Case Report

PURE PRIMARY SQUAMOUS CELL CARCINOMA OF THE ENDOMETRIUM WITH ISOLATED INSITU CERVICAL DYSPLASIA-A RARE ENTITY

Sabina Khan, Inara Abeer, Aruna Nigam, Sujata Jetley

Abstract:

Pure Primary Endometrial Squamous cell carcinoma (PESCC) is a rare entity with less than seventy cases reported till date. It is usually seen in postmenopausal females presenting with pyometra. Our patient was a 60 year old female presenting with postmenopausal bleeding since six months. Pap smear was reported as ASC-H (Atypical Squamous Cells-cannot exclude HSIL). Cervical biopsy showed features of Cervical Intraepithelial Neoplasia (CIN III). Endometrial curettings were suggestive of Squamous Cell Carcinoma, keratinizing type. Patient underwent radical hysterectomy following which a final diagnosis of Primary invasive Squamous cell carcinoma of uterus with isolated cervical dysplasia was made. Through this case report we wish to emphasize that although rare, diagnosis of PESCC endometrium should be considered in a postmenopausal elderly female presenting with pyometra. To the best of our knowledge, this is the first case report of endometrial squamous cell carcinoma coexistent with severe cervical primary dysplasia.

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Introduction

Primary pure squamous cell carcinoma of the endometrium (PESCC) is a very rare entity. Only about 70 cases have been reported in the world literature, according to WHO blue book.[1] PESCC is defined as a primary carcinoma of the endometrium composed of squamous cells of varying degree of differentiation. It is usually seen in postmenopausal females and shows association with pyometra. To be accepted as primary carcinoma of the endometrium it must satisfy the criteria established by Fluhmann and modified by Kay.[2] Few case reports of endometrial SCC have been sporadically published.[3] However, association of SCC of the endometrium with isolated cervical dysplasia is largely unknown.

CASE HISTORY:

A 60 year old postmenopausal female presented with pelvic pain and postmenopausal bleeding since 6 months. Per speculum examination revealed an unhealthy, congested cervix which bleeds on touch. Per vaginal examination revealed that cervix was firm, mobile, uterus was anteverted with bulky bilateral fornices. No obvious growth was seen. All routine investigations were within normal limits. On radiological examination, USG was suggestive of pyometra/hematometra. MRI showed enlarged uterus with endometrial cavity containing haemorrhagic fluid. Pap smear was reported as ASC-H (Atypical Squamous Cells-cannot exclude HSIL). Cervical biopsy showed features of CIN III. Endometrial curettings showed SCC, keratinizing type.

Patient was taken up for radical hysterectomy. On gross examination, uterine cavity showed endometrium irregularly lining the cavity. No solid growth was seen either in endometrium or cervix. Microscopic examination of the endometrium revealed extensive replacement of the entire endometrial lining by malignant squamous epithelium with tumour extending into the superficial myometrium in the region of fundus and body of the uterus. Serosa was free. Anterior cervical lip showed features of severe dysplasia but the isthmus was free from tumour.

A final diagnosis of Primary invasive Squamous cell carcinoma of uterus (pT1aN0) with isolated cervical dysplasia was made.

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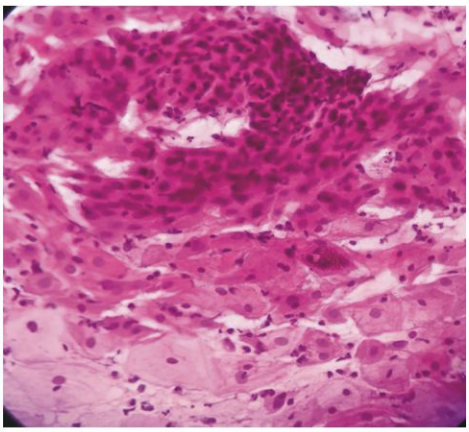

Fig 1.- Pap smear showing hyperchromatic crowded group of squamous epithelial cells without distinct cell borders (Pap stain,40X)

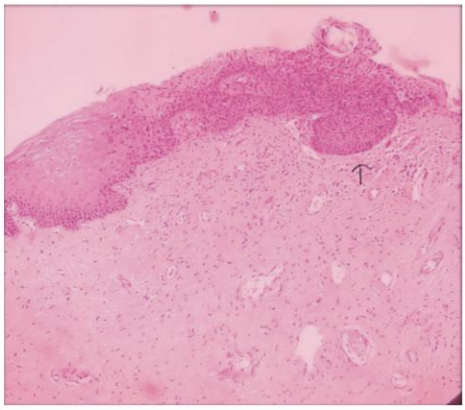

Fig 2- Cervical biopsy- Low power view showing focal CIN III changes (H& E stain, 10X)

Fig 3- Gross examination showing slightly irregular appearance of the endometrial cavity without any obvious growth.

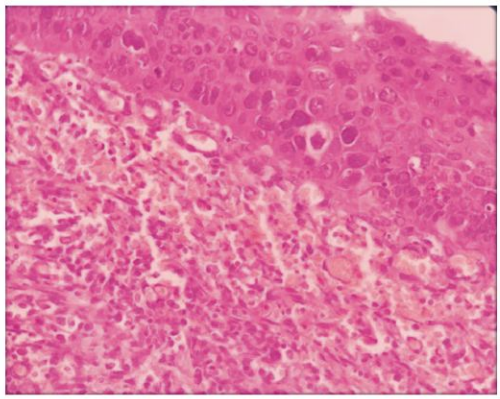

Fig 4- Microscopy shows endometrial lining completely replaced by malignant squamous epithelial cells.(H& E stain,40X)

DISCUSSION

Primary Endometrial squamous cell carcinoma is extremely rare and its exact frequency is largely unknown. The presence of squamous epithelium in the endometrium is also known as ichthyosis uteri. It is thought to arise by one of the following two mechanisms: upward spread of a primary cervical lesion or transformation of reserve or stem cells positioned between the glandular basement membrane and the endometrial columnar epithelium. Invasive squamous cell carcinoma as a result of upward spread from the cervix is very unlikely if the cervical lesion is intramucosal as was seen in our case. Primary SCC of the endometrium has been reported to show poorer prognosis than endometrioid carcinoma.[4,5] The prognosis depends mainly on tumor stage.

The pathogenesis of primary SCC of the endometrium is unknown. Several possibilities exist. First possibility is that the SCC is a complete malignant squamous differentiation of endometrioid adenocarcinoma. Secondly, HPV is involved in the pathogenesis of primary SCC of the endometrium.[6,7] Thirdly, squamous metaplasia-dysplasia-SCC sequence is involved in the pathogenesis of primary SCC of the endometrium.[8] Finally, primary SCC of the endometrium may develop from ectopic cervical tissue in the endometrium.[9] The preoperative diagnosis of endometrial squamous cell carcinoma may be difficult; since curettage specimens may show only highly differentiated squamous epithelium.

To be accepted as a primary squamous cell carcinoma of endometrium, the following criteria established by Fluhmann and modified by Kay must be statisfied. There must be no coexisting endometrial adenocarcinoma; there must be no connection between endometrial tumor and

squamous epithelium of cervix; there must be no squamous cell carcinoma of cervix; and if cervix shows an in situ carcinoma, there must be no connection between this and independent endometrial neoplasm. Our patient satisfied all these criteria.[10]

Endometrial squamous cell carcinoma usually occurs in postmenopausal women with mean age being 67 years. There is a strong association with cervical stenosis, pyometra, chronic inflammation and nulliparity which is believed to arise from ichthyosis uteri[11] Our patient was also a postmenopausal female presenting with pyometra.

The prognosis for patients with SCC depends mainly on the stage of the tumour. In a review of reported cases, 80% of stage I patients survived whereas survival for patients with stage III disease was only 20%.[5] Management for this condition usually consists of a hysterectomy with adnexectomy and radiotherapy in some cases.

CONCLUSION

Although rare, diagnosis of pure Primary Endometrial squamous cell carcinoma should be considered in postmenopausal elderly females presenting with pyometra. To the best of our knowledge this is the first case report of Pure Primary squamous cell carcinoma coexistent with cervical dysplasia.The strong relationship between tumour stage and survival suggests that early diagnosis and treatment are imperative in a case of Squamous cell carcinoma of Endometrium.

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