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Adenoid cystic carcinoma of the Bartholin's gland: a case report and literature review

Yanan Chang, Wang Wu and Hong Chen

Abstract

Objective: The present study was performed to explore the clinical features, diagnosis, and treatment of adenoid cystic carcinoma of the Bartholin's gland.

Methods: The clinical data of a case of adenoid cystic carcinoma of the Bartholin's gland were examined, and the clinical manifestation, diagnosis, and treatment were analyzed.

Results: Adenoid cystic carcinoma of the Bartholin's gland has a low incidence. The main clinical manifestations are vulvar lumps, dyspareunia, pain, itching, and bleeding. The diagnosis is based on gynecological examination and pathological biopsy, and the treatment is mainly surgical excision.

Conclusion: Adenoid cystic carcinoma of the Bartholin's gland is rare and lacks specificity. Pathological biopsy is the gold standard for diagnosis, and surgical resection should be performed for treatment.

Keywords

Bartholin's gland, adenoid cystic carcinoma, tumor, case report, pathological biopsy, surgical resection

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Introduction

Bartholin gland carcinoma (BGC) is a rare vulvar malignant tumor with a low incidence rate, accounting for 0.001% of female genital tumors and 27% of vulvar malignant tumors.[1] Squamous cell carcinoma and adenocarcinoma are the most common types of BGC, accounting for about 80% of all cases. Squamous

Department of Obstetrics and Gynecology, Zhongnan Hospital of Wuhan University, Wuhan, China

Corresponding author:
Hong Chen, Department of Obstetrics and Gynecology, Zhongnan Hospital of Wuhan University, No. 169 Donghu Road, Wuchang District, Wuhan City, Hubei Province, Wuhan 430071, China.
Email: znchgg@163.com

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adenocarcinoma and undifferentiated carcinoma are rare, accounting for about 20% of all cases.[2] We herein report a case of adenoid cystic carcinoma (ACC) of the Bartholin's gland. BGC is very rare in the clinical setting, and this case may help us to further recognize BGC and might provide some value for the diagnosis and treatment of BGC.

Case report

A 49-year-old postmenopausal woman was referred to Zhongnan Hospital affiliated to Wuhan University (Wuhan, China) with a >3-year history of BGC. Written informed consent was obtained from the patient, and this study was conducted in accordance with the Declaration of Helsinki and with approval from the Ethics Committee of Zhongnan Hospital. About 3 years previously, the patient had presented with a hard, fixed, tender black nodule with obvious attachment in the left vulva. She was then referred to our hospital. Pelvic examination showed a nodule over the left vulva measuring 1.5×1.5 cm. Physical examination revealed no other abnormalities. Chest X-ray findings and hematological, renal, and liver function were all normal. The serum level of cancer antigen 125 (CA125) was elevated at 75.9 U/mL. The pathological results of a vulva biopsy suggested ACC. Pelvic computed tomography (CT) examination revealed a mixed-density shadow on the left side of the perineum with a gas-density shadow inside and multiple lymph nodes in the bilateral groin. Magnetic resonance imaging also showed multiple lymph nodes in the bilateral groin. The initial diagnosis was a vulvar tumor, possibly BGC

On September 25, 2014, the patient underwent general anesthesia with partial vulvectomy and left inguinal lymph node dissection. The postoperative pathological finding was a left adenoid mass (ACC) with no left inguinal femoral lymph node metastasis. No adjuvant radiotherapy or chemotherapy was performed. The incision healed well after the operation. The wound size was about $5 \times 5 \times 4$ cm. The patient was discharged 2 weeks after the operation.

About 1.5 years after the surgery, the patient was admitted to Zhongnan Hospital because of the development of a left genital mass. A gynecological examination revealed a 5- × 4-cm hard, uneven, tender mass in the left genital vestibular gland. Vulvar B-ultrasound examination showed an area of low echogenicity measuring about 4.1×1.8 cm under the left vulva; its outline was unclear and shape was irregular, and no obvious color flow signals were present inside. Pelvic CT showed a low-density shadow on the left side of the vulva, and the border was unclear. A vulvar cyst was considered. The serum CA125 level was high at 65.8 U/mL. The patient was preliminarily diagnosed with a left vaginal wall cyst after the surgical treatment of BGC. On 7 April 2016, the left vulvar mass was removed under general anesthesia. Postoperative pathological examination revealed a Bartholin ACC, as shown in Figure 1. Immunohistochemical examination of the ductal epithelium revealed the following: carcinoembryonic antigen (+), cytokeratin (+), epithelial membrane antigen (EMA) (+), myoepithelial cells were p53 (+), p63 (+), S-100 (−), smooth muscle actin (SMA) (+), vimentin (+), calponin (−), and CD117 (−). The Ki-67 positivity rate was about 30%, and no tumor was found in the fibrous connective tissue at the incisional margin of the mass, as shown in Figure 2. Two weeks after surgery, the patient had recovered well. She was then discharged from the hospital and diagnosed with a vulvar malignant tumor (Bartholin ACC).

On 14 March 2018, the patient returned Zhongnan Hospital because of vulvar discomfort. Gynecological examination

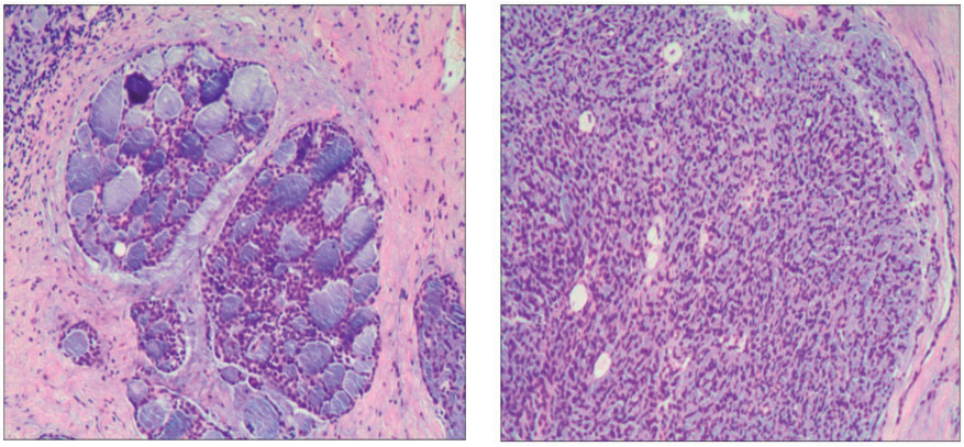

Figure 1. Left-sided Bartholin adenoid cystic carcinoma. The tumor cells were irregularly arranged with large, dark nuclei and sieve-shaped glands. Hematoxylin and eosin staining, × 400.

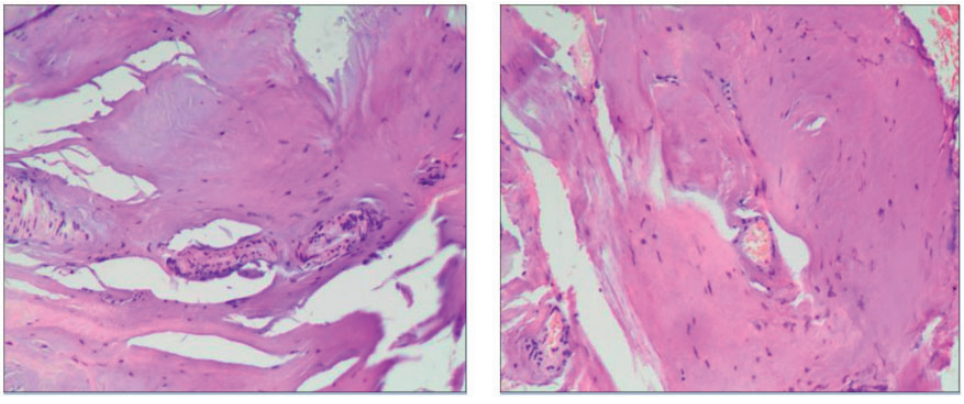

Figure 2. No tumors were found in the fibrous connective tissue of the incised margin. Hematoxylin and eosin staining, × 400.

revealed a tender mass of about 2 cm in diameter that could be palpated from the left vaginal wall. Pelvic CT showed irregular thickening of the soft tissue of the left perineum. The serum CA125 level was high at 69.9 U/mL; all other examination results were normal. The patient underwent general anesthesia for vulvovaginal tumor resection on 16 March 2018. Postoperative pathological examination revealed ACC (upper and lower vaginal tumors, posterior combined tumors), glandular epithelial calponin (+), CD117 (+), EMA (+), and p63 (+). The patient recovered well after surgery and was still clinically well at the time of this writing.

Discussion

Vulvar Bartholin ACC is extremely rare in clinical practice. Seventy-seven cases have been reported to date, accounting for

29.6% of vulvar BGC.[3] This tumor usually occurs in postmenopausal women, and the age at onset is around 50 years.[4] The pathogenesis of Bartholin ACC remains unclear. Some reports have indicated that this tumor may be related to vulvar and Bartholin gland infection, especially human papillomavirus infection.[5] The clinical manifestations lack specificity and include vulvar contact with a mass, difficulty in intercourse, pain, itching, and bleeding. Among these, a vulvar mass is the most common symptom. Bartholin ACC contains glandular, myofiber, and epithelial components. Pathological examination shows that the tumor cells are round and uniform in size with deeply stained nuclei. The noncellular area contains mucin and hyaluronic acid with duct-like or glandular cavities. Sieve-like dilation and peripheral nerve infiltration are its main features.[6] The immunohistochemistry results are mostly positive for AE1/E3, CK8/18, EMA, SMA, smooth muscle myosin, p63, S-100, CD43, CD177, and carcinoembryonic antigen.[6] Immunohistochemical examination of the excised tumor in the present case revealed p53 (+), p63 (+), SMA (+), and vimentin (+), consistent with an adenoid tumor. Bartholin ACC grows slowly and exhibits local infiltration and an early tendency to invade the nerve. Tumors may recur locally and metastasize to the lung, bone, liver, and other organs. They may also metastasize to the inguinal or pelvic lymph nodes. The most common site of metastasis is the lung. Because of the rarity of Bartholin ACC, no definite recommendation for the best treatment has been established. If the lesion is small, unilateral, and not close to the midline, the preferred treated is simple mass resection with a negative incisional margin. If the lesion is extensive, extensive radical vulvectomy is needed. No consensus has been reached on the need for lymph node dissection. Our patient underwent left lymph node dissection because of an inguinal lymph node shadow on CT examination. In a study by Yang et al.,[7] the positive rate of simple tumor resection was about 48%, and the radical resection rate was 30%. Additionally, 68.9% of the patients developed recurrence after resection of the lesion, and the recurrence rate after radical resection of the vulva was 42.9%. Our patient developed relapse more than 1 year after the first local mass excision. According to Copeland et al.,[8] patients with adenoid cystadenocarcinoma have a tumor-free rate of about 47% at 5 years after surgery, and the 5-year survival rate is about 71%. Another report indicated that postoperative adjuvant radiotherapy can help to reduce the recurrence of positive margins, but Bernstein et al.[9] stated that Bartholin ACC may be less sensitive to radiotherapy. There is a lack of information about whether chemotherapy is available after surgery. Chemotherapy is an option for patients with distant metastasis; however, the specific chemotherapeutic drugs are not yet clear. Azithromycin can reportedly induce the degeneration of salivary gland cystic adenoid carcinoma cells, but whether adriamycin has a curative effect on Bartholin ACC has not been determined.[1] The average survival time of patients with Bartholin ACC is 5 to 10 years. The overall survival rate after surgery is quite variable and is related to the tumor stage and margin status. The survival time of patients with a positive margin is <7 years, and the survival time of patients with a negative margin is 15 to 30 years longer than that of patients with a positive margin.[10]

In conclusion, Bartholin ACC grows slowly, and surgical resection is the preferred treatment. Adjuvant radiotherapy or chemotherapy can be used for patients with deep local infiltration, a positive incisional margin, or recurrence. Reasonable treatment options can provide a better prognosis.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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ORCID iD

Yanan Chang https://orcid.org/0000-0002-8845-2575

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