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Original Research Paper

Pathology

ADENOID CYSTIC CARCINOMA OF UTERINE CERVIX IN A YOUNG PATIENT: A CASE REPORT

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ABSTRACT Adenoid cystic carcinoma of the cervix is a very rare condition that occurs more frequently in postmenopausal women, although it can occur infrequently in people under the age of 40. In the present case, a 29-year-old woman complained mainly of post-coital bleeding and bloody vaginal discharge. Per speculum examination revealed a firm mass in the uterine cervix. Ultrasonography revealed a 2.4 cm x 1.9 cm hypoechoic lesion in the posterior lip of the cervix. Liquid based cytology was suggestive of squamous cell abnormality ASCUS. On HPV testing HPV16/31 was detected high, while HPV18/45 was not detected. The diagnosis of adenoid cystic carcinoma was confirmed by the histopathological examination of the cervical biopsy. Immunohistochemistry showed tumor cells with positive expression of p16, p63, and S-100. We report this case because of its rarity, particularly in young patients.

KEYWORDS : Adenoid cystic carcinoma, cylindroma, Adenoid basal carcinoma, Human papilloma virus.

INTRODUCTION

Primary adenoid cystic carcinoma (ACC) of the cervix is extremely rare, accounting for less than 1% of all cervical carcinomas and approximately 3% of all primary cervical adenocarcinomas.[1] In female reproductive tract, ACC is seen more commonly in the Bartholin gland and rarely in the cervix.[2] It generally occurs in post-menopausal women, however, ACC of cervix occurring in younger age is also documented in the literature.[3] It is locally aggressive with higher capability of metastasis to local organ even when diagnosed in the early stages. Because of the rarity of the disease, no standard treatment has been proposed so far. This case is being reported because of its rarity, especially in women of young age.

Case Report

A 29-year-old female came with the chief complaints of post-coital bleeding and vaginal discharge for 3 months. Per speculum examination revealed a firm mass of 3 cm x 3 cm observed on posterior lip of cervix. Per vaginal examination showed a firm mass hard to touch, which bleed on touch felt. Ultrasonography (USG) revealed 2.4 cm x 1.9 cm hypoechoic lesion in posterior lip of cervix. Liquid based cytology was performed. Smear examined are composed of mainly intermediate and superficial squamous epithelial cells against a background of marked acute inflammation. However few cells seen with enlarged hyperchromatic nuclei. Impression of liquid based cytology was suggestive of squamous cell abnormality-ASCUS (Atypical squamous cells of undetermined significance). HPV testing HPV16/31 was detected high, while HPV18/45 was not detected. To ascertain the type and nature of pathology and decide the further line of therapy, a colposcopically guided cervical biopsy was performed and sent for histopathological examination. Microscopic examination of tissue sections of the tumor showed tumor cells dispersed in cribriform as well as in solid pattern. The tumor cells were small, uniform, composed of dense basophilic nuclei with inconspicuous nucleoli. The solid pattern was characterized by large masses or nests of uniform basaloid tumor cells. Immunohistochemical study was performed. Immunohistochemistry showed focal positivity for p16, p63, S-100, while CD56, synaptophysin, CEA were found to be negative. A histopathological diagnosis of adenoid cystic carcinoma of cervix was made.

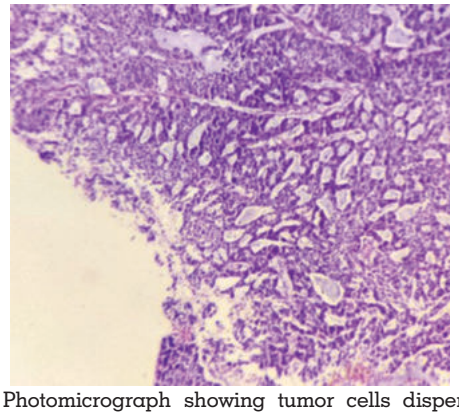

Fig.1 Photomicrograph showing tumor cells dispersed in cribriform as well as in solid pattern. (H and E, 40X)

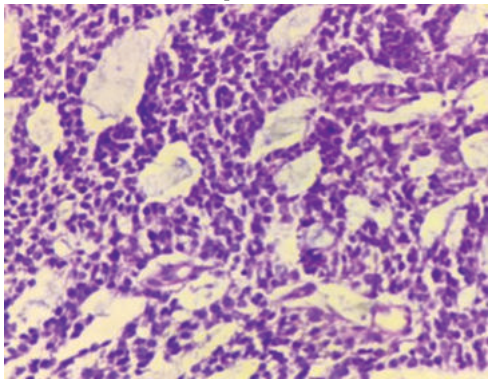

Fig.2 Photomicrograph showing tumor cells which were small and uniform composed of dense basophilic nuclei with inconspicuous nucleoli. (H and E, 100X)

DISCUSSION

Adenoid cystic carcinoma (ACC) is a malignant epithelial

neoplasm that arises in the salivary glands and can occur in a variety of other less common sites besides uterine cervix, including the breasts, lacrimal gland, prostate, esophagus, and lungs.[4] The first case of adenoid cystic carcinoma was described by Paalman in 1949 as cylindroma in 1949.[5] The origin of this disease is still unknown. Human papilloma virus (HPV) infection is thought to be an underlying cause of cervical cancer, however its role in the pathogenesis of ACC is not well established.[6] The most accepted view regarding its origin in the cervix is from "reserve cell" of endocervix.[7] A comprehensive literature review using PubMed was performed in January 2023 using the search term "Adenoid cystic carcinoma of cervix". To our knowledge about 160 cases have been described in the English literature to the duration. Based on this limited data, it has been determined that ACC of cervix is an extremely rare tumor and considered as a disease of postmenopausal women however it rarely occurs in younger patients. The main symptom of disease are usually vaginal bleeding, vaginal discharge, and uterine enlargement.Adenoid cystic carcinoma of cervix is associated with poor prognosis due to widespread lymph node and vascular metastases, especially to lung.[8]

The most common characteristic pattern in ACC is cribriform with polygonal to spindled cells forming numerous duct like structure containing extracellular matrices filled with homogenous eosinophilic or granular basophilic material. The solid type is characterized by solid sheets with prominent mitotic activity and frequently with central necrosis. In the tubular type, small ductal structures formed by one or two layers of cuboidal or polygonal tumor cells were intermingled with solid strands of tumor cells.[9]

Adenoid basal carcinoma (ABC) is the most important differential diagnosis of ACC of cervix. Since most cases of ABC have a lower risk of recurrence and metastasis than ACC, it is important to distinguish between the two. Histologically, ABC shows proliferation of nested uniform, bland basaloid cells with peripheral palisading pattern, but unlike ACC nuclear pleomorphism is minimal, with rare or absent mitoses, and no necrosis.[6]

Other differential diagnosis to consider include small cell carcinoma of the cervix, carcinoid tumor of the cervix, carcinosarcoma of the cervix, and ectopic prostatic tissue.

Because of the rarity of the disease and lack of prospective studies, no standard treatment has been proposed to date. Surgery and radiation therapy has been used in the treatment of ACC of the cervix. Chemotherapy plays a minor role in the management of this tumor but has been less effective against advanced or recurrent diseases.[10]

CONCLUSION

To conclude, this case is unique not because of the rarity of ACC occurring in cervix, but because of aggressive nature and presentation. As only some case reports are available in the literature in the younger women, further studies of the reported cases are needed to assess risk factors, treatment strategies and prognosis.

Conflict of interest

There are no conflict of interest.

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Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given her consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due effort will be made to conceal identity, but anonymity cannot be guaranteed.

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