Adenoid Cystic Carcinoma of Uterine Cervix

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Primary adenoid cystic carcinoma of the cervix is a rare condition. It is a rare variant of adenocarcinoma and is mostly seen in postmenopausal women. Here we report a case of primary adenoid cystic carcinoma of uterine cervix.

Keywords: adenocarcinoma, adenoid cystic carcinoma, uterine cervix.

INTRODUCTION

Primary adenoid cystic carcinoma of the cervix is extremely rare, accounting for less than 1% of all cervical carcinomas.[1] It is a rare and peculiar variant of adenocarcinoma and is mostly seen in postmenopausal women, is locally aggressive and capable of metastasis to other organs even in its early stage.[2] Common sites for adenoid cystic carcinoma (ACC) are the salivary glands and the respiratory tract. Occasionally it is found in the mucous membrane of head and neck, skin and breast. In the female genital tract, it has been rarely described in the Bartholin's gland and the uterine cervix.[3]

The first case was described in 1949 by Paalman.[4] As per the published data only 160 cases have been reported up to 2008.[5] It is usually seen in postmenopausal, often multigravid Iblack women.[6] Local infiltration, vascular, lymphatic and neural invasion with late metastases are its characteristic features.[7] We report a case of adenoid cystic carcinoma of uterine cervix in an elderly female.

CASE

A 55 year-old female presented with spontaneous vaginal bleeding for three months. There was no history of oral contraceptive pill's intake. Per vaginal and per speculum examination revealed an ulcerative growth of approximately 2 cm in diameter that bled on touch. Vagina appeared healthy. On bimanual recto-vaginal examination, bilateral parametrium were free of tumor infiltration. Sonography revealed an echogenic mass of 2 cm in region of cervix. The patient was staged Ib1 according to the FIGO classification for carcinoma cervix. Hemoglobin was 9 gm%. Other blood profile and biochemical examination were within normal limits. Chest X-ray was normal.

Biopsy of the cervicl growth was done and sent for histopathological examination. Microscopic examination showed fragmented biopsy with tumor cells disposed in cribriform nests and cords. The hyaline stroma forming cyst like spaces within cell nests revealed a classic Swiss cheese pattern. The tumor cells were small, uniform, composed of dense basophilic nuclei with inconspicuous

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nucleoli. Mitotic figures were rarely found. A diagnosis of adenoid cystic carcinoma was made (Figure 1).

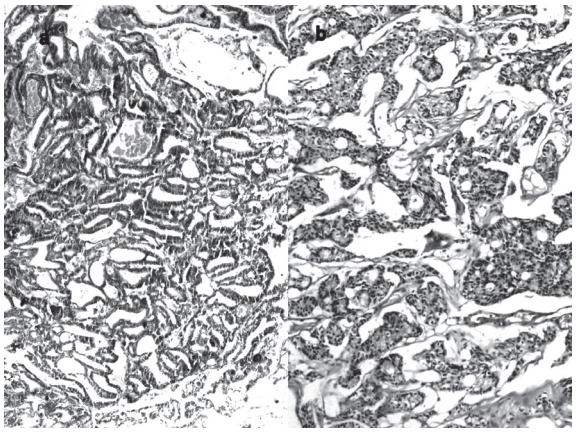

Figure 1. Photomicrograph of fragmented cervical biopsy revealing histological features of adenoid cystic carcinoma (a. H&E; x 100 , b. H&E; x 200).

The patient was managed successfully by radiotherapy followed by hysterectomy. The postoperative period was uneventful and the patient is on regular follow-up for last Six months.

COMMENT

Adenoid cystic carcinoma is a rare tumor accounting for < 1-3% of primary adenocarcinomas of the uterine cervix. It is usually seen in postmenopausal, often multigravid black women. Cervical adenoid cystic carcinoma rarely occurs in patients younger than 40 years of age.[8]

Despite the fact that cervical cancer is the commonest malignancy in Indian women, only 19 cases of ACC of the cervix have been reported from India and majority of Indian patients were reported during fifth decade of their life.[3, 9-12]

Etiology of adenoid cystic carcinoma is unknown. However, Yang and Gordon[13] reported a case of cervical adenoid cystic carcinoma coexisting with a variety of human pappiloma virus (HPV) related lesions which raises a speculation that HPV may also be the causative factor. The most accepted view regarding its origin in the cervix is from "reserve cells" of endocervix. Most of these tumors are found in multiparous women during their post-menopausal life.[8]

Clinically, it presents as a nonfriable mass on speculum examination, in contrast to the friable growth usually seen in squamous cell carcinoma of cervix. Most of the cases present with vaginal bleeding as chief complaint.[3] The morphologic appearance is similar to that of homonymous tumors of salivary gland. It is an aggressive tumor. Extensive local infiltration and invasion of lymphatic vessels as well as perineural spaces, with subsequent hematogenous spread are characteristic. Distant metastases have been reported in lungs, bones, liver and brain.[8]

Because of the rarity of the disease and the absence of prospective studies, no standard treatment has yet been proposed. Most patients were treated as squamous cell carcinoma. Surgery seems to be the treatment of choice in combination with adjuvant radiotherapy and/or chemotherapy, based on the clinical stage and presence of metastasis.[5]

ACC of the cervix is an aggressive but radiosensitive tumor.[8] Though concluding statement regarding the results cannot be made from the cases accumulated from the various reported series, nonetheless literature shows that radiotherapy too is effective in early stage and even in stage I results with radiotherapy appear to better than surgery. Cases of early stage disease having risk for metastases and local failure should be identified and managed with multimodality treatment using surgery for bulky disease with post-operative radiotherapy and chemotherapy. In advanced stages combination of chemotherapy and radiotherapy is required.[13]

CONCLUSIONS

Adenoid cystic carcinoma, a rare variant of adenocarcinomas is a rare neoplasm of the uterine cervix. As only some case reports are available in the literature, further studies and metaanalysis of the reported cases is required to assess high risk factors and to design a comprehensive treatment strategy.

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