CLEAR CELL ADENOCARCINOMA CERVIX IN A 19 YEAR OLD UNMARRIED GIRL - A RARE CASE REPORT

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Abstract
Adenocarcinoma of the cervix is a rare condition that has shown an increase in incidence. Adenocarcinoma represents about 10-15% of malignant cervical tumours. Among these, the clear cell type accounts for 3%. This type of tumour affects mainly postmenopausal women but also occurs in young women with a history of prenatal exposure to diethylstilbestrol (DES). The prognosis for adenocarcinoma of the cervix is poor overall. This article discusses a case of clear cell adenocarcinoma of the cervix, unrelated to intrauterine exposure to DES, in a 19 year old unmarried girl with no history of sexual contact and who was treated with Type II modified radical hysterectomy with bilateral pelvic lymph node dissection followed by postoperative chemoradiation.

Keywords: Clear cell adenocarcinoma cervix, Diethylstilbestrol (DES), Modified radical hysterectomy

Introduction
Carcinoma of the cervix is the most common gynaecological malignancy in India.¹ The most common histological type is the squamous cell carcinoma. Adenocarcinoma accounts only for 10-15% of malignant cervical tumours.¹ Mean age of diagnosis of adenocarcinoma cervix is 49 years although it ranges from 25-79 years.² It is not known for sure whether the same etiological factor is important in both squamous cell carcinoma and adenocarcinoma cervix. The relationship to onset of sexual activity, multiple partners and smoking is not as clear in this histological type. Now, Human papilloma virus (HPV) 16, HPV 18, HPV 45, HPV 52 and HPV 32³ are implicated, mainly in endocervical adenocarcinomas. The most frequent histological subtype is endocervical adenocarcinoma accounting for 70% of cervical adenocarcinomas.² Others are classified according to their predominant cell type as serous, endometrial, clear cell and villoglandular adenocarcinoma.² Colposcopic evaluation is of limited value and cytologic smears may also be unrevealing in adenocarcinoma cervix.³

As such there is a marked decrease in the incidence of cervical squamous cell carcinoma thanks to the wide spread use of pap smear but in contrast the incidence of adenocarcinoma cervix is showing a rising trend.⁴ Clear cell adenocarcinoma cervix accounts for 3% of all adenocarcinoma cervix.² Its occurrence both in the cervix and vagina of young woman have been linked mainly to exposure in utero to Diethylstilbestrol (DES) taken by their mothers⁵ in an attempt to prevent adverse pregnancy outcomes, which was quiet the practice during 1950s to 1960s.⁵ The relationship between intrauterine exposure to DES and clear cell carcinoma was explored in a case-control study in 1971 and further confirmed by subsequent animal experiments and a cohort study designed in the early 1990s.⁶,⁷ Fortunately the risk of clear cell carcinoma appears to have diminished as this cohort has grown older, and currently as in our case most cases are sporadic. The clinical presentation is same whether the patient had antenatal DES exposure or not. They present with abnormal vaginal bleeding or discharge. The tumour may be polypoidal and fungating or ulcerative.

Case Report
A 19-year-old unmarried girl presented to the department of obstetrics and gynaecology, Maharaja Krishna Chandra Gajapati Maharaja Krishna Chandra Gajapati (MKCG) Medical College Berhampur with complaints of intermittent irregular bleeding per vaginum of one year duration and white discharge per vaginum (PV) of six months duration. On examination patient was cachectic. Abdominal examination did not reveal any organomegaly. Speculum examination revealed a cauliflower like polypoidal growth of size 3 X 2 cm arising from the anterior lip of cervix (Figure 1 & 2).

Figure 1 & 2. Per speculum examination showing polypoidal mass arising from the cervix

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Bimanual examination revealed that the mass was friable and bled on touch. Rectal examination revealed both parametria, Pouch of Douglas and rectal mucosa to be free and not involved. Provisional diagnosis was carcinoma cervix Stage IB1.

Her routine blood investigations were found to be normal. Pap Smear revealed only inflammation. Ultrasound showed a hypoechoic space occupying lesion measuring 30X20 mm in the cervix and lower uterine region with endometrial thickness of 5mm. Both ovaries and adnexa was normal. MRI revealed a cervical mass 32 X 24mm with altered signal intensity with no obvious invasion in to the contiguous structures. There was no evidence of pelvic and paraaortic lymphadenopathy (Figure 2).

We proceeded with examination under anaesthesia (EUA) and biopsy of cervical lesion (Figure 3).

EUA confirmed the earlier findings and histopathology (HP) of biopsy specimen revealed clear cell adenocarcinoma of cervix with interval endometrium. Hence, our diagnosis was confirmed as clear cell adenocarcinoma cervix FIGO stage IB1.

We planned and proceeded with Type II modified radical hysterectomy with bilateral pelvic lymph node dissection and transpositioning of right ovary to the under surface of diaphragm (Figure 4 & 5).

No signs of extrauterine spread of the disease was noted. Unique feature was that our surgery took less than the usual time, there was no venous engorgement or oozing which usually accompanies similar cases in parous women. On histopathology gross examination of surgical specimen revealed uterus with cervix 8 X 6 X 5 cm with cut section showing lesion 3X2cm diffusely involving the cervix and extending towards the endometrial cavity (Figure 6 & 7).
Histopathology revealed adenocarcinoma with tubulopapillary architecture with conspicuous hobnailing. Focal areas of clear cell morphology was noted (Figure 8).

Figure 8. High power field showing adenocarcinoma with tubulopapillary architecture with conspicuous hobnailing and focal areas of clear cell morphology.

On immunohistochemistry the tumour was positive for CK7, occasional tumour cells showed positivity for PS2 protein while negative for ER, PR and CK20.

Impression after HP and immunohistochemistry was clear cell adenocarcinoma cervix with endomyometrium, parametrium, vaginal vault histomorphologically normal and free from tumour infiltration. Lymph nodes showed reactive hyperplasia. Postoperative period was uneventful and patient was discharged on postoperative day 10. She is now undergoing chemoradiation and is under observation in our department.

Discussion

The data from the literature indicate that in primary clear cell adenocarcinoma cervix, either radiation therapy or radical hysterectomy and bilateral lymph node dissection by an experienced surgeon results in cure rates of 85-90% for patients with small volume disease. The mode of treatment depends on patient factors and available expertise. For adenocarcinoma that expands the cervix greater than 4 cm, the primary treatment should be radiation therapy. In smaller foci as in our patient, initial surgery followed by radiation therapy should be performed.

Survival of patients with cervical adenocarcinoma was recently reported in randomized control trials to be improved when combination of postoperative radiation and platinum based chemotherapy is applied. The question of whether clear cell carcinoma and other adenocarcinoma have worse prognosis than squamous cell carcinoma of the uterine cervix remains open. Korhonen suggested, after the analysis of 163 cases of primary cervical adenocarcinoma of different subtypes that the prognosis of clear cell carcinoma is similar to that of non-clear cell adenocarcinoma. Reich et al did not find a statistically significant difference in the prognosis of surgically treated patients with Stage IB-IIB clear cell carcinomas, squamous cell carcinomas and non clear cell carcinomas. Niibe et al based on literature review, suggested that 5-year survival rate in cervical adenocarcinoma is worse than cervical squamous cell carcinoma. A similar opinion was presented by Quinn and Freitag et al. All authors agree that the most important factor influencing the prognosis of each histopathological type is the stage of the disease.

Conclusion

The incidence of malignant neoplasm of genital organs in young woman under 20 years is relatively low. In our presented case, there was no family history, lack of sexual exposure and lack of epidemiological risk factors for carcinoma cervix such as human papilloma virus, oral contraceptive pill use, multiple sexual partners and smoking. No history of prenatal DES exposure is yet another unique feature, since it is known that clear cell adenocarcinoma cervix is rare in women without in utero DES exposure and if it occurs in the absence of DES exposure is usually in postmenopausal age group. Hence, clear cell adenocarcinoma cervix presenting in a 19 year old unmarried girl with no history of sexual contact and no prenatal DES exposure is such a very rare entity.

Editor’s comment

This case highlights the fact that clear cell adenocarcinoma of the cervix may present in younger patients without exposure to DES in utero. A high index of suspicion and scrupulous attention to histological detail will confirm diagnosis.

References


