

# Adenoid Cystic Carcinoma of the Cervix: Case Report

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## Abstract

Adenoid cystic carcinoma (ACC) is usually an epithelial neoplasm of the salivary glands but can also occur in the lacrimal glands and the mucous glands of the respiratory tract, breasts and skin. Very rarely, it may affect the uterine cervix. We report here a case of a woman with postmenopausal bleeding for 7 months, who on physical examination presented with a vegetative and friable tumor lesion in the uterine cervix, measuring about 5 cm in diameter. The abdominal and pelvic computed tomography scans showed a hypodense and heterogeneous mass in the cervix and uterine isthmus. Histopathological examination and immunohistochemical assays confirmed the diagnosis of ACC. The different pathological aspects, therapeutic options and prognostic factors of ACC are discussed.

## Keywords

Adenoid Cystic Carcinoma, Cervix, Uterine Cervix Cancer

## 1. Introduction

Adenoid cystic carcinoma (ACC), was first described between 1853 and 1854 by three French scientists: Robin, Lorain and Laboulbène [1]. However, the first case of a cervical ACC was only reported in 1949 by Paalman and Counseller [2]. ACC is an uncommon form of malignant neoplasm that arises in secretory glands, commonly associated with major and minor salivary gland neoplasms; however, it can occur in a variety of other sites such as the respiratory tract, breasts and skin. This neoplasm is defined by its distinctive histological appearance [1].

A wide age range has been reported for adenoid cystic carcinoma, including

cases in the pediatric age group. Most individuals are diagnosed with the disease in the fourth through sixth decades of life. There is a slight female preponderance and no strong genetic or environmental risk factors have been identified. The specific molecular abnormalities that underlie this disease process are unknown [1].

Primary ACC of the cervix is uncommon, accounting for less than 1% of all cervical carcinomas, and still has an uncertain etiology [3]. In the absence of clinical trials or case reports in the literature, there are no recommended treatment guidelines for ACC of the cervix and patients are treated like any other carcinoma of the cervix. Even the prognosis is still unknown [4].

Here, we report an extremely rare case of a patient who was diagnosed with cervical ACC. The patient had given her consent for the case report to be published.

## 2. Case Report

An 82-year-old white woman came to our hospital with continuous post-menopausal vaginal bleeding for 7 months, with tiredness, inappetence, and weight loss (about 20 kg) in that period. She had no significant medical history, and one year prior to her visit, a Pap smear performed by her general practitioner was normal. At the time of the consultation, the patient denied the presence of abdominal pain or changes in urinary or intestinal habit. On speculum examination, a vegetative and friable tumor lesion was visualized in the uterine cervix, measuring 5 cm in diameter, with numerous atypical vessels. A bimanual pelvic examination showed that the vagina and the bilateral adnexal structures were normal. There was no evidence of parametrial infiltration or involvement of the rectal mucosa. Histological examination of the cervical biopsy showed a moderately differentiated adenocarcinoma with papillary and solid areas. In addition, the immunohistochemical profile revealing positivity for vimentin and P16, besides progesterone receptor positivity, supported the hypothesis of endometrial adenocarcinoma stage I.

Transvaginal ultrasonography revealed a solid expansive lesion in the cervix and an echogenic endometrium measuring 0.2 cm thick. Pelvic and abdominal computed tomography showed a hypodense and heterogeneous tumor in the cervix and uterine isthmus, measuring 5 × 5 cm. In addition, laboratory tests, mammography, and chest radiography were performed, and no metastatic dissemination was found. On the basis of the clinical picture and additional investigations, an endometrial neoplasm stage I with uterine cervix extension was initially suspected.

The patient underwent surgery for staging including radical hysterectomy and bilateral adnexectomy and bilateral pelvic lymphadenectomy. Macroscopic examination of the surgical specimen revealed a multinodular tight lesion measuring 6.0 × 5.0 × 4.5 cm, occupying about 80% of the circumference of the cervix, compromising the chorion and causing bulging of the endocervix and isthmus

(**Figure 1**). Definitive biopsy of the surgical specimen showed a 6-cm long tumor at its largest diameter, extensive solid areas and foci of necrosis. There was invasion of the uterine wall, the cervix proximally and the endometrium distally. There was also a presence of angiolymphatic tumor invasion.

Microscopy showed a proliferation of basaloid cells arranged predominantly in a cribriform pattern (**Figure 2**). Immunohistochemistry revealed positivity for CAM 5.2 and collagen IV in the hyaline material. Thus, the final diagnosis was ACC of the cervix.

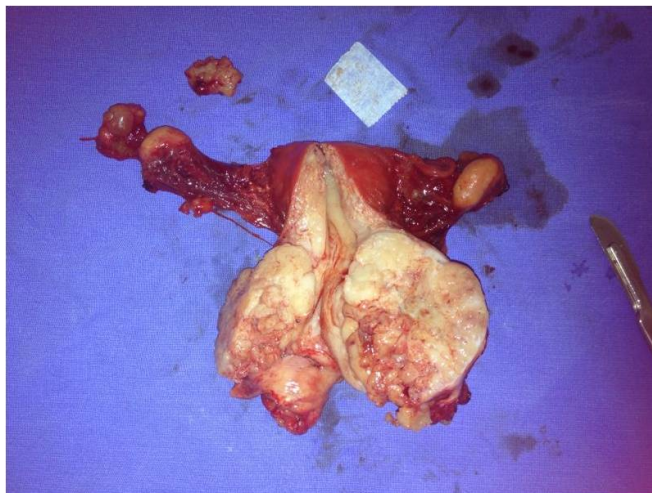
After surgery, the patient underwent 25 sessions of adjuvant radiotherapy and 5 cycles of chemotherapy with cisplatin and brachytherapy. During the 2-year follow-up after the initial treatment, the patient did not show evidence of tumor recurrence and remained asymptomatic after more than 5 years of treatment.

### 3. Discussion

ACC of the cervix is very rare and it usually occurs in postmenopausal women,



**Figure 1.** Cervical tumor after surgery.



**Figure 2.** Macroscopic examination of the cervical tumor.

especially between the sixth and seventh decade of life, but it can also occur in young women of reproductive age. Some authors report a higher prevalence of the tumor in African Americans and multiparous women. There is a strong association of ACC with HPV infection like most of the other cervical cancers [4] [5]. Although tumor etiopathogenesis remains uncertain, Shi *et al.* [6] found 27 cases described in the literature of cervical ACC associated with cervical carcinomas, supporting the hypothesis that there is a common cellular origin and/or causative agent. Probably, the chronic HPV infection [6].

In fact, some immunohistochemical evidence suggests that ACC develops from the multipotent basal cells of the cervical epithelium, which posterior squamous or glandular differentiation [7].

Clinically, as reported in our case, this tumor is often manifested by continuous or irregular vaginal bleeding. It should be noted that the presence of constitutional symptoms reported by our patient, including substantial weight loss in recent months, contributes to the diagnostic suspicion of a malignant neoplasia. A palpable mass of hard consistency, friable or not, may be a finding in the physical examination. Other clinical findings differ according to tumor extension and invasion of adjacent tissues, lymph node involvement or presence of distant metastases [4] [5]. In our case, the patient was very old and the constitutional symptoms were attributed to the old age. Even the absence of sexual partner in our patient can be considered a risk factor for the delay in diagnosis because, most of the time, vaginal bleeding starts after sexual intercourse in women with any kind of cervical cancer. Spontaneous bleeding occurs only in locally advanced tumors like the one presented above.

Histological examination of this type of tumor commonly reveals the presence of islets of pleomorphic basaloid cells in the medium of amorphous or mucinous eosinophilic hyaline material, forming adenoid structures, characterizing a cribriform pattern. In addition to this, other patterns can be described through examination, such as the tubular solid and undifferentiated pattern. The finding of areas of necrosis in the anatomopathological examination in our case, seems to be a common finding according to some authors [4] [7]. Lymphovascular invasion is also common in this type of tumor and was also detectable in our case. This common fact in this type of tumor can contribute to a worsening in prognosis [7].

On the basis of morphology, pleomorphism, mitotic index and stromal hyalinization of ACC, it is possible to distinguish it from small cell carcinoma, basal adenoid carcinoma and nonkeratinized squamous cell carcinoma, considered to be its main differential diagnoses. In addition, immunohistochemical examination of ACC reveals diffuse positivity for CD 117, cytokeratin and CAM 5.2 [7]. In the case reported, these differential pathologic aspects could be noted as described previously.

Cervical ACC is characterized by an aggressive behavior, since it has a high predisposition for the invasion of adjacent organs, lymphatic vessels and perineural spaces and subsequently for hematogenous metastasis. There is evidence

that supports the aggressive nature of this cancer, even in its early stages. A review showed that 74% (14/19) of women diagnosed with stage I ACC had lymph involvement after lymph node dissection [8]. In fact, a preliminary review of 43 cases found overall survival for patients with stage I disease to be 56% at 3 - 5 years. Even though some studies report a higher survival rate, this still appears to be considerably lower than in other types of stage I cervical tumors [9]. Thus, it seems logical that the treatment of ACC be instituted in a less conservative way than with other tumors of the uterine cervix.

Due to the rarity of this type of cervical cancer and the absence of prospective studies, no guidelines for treatment have been proposed so far. Thus, most of the patients are treated in a similar way as recommended for squamous cell cancer of the cervix. This aspect was considered in our patient and a radical treatment was performed. The management of the initial stages of the cervical ACC (stages I-II according to International Federation of Gynaecology and Obstetrics classification) involves surgical intervention, and radiotherapy is generally recommended as an adjuvant treatment. This recommendation is due to the fact that uterine cervix are considered radiosensitive tumors, and in previously reported cases, even in the early stages, better results were seen with adjuvant radiotherapy compared to surgical treatment alone [8]. The role of chemotherapy as adjunctive or primary treatment is still undefined, although in some case reports, chemotherapy has been indicated only for recurrent disease [7]. In our case, chemotherapy was used during radiotherapy.

Elhassani *et al.* reported 13 cases of cervical ACC stage IIIB, where the first case was managed with chemotherapy and concomitant radiotherapy, achieving success [4]. In contrast, Nishida *et al.* treated a case of stage IIIB only with radiotherapy; the patient did not show evidence of recurrent tumor at 5 years after such therapeutic management [5].

Considering that the main determinant of a patient's survival is metastasis, an aggressive local and systemic therapy is essential, considering local and distant recurrences. After surgery, we chose chemotherapy for our patient with weekly cisplatin during radiotherapy of 50 Gy in 25 fractions for 5 weeks and intracavitary brachytherapy at a dose of 7 Gy per fraction weekly for 3 weeks, which resulted in no recurrence during the 2-year follow-up. According to the literature, combined treatment appears to be necessary to obtain cure.

#### 4. Conclusions

Our case report showed the characteristics of an infrequent tumor in an 82-year-old woman with a 6-cm mass in the uterine cervix, initially diagnosed as endometrial adenocarcinoma, on the basis of clinical and initial examinations. The diagnosis changed completely after pathological examination and therefore, even patients with a compatible clinical setting, should be carefully investigated.

The use of radiotherapy and adjuvant chemotherapy is being evaluated in this type of tumor but since it has a poor prognosis, most of the oncologists are using

adjuvant therapy after surgery. Gynecologists should be familiar with the diagnosis of ACC and its treatment, so that treatment can be quickly initiated to limit damage to other organs and systems.

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### Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

### References

- [1] Biswas, K.D., *et al.* (2014) Unusual Presentations of Adenoid Cystic Carcinoma in Extra-Salivary Gland Subsites in Head and Neck Region: A Case Series. *Indian Journal of Otolaryngology and Head & Neck Surgery*, **66**, 286-290. <https://doi.org/10.1007/s12070-012-0490-0>
- [2] Paalman, R.J. and Counselor, V.S. (1949) Cylindroma of the Cervix with Procidencia. *American Journal Obstetrics Gynecology*, **58**, 184-187.
- [3] Sinaa, M., Oukabli, M. and Albouzidi, A. (2014) Adenoid Cystic Carcinoma of Cervix in Younger Women: Report of Two New Cases. *The Pan African Medical Journal*, **19**, 99. <https://doi.org/10.11604/pamj.2014.19.99.5241>
- [4] Elhassani, K.K., *et al.* (2009) Advanced Adenoid Cystic Carcinoma of the Cervix: A Case Report and Review of the Literature. *Cases Journal*, **2**, 6634. <https://doi.org/10.4076/1757-1627-2-6634>
- [5] Nishida, M., Nasu, K., Takai, N., Miyakawa, I. and Kashima, K. (2005) Adenoid Cystic Carcinoma of the Uterine Cervix. *International Journal of Clinical Oncology*, **10**, 198-200. <https://doi.org/10.1007/s10147-004-0466-5>
- [6] Shi, X., *et al.* (2015) 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. *Diagnostic Pathology*, **10**, 145. <https://doi.org/10.1186/s13000-015-0376-z>
- [7] Rais, M., Kharmoum, J., Ech-Charif, S. and El Khannoussi, B. (2017) Adenoid Cystic Carcinoma of the Uterine Cervix: A Report of 2 Cases. *Case Reports Pathology*, **2017**, 8401741. <https://doi.org/10.1155/2017/8401741>
- [8] Dingh, T.V. and Woodruff, J.D. (1985) Adenoid Cystic and Adenoid Basal Carcinomas of the Cervix. *Obstetrics & Gynecology*, **65**, 705-709.
- [9] Dixit, S., Singhal, S., Vyas, R., Murthy, A. and Baboo, H.A. (1993) Adenoid Cystic Carcinoma of the Cervix. *Journal of Postgraduate Medicine*, **39**, 211-215.