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CASE REPORT

Adenoid Cystic Carcinoma (ACC) Cervix in a Young Female- A Rare Occurrence

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Abstract

Adenoid cystic carcinoma cervix (ACC) is an extremely rare (representing <1% of cervical carcinomas), highly aggressive cancer of old, macroscopically appearing as an irregular, friable mass and histologically showing a pseudo glandular or cribriform growth pattern. The tumor is highly infiltrative, often associated with vascular, lymphatic as well as perineural invasion and early recurrence. We report a case of ACC Cervix in a 20-year-old female who presented with vaginal bleeding and cervical biopsy suggested ACC. The patient underwent total abdominal hysterectomy with bilateral adnexectomy and the histopathological examination revealed adenoid cystic carcinoma cervix extending and inolving endometrium and greater than 2/3rd of the myometrium with uninvolved adnexa. We report this case because of its rarity and particularly unusual occurrence in young females.

Key Words

Adenoid Cystic Carcinoma, Cervical Carcinoma, Squamous Cell Carcinoma

Introduction

Adenoid cystic carcinoma (ACC) is a rare, malignant neoplasm with a distinctive histological appearance, despite its varied anatomic location, the foremost common sites of involvement appear to be the minor salivary glands, major salivary glands, and the extraoral seromucinous glands. [1] In the female reproductive tract, ACC occurs most typically in the Bartholin gland and infrequently in the cervix, accounting for fewer than 1% of all cervical carcinoma. [2] Adenoid cystic carcinoma appearing within the cervix was first reported by Paalman and Counseller in 1949. [3] It's most commonly seen in patients between the sixth and seventh decades (mean age of 71 years in the largest series) and is more common in African-American women. [4] Very rare cases are reported in women under the age of 40 years. [5] Histologically, cervical adenoid cystic carcinoma has an appearance just like that arising in the exocrine gland, consisting of basaloid cells arranged in cribriform , tubular and solid growth patterns. Adenoid cystic behaves aggressively with frequent local recurrences or metastatic spread. [4]

Case Report

A 20-year-old woman presented with the complaint of vaginal bleeding and pelvic pain for the past two months. She had no significant prior medical and menstrual history. Per vaginal and per speculum examination revealed an exophytic friable mass arising from the cervix, which bled heavily on touch involving the upper part of the vaginal cavity. Cervical biopsy and total abdominal hysterectomy

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Fig 1. Gross Appearance of Adenoid Cystic Carcinoma Cervix. The Grayish White Gelatinous Cervical Growth is Seen Involving the Uterus

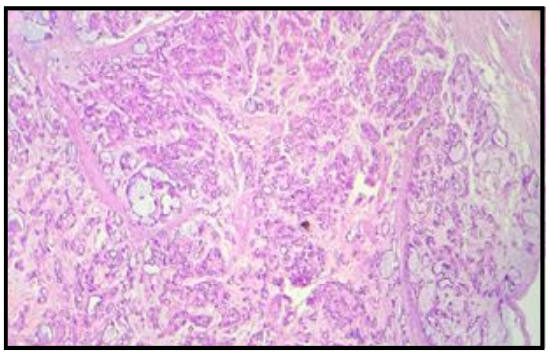

Fig 2. Photomicrograph Showing Infiltrative Pattern-Adenoid Cystic Carcinoma (40×, H&E).

specimen were sent for histopathological examination.

Gross examination of the total hysterectomy with bilateral adnexectomy specimen (*Fig 1*) showed a large, grayish-white, gelatinous growth measuring 5×3cm arising from the cervix which has eaten up the cervix and involved part of the uterus with grossly uninvolved adnexa.

Microscopic examination of tissue sections of the tumor showed proliferation of basaloid cells arranged in a cribriform and tubular pattern with abundant luminal mucin (*Fig 2*). The tumor cells were small and only mildly pleomorphic with hyperchromatic nuclei, inconspicuous nucleoli, and occasional mitotic figures (*Fig 3,4*). A histopathological diagnosis of Adenoid cystic carcinoma, cervix involving greater than 2/3rd of uterine stroma was made.

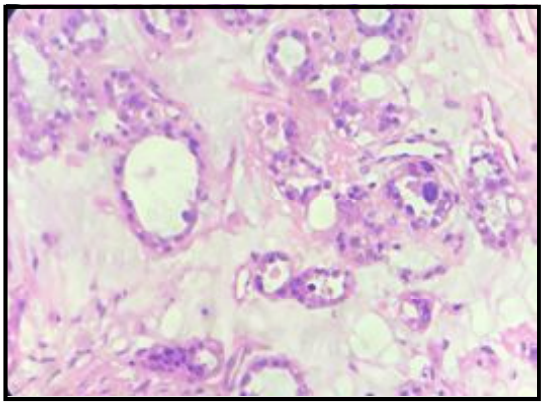

Fig 3. Photomicrograph Showing Tubular Arrangement with mild Cellular Pleomorphism (400×, H&E)

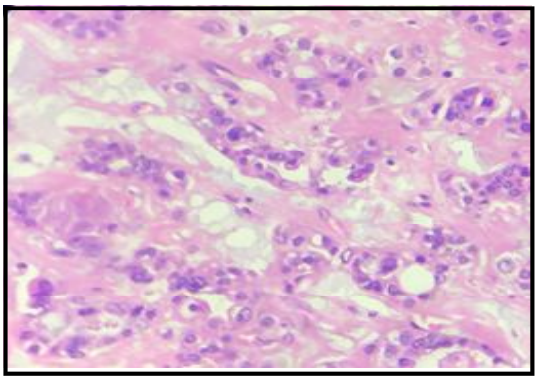

Fig 3. Photomicrograph Showing Eosinophilic Material Present in Cyst like Spaces (400×, H&E).

Discussion

We present a case of adenoid cystic carcinoma in a young female. Previously, adenoid cystic carcinoma of the cervix was considered as the disease of postmenopausal women, with an average age of presentation found to be 20 years older than squamous cell carcinoma of the cervix. However, later reviews mentioned it in females below 40 years of age also but such examples are very few [5] Clinically, it presents as a hard mass that can be ulcerated or friable. The main symptom is usually vaginal bleeding. [6] These clinical features are consistent with the case presented here, as the patient presented with vaginal bleeding and had a palpable, friable mass.

The most accepted view regarding its origin in the cervix is from the "reserve cells" of the endocervix. The origin

of this disease remains unknown. Also, Schi *et al.* [7] demonstrated that the high-risk Human Papilloma Virus (HPV) might plays a major role in ACC tumor pathogenesis.

Histologically our patient had characteristic features of ACC. Grossly it always features a solid appearance and an infiltrative pattern of growth. Microscopically, cervical adenoid cystic carcinoma has an appearance similar to that arising in the salivary gland, consisting of nests, islands, cords, and trabeculae of crowded cells with little cytoplasm and hyperchromatic nuclei. Characteristically, the cellular nests have a cribriform pattern with palisading nuclei, surrounding rounded spaces filled with eosinophilic hyaline or mucinous material [8] Other less frequent patterns including tubular, trabecular, solid, or undifferentiated may be observed with surrounding hyaline material. Lymphovascular invasion is frequent.

Adenoid basal cell carcinoma is an important differential since the majority of cases of adenoid basal cell carcinoma are benign owing to the low potential for recurrence and metastasis. On morphology; pleomorphism, numerous mitoses, necrosis, and a desmoplastic stromal response are common in ACC; in contrast, ABC lacks intraluminal hyaline material frequently present in ACC and has less pleomorphic nuclei showing less mitotic activity. Besides, ABC is often distinguished by the absence of CD117 immunostaining.

ACC of the cervix seems to be more aggressive than squamous cell carcinoma with a propensity tendency to local and metastatic recurrence. [8] .The most commonly reported sites of distant metastasis for carcinoma cervix include the lungs, bones, mediastinal and supraclavicular lymph nodes and the liver. [9]

Because of the rarity of the disease and the absence of prospective studies, no universal consent has been proposed in the standard treatment. However, ACC of the cervix is taken into account as a radiosensitive tumor and better results are reported in early stages patients that are treated with adjuvant radiotherapy. [10]

Conclusion

ACC of the cervix is a rare and aggressive neoplasm that is capable of distant metastasis in its early stage. It usually presents in the elderly age group. Very few cases have been reported in the young age group. Enhancement of postoperative treatment regimens and careful follow-up makes it essential to distinguish it from other tumors with similar histologic aspects.

References

1. Jaso J, Malhotra R. Adenoid cystic carcinoma. Arch Pathol Lab Med 2011;135(4):511-15.
2. Benhayoune K, El Fatemi H, Bannani A, Melhouf A, Harmouch T. Adenoid cystic carcinoma of cervix: two cases report and review of the literature. Pan Afr Med J 2015;20:77
3. Paalman RJ, Counseller US. Cylindroma of cervix with procedentia. Am J Obstet Gynecol 1949;58:184-87.
4. Pirog EC, Wright TC, Ronnett BM, Kurman RJ. Carcinoma and other tumors of the cervix. In: Kurman RJ., editor. Blaustein's Pathology of the Female Genital Tract. 7th. New York, NY, USA: Springer; 2019. pp. 361-63.
5. King LA, Talledo OE, Gallup DG, Melhus O, Otken LB. Adenoid cystic carcinoma of the cervix in women under age 40. Gynecol Oncol 1989;32(1):26-30.
6. Dixit S, Singhal S, Neema J, Soornarayan R, Baboo HA. Adenoid cystic carcinoma of the cervix in a young patient. J Postgrad Med 1994;40:94-95
7. Shi X, Wu S, Huo Z, Ling Q, Luo Y, Liang Z. Co-existing of adenoid cystic carcinoma and invasive squamous cell carcinoma of the uterine cervix: a report of 3 cases with immunohistochemical study and evaluation of human papillomavirus status. Diagn Pathol 2015;10:145.
8. Nucci M.R, Lee K.R, Crum C.P. Tumors of the Female Genital Tract cervix. In: D. M. Fletcher, editors. Diagnostic Histopathology of Tumors. 4rd ed. Elsevier; 2013.pp. 827-28.
9. Bhardwaj S, Mahajan D, Vir Gupta Y. Metastatic squamous cell carcinoma of the cervix presenting as a splenic cyst. J K Science 2008;10(3):146 -8.
10. Kaur P, Khurana A, Chauhan AK, Singh G, Kataria SP, Singh S. Adenoid cystic carcinoma of cervix: treatment strategy. J Clin Diagn Res 2013;7(11):27