Case Report

Primary endometrial squamous cell carcinoma with extensive icthyosis uteri: A rare case report

Bhide S.P.[1], Jaison J.[2], Joshi S.R.[3]

[1]Dr. Smita P. Bhide, Professor, [2]Dr. Janice Jaison, Assistant Professor, [3]Dr. S.R. Joshi, Professor and HOD; all authors are attached with Department of Pathology, MIMER Medical College, Talegaon Dabhade, Pune, Maharashtra, India.

Corresponding Author: Dr Janice Jaison, Assistant Professor, Department of Pathology, MIMER Medical College, Talegaon, Dabhade, Pune, Maharashtra, India. E-mail: drjanicej@gmail.com

Abstract

Primary endometrial squamous cell carcinoma is an extremely rare tumour with less than 100 cases reported till date. It usually occurs in postmenopausal women and has strong association with pyometra, cervical stenosis, chronic inflammation and icthyosis uteri. We report a rare case of primary endometrial squamous cell carcinoma arising in Icthyosis uteri in a 60-year-old lady presenting with postmenopausal bleeding.

Keywords- Endometrium, Icthyosisuteri, Squamous cell carcinoma

Introduction

The presence of squamous epithelial lining in the endometrium, termed as icthyosis uteri, a condition in which the endometrial lining is replaced by keratinised squamous epithelium has been described in many conditions and is benign in most of the cases [1].

Primary endometrial squamous cell carcinoma arising in icthyosis uteri is very rare with only few cases reported in literature [2]. Here we report a rare case of primary endometrial squamous cell carcinoma arising in icthyosis uteri in a 60 year old lady presenting with postmenopausal bleeding.

Case Report

A 60 year old postmenopausal female, presented with bleeding per vaginum and foul smelling vaginal discharge since 6 months. She was 3rd para with uneventful obstetric history. She has been postmenopausal since 10 years.

When she was examined it was noted that there was an IUCD string present which was in place for 25 years.

The patient admitted that she had forgotten all about it. Per speculum examination showed atrophic cervix. Pap smear was taken which was negative for Intraepithelial lesion or malignancy.

D&C and cervical biopsy was done. D&C showed very few endometrial glands and atypical squamous cells.

Cervical biopsy was negative for intraepithelial or invasive malignancy and showed chonicnon specific cervicitis. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done and specimen was sent to Department of Pathology.

On gross examination, the uterus was enlarged, measuring 11x9x6 cm with atrophic & fibrotic cervix.No growth or ulcerative lesion was seen on cervix. Endometrial cavity was lined by irregular & shaggy endometrium.

There was an ill-defined mass of size 4x3x3cm seen invading more than half of the myometrial thickness. Bilateral adnexae were unremarkable.

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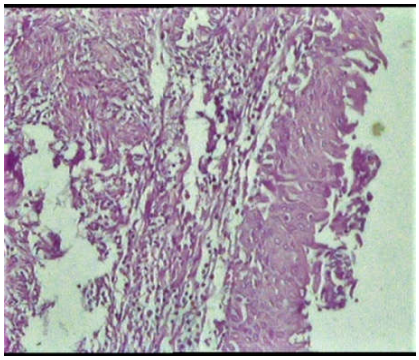

Fig No-1: Photomicrograph showing replacement of endometrial lining by stratified squamous lining epithelium (H&E 40X)

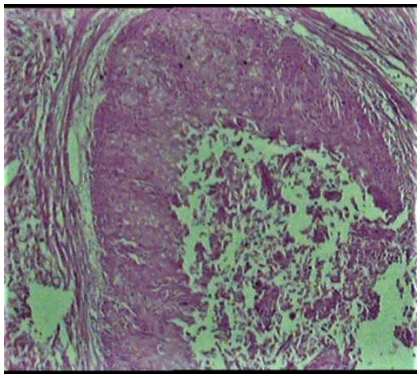

Fig No-2: Photomicrograph showing islands of squamous cell carcinoma invading the myometrium (H&E 10X)

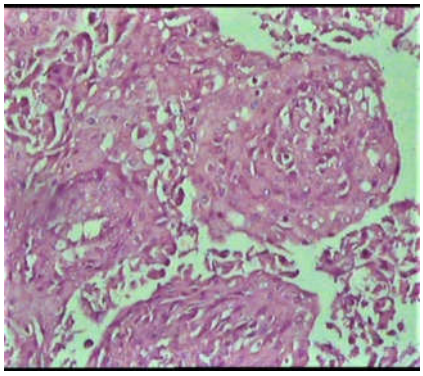

Fig No-3: Photomicrograph showing islands of invasive squamous cell carcinoma (H&E 40X)

Multiple sections studied from different parts of endometrium and corpus uteri showed extensive replacement of the entire endometrial lining by squamous epithelium which showed transition from dysplasia, carcinoma in situ to areas of frankly invasive keratinising squamous cell carcinoma invading the myometrium. Few normal endometrial glands and few endometrial glands showing squamous metaplasia were seen. However no focus of adenocarcinoma was found. Sections from the cervix were unremarkable.

There was no evidence of intra-epithelial or invasive squamous cell carcinoma in cervix. There was no cervical lesion continuous with that of uterus. Bilateral adnexae were unremarkable.

Hence the diagnosis of primary endometrial squamous cell carcinoma (PT1bNxMx FIGO stage at least IB) with extensive icthyosisuteri was made.

Discussion

Zeller in 1885 first described the term icthyosis uteri in which the entire surface of endometrium is replaced by stratified squamous epithelium following intrauterine iatrogenic introduction of substances like formalin or iodine [2]. It is usually associated with tuberculous endometritis, puerperal endometritis, endometrial polyps, endometrial hyperplasia, cervical stenosis & pyometra [3]. According to some authors, icthyosis uteri lacks malignant potential, However dysplastic and anaplastic changes have been reported with just few exceptional case reports describing the occurrence of primary endometrial squamous cell carcinoma in the background of icthyosis uteri [2].

Primary endometrial squamous cell carcinoma is extremely rare, with only 64 cases being reported in the literature. In 1996, Goodman et al reported 8 cases of endometrial squamous cell carcinoma & reviewed previously reported 56 cases of it [4].

To reach a diagnosis of primary endometrial squamous cell carcinoma it is important to exclude cervical squamous cell carcinoma extension into the endometrium & squamous differentiation of an endometrioid adenocarcinoma. To be accepted as Primary Squamous Cell Carcinoma of the endometrium the tumor must satisfy the criteria established by Fluhmann & modified by Kay.

1. There must be no coexisting endometrial adenocarcinoma.
2. There must be no connection between endometrial tumor & squamous epithelium of cervix.
3. There must be no primary squamous cell carcinoma of cervix.
4. If cervix shows an in situ carcinoma, there must be no connection between this & independent endometrial neoplasm [4, 5].

In the present case, cervix did not show in situ or invasive squamous cell carcinoma. Hence the possibility of direct extension from cervical squamous cell carcinoma was ruled out. Also there was no focus of endometrial adenocarcinoma in present tumor. Thus, the present case fulfilled all the above criteria of primary squamous cell carcinoma of Endometrium.

Primary squamous cell carcinoma of endometrium usually occurs in postmenopausal women (mean age 67years). The condition has been strongly associated with pyometra, cervical stenosis, chronic inflammation and irritation. Clinically, majority of the patients present with vaginal bleeding [6]. Our patient was also a postmenopausal female, who presented with vaginal bleeding & foul-smelling vaginal discharge.

The pathogenesis of primary squamous cell carcinoma of endometrium is unknown. Several possibilities exist. First the squamous cell carcinoma is a complete malignant squamous differentiation of endometrial adenocarcinoma. Second, HPV is involved in the pathogenesis of Primarysquamous cell carcinoma of endometrium.

Thirdly, squamous metaplasia-dysplasia-squamous cell carcinoma sequence is involved in the pathogenesis of primarysquamous cell carcinoma of endometrium. Finally, primary endometrial squamous cell carcinoma may develop from ectopic cervical tissue in the endometrium [7].

In the present case, a retained IUCD was found and microscopy demonstrated Icthyosis Uteri with areas of dysplasia and invasive squamous cell carcinoma. Chronic irritation induced by retained IUCD might have led to the sequence of squamous metaplasia- dysplasia-invasive squamous cell carcinoma.

Valente P T et al had reported one case of primary endometrial squamous cell carcinoma following 25 years of IUCD retention [8]. Sharmaine Mitchell reported extensive squamous metaplasia & in situ squamous cell carcinoma of endometrium in a 65 year old patient who gave history of retained IUCD for 20 years [9].

The prognosis for patients with primary endometrial squamous cell carcinoma depends upon mainly the stage of the tumor. Primary endometrial squamous cell carcinoma has been reported to show a poorer prognosis than endometrioid carcinomas [6]. Survival rate for patients with stage I disease is 80% & for stage III is 20%. Hence early diagnosis and prompt treatment is imperative to improve the survival rate [2].

No final treatment recommendations have been given so far, therapy usually consists of surgical hysterectomy with adnexectomy and radiotherapy in some cases [6].

Our patient was treated with hysterectomy with bilateral salpingo-oophorectomy.There was no clinical suspicion of malignancy so lymphnode dissection was not done. She is alive without any recurrence and metastases 10 months after the operative treatment.

Conclusion

Rarely primary endometrial squamous cell carcinoma can arise in icthyosis uteri. Hence the possibility of primary endometrial squamous cell carcinoma should be kept in mind when atypical squamous epithelial cells are seen in endometrial curettage specimen, especially in a postmenopausal woman with history of retained IUCD.

Findings: Nil; **Conflict of Interest**: None initiated
Permission from IRB: Yes

References

1. Sood N, Sinha KS. Superficial Spreading Squamous Cell Carcinoma and Icthyosis Uteri with CIN III with p 16 Expression. JKIMSU.2017;6(4):126-30

2. Amita K, Padmini J. Squamous Cell Carcinoma of Endometrium with Extensive Icthyosis Uteri.Online J Health Allied Sci.2010;9(4):25

3. Kudrimoti JK, Chopdar AS, Puranik SC. Icthyosis Uteri eith in situ Carcinoma- Report of 2 cases.Annals of Pathology and Laboratory Medicine.2018;5(11):C-172-4. DOI: 10.21276/APALM.2166

4. Bagga PK, Jaswal TS, Datta U, Mahajan NC. Primary endometrial squamous cell carcinoma with extensive squamous metaplasia and dysplasia. Indian J Pathol Microbiol. 2008;51(2):267-8.

5. Jain M, Kashyap A, Biswas R. Primary Endometrial Squamous Cell Carcinoma in situ with Extensive Icthyosis Uteri: A Rare Case Report. J Clin Diagn Res. 2017; 11 (8:) ED13-4. DOI: 10.7860/JCDR/ 2017/ 29967.10384

6. Jetley S, Jairajpuri ZS, Hassan MJ, Madaan G, Jain R. Primary endometrial squamous cell carcinoma in situ: report of a rare disease. Sultan Qaboos University Med J. 2015 Nov;15(4):e559. DOI: 10.18295/squmj. 2015. 15.04.021

7. Terada T, Tateoka K. Primary pure squamous cell carcinoma of the endometrium: A case report. International J Clinic Experiment Pathol. 2013; 6 (5): 990.

8. Valente PT, Ahmad MM. Low grade uterine stromal sarcoma and squamous cell carcinoma associated with an IUD: a report of 2 cases. J Reprod Med. 2004;49 (4):324-8.

9. Mitchell S, Fletcher H, Williams NP. In situ squamous cell carcinoma of the endometrium associated with long-term intrauterine device (Dalkon Shield) usage. J Obstet Gynaecol. 1999; 19(1): 88-9. DOI:10.1080/ 01443619966119.

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