A Rare Variant of Carcinoma Cervix “Adenoid Cystic Carcinoma” – Case and Brief Review of Literature

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Citation: Neha Varun., et al. “A Rare Variant of Carcinoma Cervix “Adenoid Cystic Carcinoma” – Case and Brief Review of Literature”. EC Gynaecology 6.2 (2017): 62-66.

Abstract

Introduction: Adenoid cystic carcinoma (ACC) of the cervix is the rare and the unusual variant of adenocarcinoma. It is a malignant epithelial neoplasm with a distinctive histological appearance accounting for 0.5 - 2% of all cervical carcinomas. It is generally a locally aggressive tumor and has a high tendency for local recurrence and distant metastasis. In this article we are reporting a case of a primary adenoid cystic carcinoma of the cervix and discuss a brief review of related literature.

Case Presentation: A forty-six-year-old postmenopausal female presented in the outpatient department with the history of postmenopausal vaginal bleeding. On examination, a 3 × 3 cm hard growth arising from the cervix was visualized. A clinical diagnosis of carcinoma cervix stage 2B was made and histopathological examination report of the cervical biopsy revealed adenoid cystic carcinoma of the cervix. The patient was successfully treated with the radiotherapy.

Conclusion: Standard treatment of ACC has not been proposed yet because of the rarity of this condition. From the review of the literature, it appeared that the combination of all the modalities (surgery, radiotherapy, and chemotherapy) is required for the successful management and the long-term remission. For the locally advanced diseases, concurrent chemo-radiotherapy seems to be the logical option.

Keywords: Adenoid Cystic Carcinoma; Radiotherapy; Surgery

Abbreviations

ACC: Adenoid Cystic Carcinoma; ABC: Adenoid Basal Carcinoma

Introduction

Adenoid cystic carcinoma of the cervix (adenocystic carcinoma, cylindroma or cylindromatous adenocarcinoma) is the rare and the unusual variant of adenocarcinoma [1]. It is a malignant epithelial neoplasm with a distinctive histological appearance accounting for 0.5 - 2% of all cervical carcinomas [2]. ACC of the cervix is usually seen in the postmenopausal age group and very few cases have been reported in women below the age of 40 years [3]. ACC is generally a locally aggressive tumor and has a high tendency for local recurrence and distant metastasis - vascular, lymphatic and neural invasion, with the subsequent hematogenous spread [3]. Because of the rarity of...
the disease, no standard treatment has yet been proposed. Surgery and radiation therapy, either alone, or in a combined setting, have been used in the treatment of these cases. Considering high local failures and distant relapses, an aggressive local and systemic treatment is recommended in an advanced stage and inoperable disease.

**Case Presentation**

A forty-six-year-old postmenopausal female presented in the outpatient department with the chief complaints of postmenopausal vaginal bleeding. She was a known case of type 2 diabetes mellitus. Speculum examination revealed a 3 x 3 cms fungating growth arising from the cervix and involving the proximal vagina. Bimanual rectovaginal examination revealed a hard growth arising from the cervix, growth was bleeding on touch, no parametrical involvement and uterosacral were thickened and seems to be infiltrated by a tumor. A clinical diagnosis of carcinoma cervix stage 2B was made. Magnetic resonance imaging showed a bulky cervix with a lobulated soft tissue lesion measuring 2.8 × 1.8 × 3.1 cm involving the posterior wall of the cervix and endocervical canal. It was seen to infiltrate the lower uterine corpus and distally into the proximal vaginal cavity. The lesion was focally abutting the rectum with loss of fat planes and focal anterior wall involvement. The histopathological finding of the deep cervical biopsy specimen revealed adenoid cystic carcinoma cervix (Figure 1-3). The patient was successfully treated with the radiotherapy. The patient is on follow up with no recurrence.

*Figure 1: Photomicrograph of cervical biopsy specimen (lower magnification), showing adenoid cystic carcinoma (glandular spaces filled with eosinophilic material).*
Discussion

ACC is a rare malignant tumor. It is most commonly found in the salivary glands, the respiratory tract and can involve other sites containing secretory gland component [4,5]. It constitutes around less than 1% of all cervical carcinomas [4]. First case of ACC of the cervix has been diagnosed in 1949 [6,7]. ACC has a typical histological appearance, regardless of its anatomical location [1]. This carcinoma generally occurs in older women in their sixth and seventh decades [4]. The most accepted theory regarding its cervical origin is from “multipotent reserve cells” of endocervix which can have squamous or glandular differentiation [4]. The origin of this disease is still unknown. Human papillomavirus (HPV) infection is known to be a cause of cervical cancer and some authors have displayed the association of HPV with the ACC and squamous cell carcinoma, indicating a notable role of HPV in the pathogenesis these tumors [8]. Some authors have suggested the association of ACC of the cervix with the high parity and black race [8-10].

The main clinical symptom is usually an abnormal vaginal bleeding [4,11]. In some patients enlargement of the uterus and vaginal discharge can be there. This tumor commonly presents as a hard palpable mass clinically which can be a ulcerated or friable growth but more commonly a non-friable growth. This tumor is more aggressive than squamous cell carcinoma with a higher propensity for local and metastatic recurrence [3]. Grossly on examination it didn’t differ much from the usual squamous cell carcinoma [8]. These tumors cannot be simply classified into squamous or glandular cell lesion.

The grading system of ACC was reported by Perzin., et al. based on the characteristic histological pattern: solid, tubular and cribriform [12,13]. The most typical pattern of ACC is cribriform [14,15]. Histologically it is characterized by the presence of uniform round or ovoid cells with varying amounts of cytoplasm and discrepancy in the size and staining standard of the nuclei. The outermost cells are paling around the central core and are arranged into innumerable cystic cavities fill up with a homogeneous transparent substance representing typical hyaline bodies. The stromal reaction consists of two to three layers of fibrocytes encasing the cellular nests, and lymphocytes or plasma cells infiltration [1]. Cells of ACC immunohistochemically stains positive for many cytokeratins, EMA, CAM 5.2, CK7 and CD 117.

CEA may also be demonstrated. There are no myoepithelial cells in case of cervical ACC, which could differentiate this tumor from ACC at other sites [4]. It has been reported that the three histologic patterns of ACC represent the aggressiveness of the tumor. It has been scarcely reported to be related to the cervical intraepithelial neoplasia, squamous cell carcinoma, adenocarcinoma and uterine cervical sarcoma [16]. Some authors reported that CK 14 stained in cases of adenoid basal carcinoma (ABC); therefore, this aids to differentiate between ACC and ABC [1]. Differential diagnosis of this tumor comprises of adenoid basal carcinoma and basaloid squamous cell carcinoma. ABC is much less aggressive than the ACC.

The solid subtype of the ACC is associated with worse prognosis in terms of the development of distant metastases and overall survival rate. The cribriform subtype is associated with significant local aggressiveness and a decreased survival rate as compared with the tubular subtype [7]. Other particular tumor characteristics that are related to the high probability of recurrence and mortality in stage Ib cervical cancers, in general, should be appraised in ACC. These features consist of the large size of the tumor, deep stromal invasion, and the capillary lymphatic spaces infiltration by tumor establish on the microscopic examination [17]. The poor outcome has been reported in cases of advanced disease (stage III and IV) [18].

There is a paucity of prospective studies as well as because of the rarity of this condition, no standard treatment has yet been suggested. Majority of patients were treated on the line of squamous cell carcinoma. Combined modalities have been used in the treatment of ACC of the cervix [3,17]. The best modality of treatment is surgery, chiefly when the surgical margins of resection could be determined histologically. Radiotherapy is reported to be a useful method of treatment for surgically operated patients where surgical borders were inadequate, patients who had recurrent disease and in inoperable patients.

ACC of the cervix is reported to be a radiosensitive tumor and appropriate results have been seen in early-stage patients that have been treated with adjuvant radiotherapy, as compared to those patients where only surgery has been done [4]. Chemotherapy is not very successful in cervical ACC and it is indicated only in advanced/recurrent/metastatic disease without much result. Adenoid cystic carcinoma of the cervix is known to be associated with poor prognosis due to extensive lymph node and vascular metastases, especially to lungs, which is the frequent site of metastasis [4]. Distant metastasis of a tumour is the main predictive factor of the survival. Therefore, the majority of patients require vigorous local and systemic therapy because of the high incidence of local and distant relapses. The recurrence rate was 59%, 89% and 100% for the tubular tumors, classic cribriform lesions, and the solid variety respectively.

Conclusion

ACC of the cervix is a rare variety of a cervical carcinoma, particularly an aggressive neoplasm. It requires aggressive treatment, intensification of postoperative treatment regimens and careful follow-up, thus needs to be distinguished from other tumors with similar histologic findings.

Acknowledgement

Nil.

Conflict of Interest

Nil.

Bibliography


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