INTRODUCTION

Papillary squamous cell carcinoma and adenoid cystic carcinoma (ACC) are rare variants of squamous cell carcinoma and adenocarcinoma of the cervix. Papillary squamous cell carcinoma was described by Randall et al. Histologically, papillary squamous cell carcinomas are composed of papillary projections covered by several layers of atypical epithelial cells. The behavior of papillary squamous cell carcinoma is similar to that of invasive squamous cell carcinoma with the exception of late recurrences in various larger series. ACC of the cervix is an uncommon tumor accounting for < 1% of all cervical adenocarcinomas. ACC appearing in cervix was first reported by Paalman and Counseler. The etiology of cervical ACC is not known. Yang and Gordon reported a case of cervical ACC coexisting with human papillomavirus (HPV) related lesions. ACC of the uterine cervix is associated with poor prognosis due to widespread metastasis.

We report these two rare variants of cervical carcinoma with review of the literature.

CASE HISTORY

This was a case of two menopausal woman with age range between 51 and 49-years respectively presented with post-menopausal bleeding. Hysterectomy was performed in both cases. In the first case, grossly, the cervix was everted, keratinized and hypertrophic. The cut surface was unremarkable. Microscopy revealed multiple papillae with broad fibro-vascular cores covered by several layers of atypical epithelial cells. The tumor cells showed various degree of atypia while in others areas they were very delicate fibro-vascular cores covered by mono-layered epithelium. The tumor cells were mildly pleomorphic, had dense basophilic cytoplasm, moderately pleomorphic nuclei with inconspicuous nucleoli. Mitotic figures were scanty. Foci of moderately differentiated squamous cell carcinoma were also identified. The adenoid cystic component was positive for actin and the hyaline material was positive for periodic-acid-Schiff stain. A diagnosis of ACC of the cervix with foci of squamous cell carcinoma was given.

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ABSTRACT

Cervical cancer is the most common malignancy in females in the developing world. Squamous cell carcinoma and adenocarcinoma are the most common histological types. Papillary squamous cell carcinoma is a rare variant of squamous cell carcinoma associated with recurrence and more aggressive behavior. Adenoid cystic carcinoma is a rare variant of adenocarcinoma which is associated with early lymph node and vascular metastasis.

Key words: Adenoid cell carcinoma, cervix, papillary squamous cell carcinoma

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DISCUSSION

Non-glandular papillary carcinoma of the cervix are uncommon tumors with rather sketchy literature. These tumors were initially described by Randall et al., who reported that they had a histologic resemblance to transitional cell carcinoma of the bladder. These papillary tumors of the cervix can be subdivided into three groups based on their histologic appearance: Predominantly squamous, predominantly transitional, and mixed squamous and transitional. However, all three histologic subdivisions demonstrated positivity for cytokeratin 7 and negativity for cytokeratin 20, which is a pattern more typical of cervical squamous lesions than of transitional cell tumors of the urinary tract. Because they demonstrate a spectrum of histologic appearances, these tumors have been referred to as papillary squamotransitional carcinomas.

Clinically most of the cases were older women (mean age 46.3) and a great proportion presented with abnormal vaginal bleeding, similar to cases reported by Randall et al. and Koenig et al. Histologically, papillary squamous cell carcinomas are composed of papillary projections covered by several layers of atypical epithelial cells. In cases that appear more transitional, the cells are oval with their long axis oriented perpendicular to the surface and there is a flattening of the cells as they reach the surface. The behavior of papillary squamous cell carcinoma is similar to that of invasive squamous cell carcinoma with the exception that late recurrences have been reported in the larger series. Papillary squamous cell carcinoma can be mistaken for a papillary squamous cell carcinoma in situ on a superficial biopsy. Because papillary squamous cell carcinoma is capable of acting aggressively and metastasizing, it is important that a cone biopsy performed to rule out invasion whenever a papillary squamous cell carcinoma in situ is diagnosed. These tumors can also be mistakenly diagnosed as verrucous carcinoma or as condyloma acuminate with atypia. Papillary squamous cell carcinomas lack the condylomatous changes and degree of cytoplasmic differentiation present in the verrucous carcinoma.

The common site for ACC is the salivary glands and the respiratory tract, 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. ACC appearing in cervix was first reported by Paalman and...
Counseller. Primary ACC of the cervix accounts for less than 1% of all cervical carcinomas.

ACC of the cervix was previously considered as the disease of the post-menopausal women, with an average age of presentation to be 20 years older than squamous cell carcinoma of the cervix. However, later reviews mentioned it in below 40 years of age also but such examples are very few. The etiology of cervical ACC is not known. Yang and Gordon in their study reported a case of cervical ACC co-existing with a variety of HPV related lesions—condyloma acuminata, vulvar intraepithelial neoplasm, cervical intraepithelial neoplasm, and invasive basaloid squamous cell carcinoma which raises the possibility that HPV may also be the causative factor for ACC of the uterine cervix. In our case also, cervical ACC is associated with high grade squamous cell carcinoma of the cervix. The tumor probably originates from the endocervical multi-potent reserve cells. Presence of associated squamous cell differentiation in most of the cases suggest that tumor arises from some subcolumnar reserve cells in the transformation zone, which are thought to be the progenitor of most other cervical carcinomas.

The cervical ACCs show necrosis, a high mitotic rate, and greater nuclear pleomorphism in contrast to its salivary gland counterpart. ACC of the uterine cervix is associated with poor prognosis due to widespread lymph node and vascular metastases, especially to lungs, which is the commonest site of metastasis. Cause of early metastasis is unclear. In a study conducted by Ishiko et al., it was shown that the expression of cyclo-oxygenase-2 in the tumor might induce the expression of vascular endothelial growth factor thus, increasing angiogenesis and enhance tumor growth and invasion. The most important differential diagnosis of cervical ACC is adenoid basal carcinoma (ABC). Both tumors are common in the postmenopausal women but patients with ABC are usually asymptomatic. Histologically, ABC has proliferation of nested uniform, bland basaloid cells with a peripheral palisading pattern. Cellular pleomorphism, mitoses, necrosis, stromal hyalinization and metastasis are commonly identified in ACC but are rare or absent in ABC. Immunohistochemical studies show ACC to be positive for actin, collagen IV, and laminin, whereas these stains do not highlight the tumor cells of ABC though both show positivity to CEA, EMA, CK7, CK20 and CAM 5.2. Treatment is a judicious combination of radical pelvic surgery, radiotherapy and chemotherapy except in the patients with stage I (especially those with histological features associated with a higher risk of recurrence) who should receive aggressive local therapy alone.

CONCLUSION

Papillary squamous cell carcinoma and ACC of the cervix are rare primary tumors of the cervix associated with poor prognosis due to the occurrence of late recurrence and distant metastasis respectively.

REFERENCES