

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