Clinical Case Report

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Adenoid cystic carcinoma of the vagina

A case report

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Abstract

Rationale: Squamous carcinoma is the most common malignancy of vagina. Adenoid cystic carcinoma (ACC) in the vagina is very rare.

Patient concerns: In the present study, we present a 45-year-old woman with a palpable swelling in the vagina. The patient reported body paresthesia, chest congestion, expiratory dyspnea, and itching in the thigh root.

Diagnosis: The ultrasound results revealed inhomogeneous echoes of the muscular layer in the middle and distal of the vagina, and probed a slightly richer blood flow signal. Then biopsy was performed. On microscopic examination, it was observed that tumor cells were arranged in a tubular or cribriform pattern, and exhibited a consistent size, small nuclei, and nuclear fission. The myoepithelium was lined around the glandular cavity, but the myoepithelium was tumorous. Immunohistochemistry was performed for further verification. Vimentin was positive in mesenchyme and CK-P was positive in epithelial cells. P63 and calponin were spotted, which were focal positive around the glandular cavity. Finally, the patient was diagnosed as ACC.

Interventions: At last, the patient chose chemoradiotherapy, not surgical excision.

Outcomes: The patient is alive and well 13 months after the initial diagnosis.

Lessons: ACC in the vagina is extremely rare. To our knowledge, this report is the first case of ACC arising from the vagina in English-language literature. Extensive surgical section of the tumour and chemoradiotherapy are recommended for therapy. Because of rarity, the prognosis of ACC in vagina is not known.

Abbreviation: ACC = adenoid cystic carcinoma.

Keywords: adenoid cystic carcinoma, biopsy, clinical treatment, clinicopathologic diagnosis, vagina

1. Introduction

Adenoid cystic carcinoma (ACC) has been described in many organs, such as the trachea, mammary glands, uterine cervix and, sinuses.[1–4] Indeed, it is a epithelial tumor that usually originates in the salivary glands, submandibular glands, and minor salivary glands.[5,6] This tumor is malignant, has low local recurrence, and rarely leads to distant metastasis,[7,8] but is strictly location-dependent. Typically, it consists of small basaloid cells with a solid cribriform pattern or epithelial cells with a tubular growth pattern in histology.[9] The occurrence of ACC is fairly scarce in the vagina, and it has not been reported in literature. The present report intends to share 1 case of ACC that occurred in the vagina.

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2. Case presentation

The present study was conducted in accordance with the declaration of Helsinki and with approval from the Ethics Committee of our Hospital. A written informed consent was obtained from the patient.

A 45-year-old woman complained of a palpable swelling in the vagina while taking bath 2 months ago. She reported body paresthesia, chest congestion, expiratory dyspnea, and itching in the thigh root. The outpatient examination revealed a 2 to 3 cm nodule from the vaginal orifice, which was fixed with the pubis. The nodule was approximately 2 × 3 cm in size. Biopsy by local hospital provided a description, but there was no definitive diagnosis. Therefore, the patient visited West China Second Hospital (Chengdu, Sichuan) for consultation in August 2017. An ultrasound was first performed on the patient. The results revealed inhomogeneous echoes of the muscular layer in the middle and distal area of the vagina, and a slightly richer blood flow signal (RI = 0.64, Fig. 1) was probed at the same site (Fig. 1). This result could provide guidance for consideration of a malignant tumor. However, the clinical tumor marker detection results were all within the normal range. Positron Emission Tomography-Computed Tomography (PET-CT) indicated a local malignant tumor in the left behind the pubic bone and left vagina. In addition, osteolytic lesions were found in the 10th thoracic vertebra, which was considered as bone metastasis (Fig. 2).

Pathological consultation revealed that tumor cells were arranged in a tubular or cribriform pattern, and exhibited a consistent size, small nuclei, and nuclear fission. The myoepithelium was lined around the glandular cavity. These histological features suggested a diagnosis of ACC. Immunohistochemistry

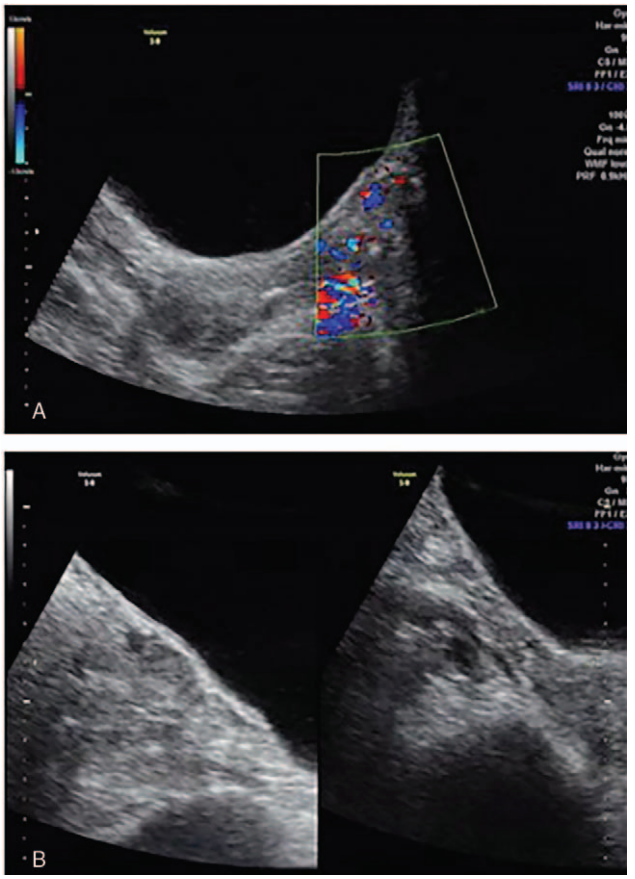

Figure 1. Ultrasonographic image showing inhomogenous echoes in the vagina (A, B).

was performed for further evidence. It was found that vimentin was positive in mesenchyme cells and CK-P was positive in epithelial cells. Furthermore, AR, CgA, Syn, CD56, S-100, calretinin, and BerEP4 were all negative, hinting that it was exactly an epithelial tumor. It is noteworthy that P63 and calponin were spotted. These were focal positive around the glandular cavity, which verified the existence of the myoepithelium. Indeed, Ki-67 was detected and positive in approximately 20% of the tumor cell nuclei. Based on the pathological report, the patient was advised by clinical doctors to receive chemo-radiotherapy (Fig. 3). The patient is alive and well 13 months after the initial diagnosis.

3. Discussion

Theodor Billroth was the first to describe ACC as cylindromas in his histological studies in 1856. The incidence of ACC is not very high, in general.[10] These are usually observed in the salivary gland. The average age of onset was 57.4 years old, and approximately 60% of patients are women.[11] ACC makes up approximately 6% of all salivary gland tumors. In addition, these make up 15% to 30% of submandibular gland tumors, 30% of minor salivary gland tumors, and 2 to 15% of parotid gland tumors.[12]

Furthermore, this has been reported in many organs in recent years. For instance, lacrimal sac tumors are rare but was reported in 1 case of ACC. The case was a 41-year-old woman with late diagnosis, who underwent ophthalmological examination and multidisciplinary treatment, and was finally confirmed as primary ACC.[13] The ACC also occurred in the female reproductive system. For instance, a 23-year-old woman was diagnosed ACC in left ovary, the tumor had the typical cribriform pattern of ACC, lacked any component of surface epithelial carcinoma, and showed myoepithelial differentiation. And there is a higher incidence in vulva of ACC according to the literature. In another example, ACC of the buccal mucosa has also been reported. Among intraoral ACC, the buccal mucosa was among the rarest sites.[14] In the present case, ACC of the vagina is also very rare. Literatures were searched, but a similar case report could not be found.

In the early stage of ACC, the most common symptom is painless mass, in which few experience this with pain. The level of

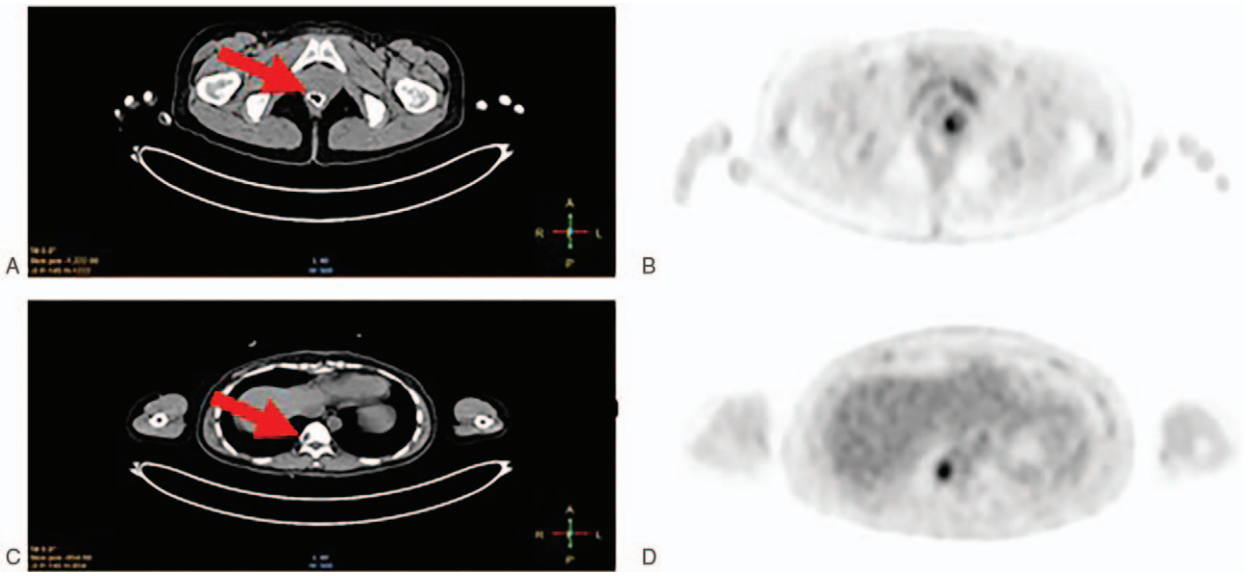

Figure 2. PET-CT scans of the lesion in the vagina (A: PET, B: CT) and the osteolytic lesions in the 10th thoracic vertebra (C: PET, D: CT). PET-CT, Positron Emission Tomography-Computed Tomography.

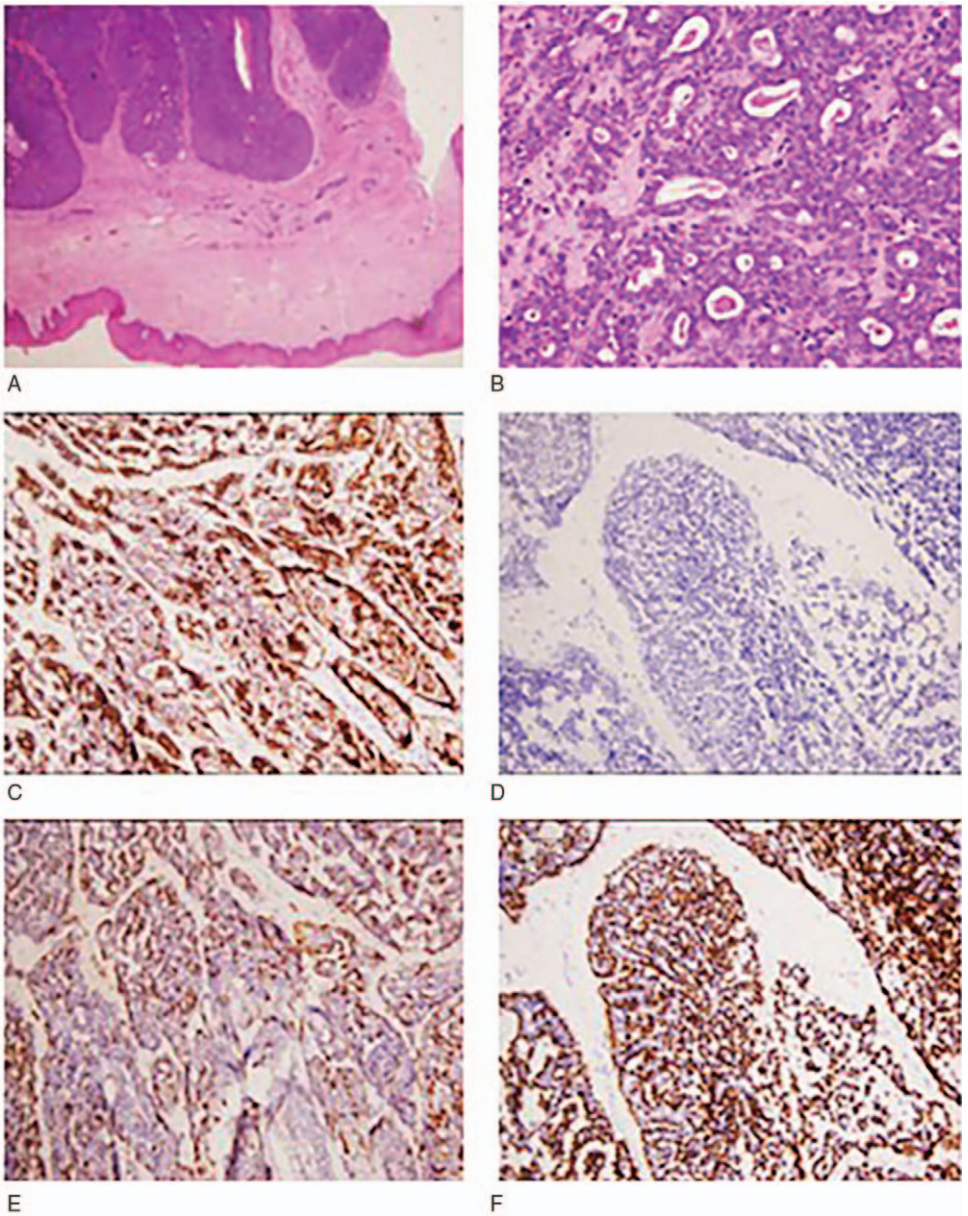

Figure 3. Hematoxylin and eosin (H&E) and immunohistochemical staining findings. H&E staining reveals the typical histological features in 40× (A) and 400× (B) microscopy. (C) Positive CK-P staining in glandular epithelial cells (200×). (D) Negative EP-CAM staining in tumor cells (200×). (E) Positive Calponin staining in myoepithelium cells (200×). (F) Positive p63 staining in myoepithelium/basal cells (200×).

pain and the process of illness are not the same in different situations. However, tumor cells always spread along the nerve. For instance, it can cause facial nerve paralysis when it occurs in the parotid gland. Correspondently, the major complaint of the patient in the present study was paresthesia of the left body. This symptom was an important tip for consideration of ACC. ACC commonly metastasizes to the lung, bone, and viscera, even with adequate locoregional control,[15] and the imaging tests of the present patient revealed that distant metastasis to bone had already occurred. The preoperative diagnosis is very hard for clinicians, in general, and most cases are dependent on biopsy. Despite its microscopic features, the gross type of ACC should also be given attention, such as painless nodules, and indurated, firm, immobile and small telangiectatic vessels.

The treatment aspect of this tumor was chiefly surgery or/and coupled with radiotherapy. Since the tumor in the present case had distant metastasis, the patient was advised to receive chemoradiotherapy before surgery. However, the ACC was unresponsive to chemotherapy, although palliative chemotherapy might have been used in symptomatic patients.[15] Hence, local mass resection is the main radical cure for ACC. Furthermore, adjuvant radiotherapy appears to be a useful way to improve locoregional control and disease-free survival.[16,17] However, extensive excision was not performed in this case of ACC considering the actual condition of the patient.

Basic studies on ACC have not received much attention at present. This has only been mentioned in one literature, in which the Notch signaling pathway, including *NOTCH1* and *NOTCH2*, mutated functionally in ACC, while *TP53*, *KRAS*, and *BRAF*, as common cancer genes, were unexpectedly identified without mutations.[18] Furthermore, ACC is often driven by the MYB-NFIB fusion gene, resulting in the overexpression of the

proto-oncogene *MYB*.[19,20] This may provide as a new marker for clinical diagnosis and treatment.

Author contributions

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